Atlas of Adolescent Dermatology

Patricia Treadwell
Julie Prendiville
Michael Lee Smith
Editors

Second Edition

Springer

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ISBN 978-3-031-86176-5 ISBN 978-3-031-86177-2 (eBook)
https://doi.org/10.1007/978-3-031-86177-2

This Springer imprint is published by the registered company Springer Nature Switzerland AG
The registered company address is: Gewerbestrasse 11, 6330 Cham, Switzerland

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Part I
Acne and Perioral Dermatitis

Chapter 1
Acne

Michael Lee Smith

Introduction

Acne vulgaris is a chronic inflammatory dermatosis with a complex interplay of four known pathogenic factors. There is follicular hyperkeratinization leading to follicular plugging in the pilosebaceous unit (comedones). This is coupled with an androgen-stimulated increase in sebum production and alteration of the sebum composition. Proliferation of *Cutibacterium acnes* and an inflammatory response complete the tetrad.

Epidemiology

Acne vulgaris is the most common skin disorder in the United States, and affects about 9% of the world population. The most common ages are adolescence and young adulthood. It affects approximately 85% of individuals aged 12–24 years and almost 50% of individuals aged 20–29 years.

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_1

Clinical Findings

The characteristic lesions of acne are blackheads (open comedones) (Fig. 1.1), whiteheads (closed comedones), papules (Fig. 1.2), pustules, and nodules (Fig. 1.2). The distribution tends to be in areas of greatest density of sebaceous glands, which are the face, the upper back, and the chest. A combination of lesion types may be present in any particular area.

Laboratory

Diagnosis is most often based on clinical findings. No laboratory test is pathognomonic.

Treatment

This publication is not intended as a comprehensive discussion of all available treatments. A summary below will list many of the therapeutic options. The reader is directed to the references below for guidelines for treatment, including algorithms, mechanisms of action and side effects.

Many patients have already used OTC products when they arrive at the office. Nonprescription recommendations include gentle cleansing, use of non-comedogenic moisturizers and sunscreens, and frequent pillowcase changes.

Acne prescription treatment regimens may include any of the following:

Topical retinoids
Topical salicylic acid

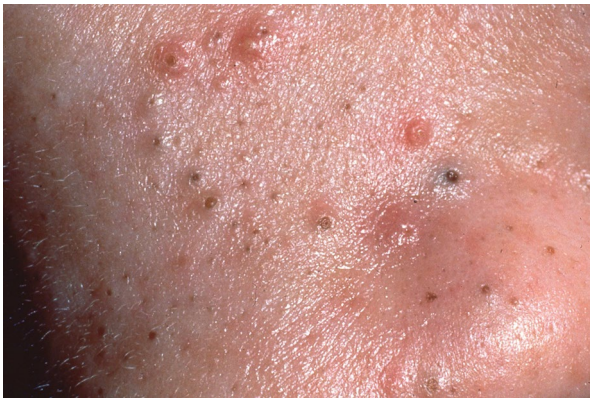

Fig. 1.1 Open comedones and red papules

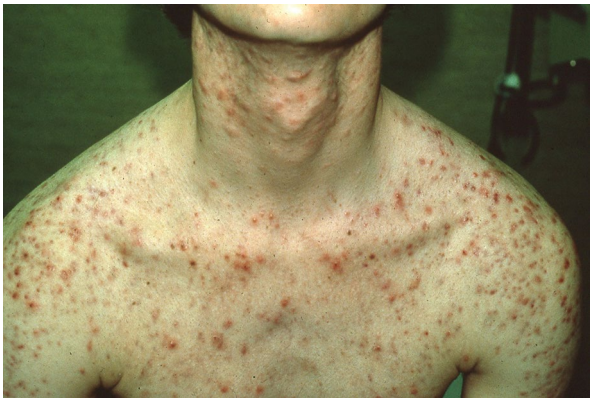

Fig. 1.2 Papules and nodules

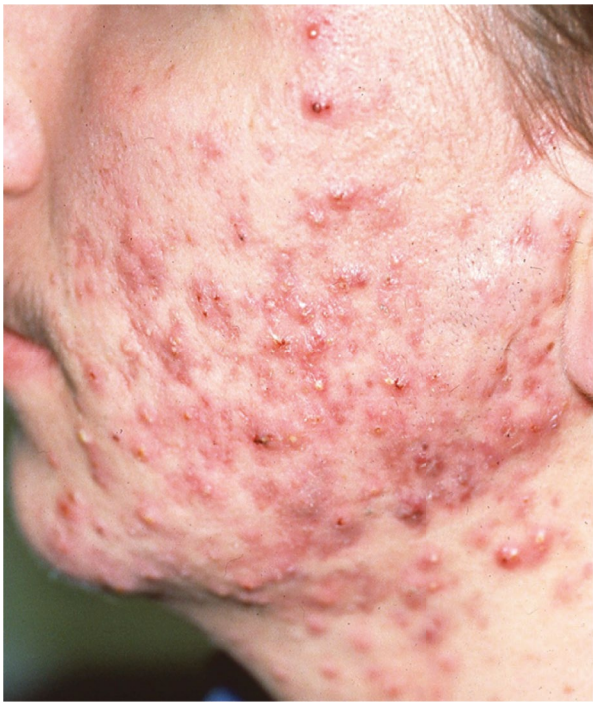

Fig. 1.3a Pre-isotretinoin

Topical antibacterial agents such as benzoyl peroxide or topical antibiotics
Systemic antibiotics
Hormonal therapies
Oral isotretinoin (Figs. 1.3a and 1.3b)

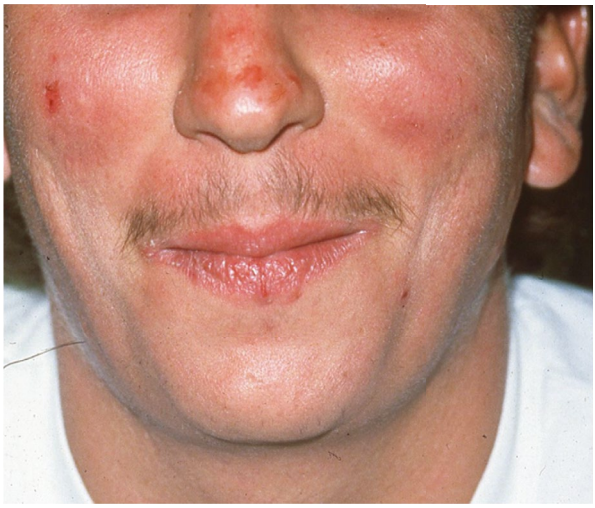

Fig. 1.3b Post-isotretinoin

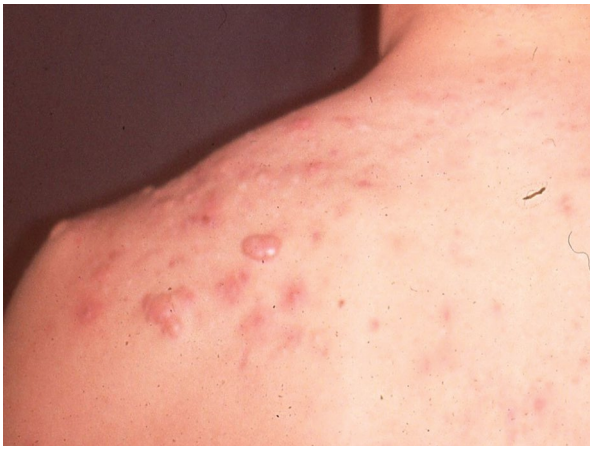

Fig. 1.4 Keloidal scarring after nodular acne

Prognosis

Prompt treatment of acne can minimize long-term scarring (Fig. 1.4). Post-inflammatory hyperpigmentation (especially in skin of color) is particularly bothersome. This can be treated in addition to the use of meticulous sun protection.

Further Readings

1. Eichenfield DZ, Sprague J, Eichenfield LF. Management of acne vulgaris: a review. JAMA. 2021;326(20):2055–67.
2. Harper JC. Acne vulgaris: what's new in our 40th year. J Am Acad Dermatol. 2020;82(2):526–7.
3. Thiboutot D, Dréno B, Sanders V, Rueda MJ, Gollnick H. Changes in the management of acne: 2009–2019. J Am Acad Dermatol. 2020;82:1268–9.

Chapter 2
Perioral Dermatitis

Michael Lee Smith

Introduction

Perioral (aka periorificial) dermatitis is an acneiform inflammatory eruption of the face that has been associated with a variety of triggers, including topical steroids; inhaled steroids used for allergic rhinitis or asthma; toothpastes containing cinnamon, mint, or fluoride; chewing gums; and some sunscreens. A large number of other putative causes include infectious agents like *Demodex*, cosmetics, hormones, and systemic steroids.

Epidemiology

Although perioral dermatitis has been reported in childhood, adolescence, and adulthood, the more common presentation is in adolescents and young adults. There seems to be a slight female predominance, but it is not significant. In younger patients, particularly with skin of color, a granulomatous variant is possible.

Clinical Findings

Discrete small papules and papulopustules are found in clusters around the perioral area (Figs. 2.1 and 2.2). Mild superficial scaling is seen in some patients. There is often an area of sparing at the vermillion border, and lip vermillion is not involved.

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_2

There may be involvement of the perinasal and periorbital areas, leading to the diagnostic term of periorificial dermatitis. There is usually very little pruritus, but we do have teens complain of mild stinging at times.

A variant of perioral dermatitis seen mostly in preadolescent children is granulomatous periorificial dermatitis. It may extend into early adolescence. It typically consists of shiny, firm, 1–2 mm erythematous to skin color to yellowish-brown papules in all periorificial locations (Fig. 2.3). This may be synonymous with Facial Afro-Caribbean Eruption. Extra-facial lesions may be seen in other areas such as the neck, trunk, and genital area. The facial lesions are rarely symptomatic, but the genital papules may be painful.

Laboratory

The diagnosis is most often made from the characteristic clinical findings. If a biopsy is performed, histopathology shows perivascular and perifollicular lymphocytic infiltrates. In the granulomatous form, additional findings include giant cells and dermal granulomas. The granulomas often have a sarcoid-like appearance.

Treatment

Avoidance of topical corticosteroids or weaning the potency of any topical corticosteroid being applied is useful. Treatment options include the following: (1) topical metronidazole, (2) topical and/or systemic antibiotics, or (3) topical calcineurin inhibitors.

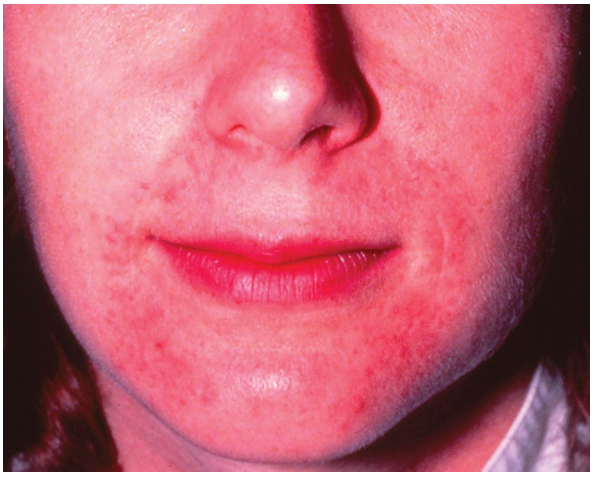

Fig. 2.1 Erythematous papules with scale seen in perioral dermatitis

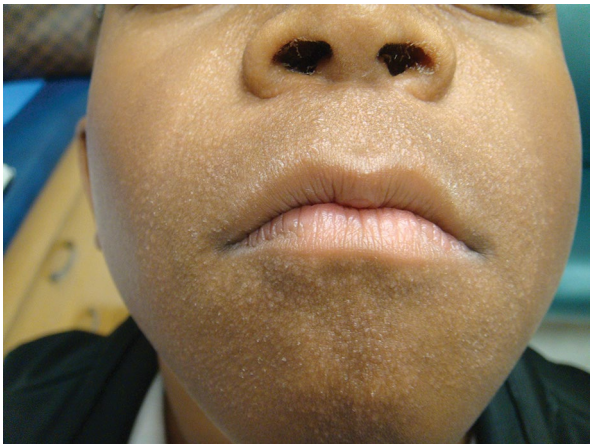

Fig. 2.2 Skin colored discrete papule with scale in perioral dermatitis

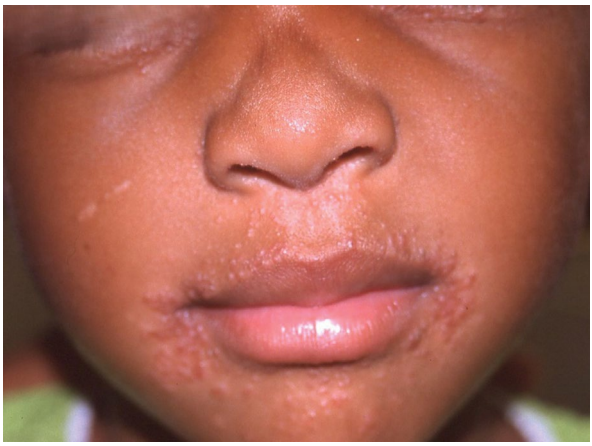

Fig. 2.3 A nine-year-old with granulomatous periorificial dermatitis

In addition, a non-comedogenic moisturizer can be helpful to decrease the amount of visible scale.

Prognosis

Generally, improvement is more rapid with treatment. However, recurrences may be noted, especially upon repeated exposure to the triggering agents.

Suggested Readings

1. Ollech A, et al. Topical calcineurin inhibitors for pediatric periorificial dermatitis. J Am Acad Dermatol. 2020;82(6):1409–14.
2. Searle T, et al. Perioral dermatitis: diagnosis, proposed etiologies, and management. J Cosmet Dermatol. 2021;20:3839–48.
3. Tempark T, et al. Perioral dermatitis: a review of the condition with special attention to treatment options. Am J Clin Dermatol. 2014;15:101–13.

Part II
Cutaneous Infections and Infestations

Chapter 3
Meningococcal Infections

Patricia Treadwell

Introduction

The organism *Neisseria meningitidis* can cause meningitis, septicemia, and, less commonly, pneumonia in adolescents. Of the patients who develop meningococcemia, the majority will have cutaneous manifestations.

Epidemiology

Adolescents are a group that has a higher incidence of risk for *Neisseria meningitides* infections. Approximately 85% of the cases in adolescents and young adults are caused by serogroups B, C, W, Y, or nongroupable meningococci [1].

Meningococcal infections are spread through respiratory droplets. Household and other close contacts are affected at a much higher rate than other contacts. The length of the incubation period is 1–10 days.

Clinical Findings

Initially, the symptoms may be nonspecific, e.g., fever, headache, malaise, myalgias, and/or cold hands and feet. Some patients may have meningismus, photophobia, and/or vomiting (findings in meningitis). Patients are typically ill-appearing.

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_3

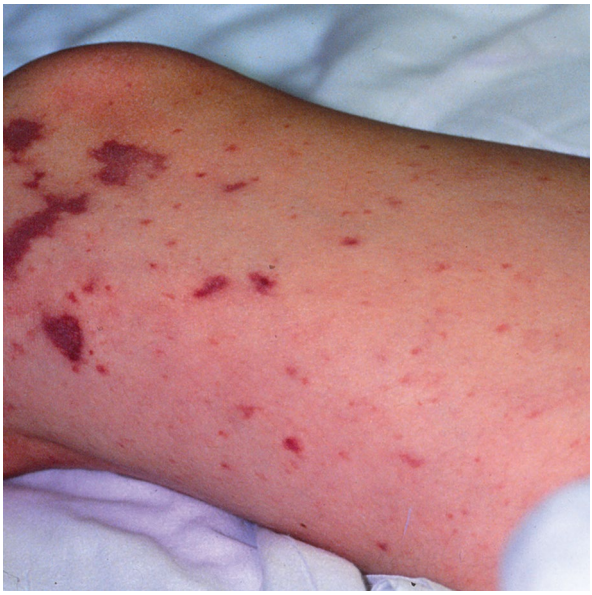

Fig. 3.1 Purpuric lesions with jagged borders

Cutaneous manifestations may be morbilliform macules and papules in the beginning, and then later petechiae, purpura, vesicles, or pustules are noted. Characteristically, the purpuric lesions have jagged edges (Fig. 3.1). Necrotic lesions and ulcers may develop (Fig. 3.2).

With further progression, patients may develop hypotension, shock, or disseminated intravascular coagulation (DIC). Purpura fulminans (Fig. 3.3) is a term used to describe DIC accompanied by necrotic and purpuric plaques.

Laboratory

Positive blood and/or cerebrospinal fluid cultures can establish the diagnosis. Some public health or research laboratories may have PCR assays.

Treatment

Per the 2024 *Red Book: Report of the Committee on Infectious Diseases* (www.aapredbook.org) treatment recommendations are as follows: -Empiric therapy with ceftriaxone or cefotaxime is recommended. Once a microbiological diagnosis is established, intravenous penicillin G is recommended at a dose of 300,000 U/kg per day up to a maximum of 12 million units per day divided every 4–6 h for 5–7 days. Cefotaxime, ceftriaxone, and ampicillin are acceptable alternatives. In a patient

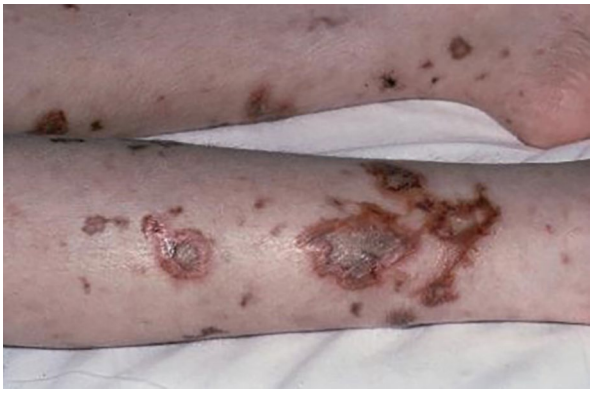

Fig. 3.2 Progression of meningococcal lesions with crusting

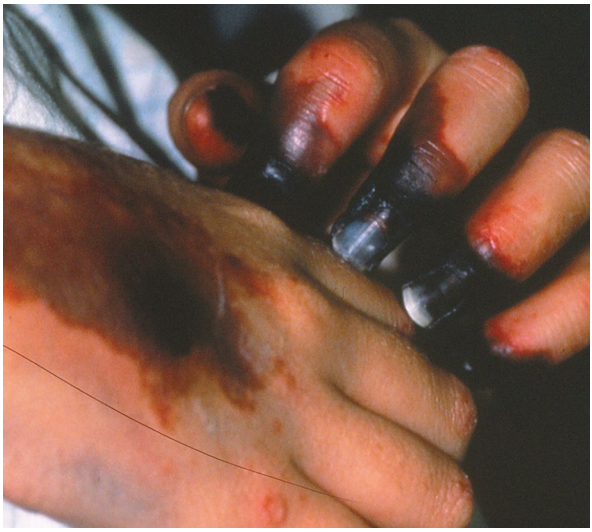

Fig. 3.3 Purpura fulminans in a patient with meningococcemia

with a life-threatening anaphylactic penicillin allergy, meropenem or ceftriaxone can be used, recognizing that the rate of cross-reactivity in penicillin-allergic adults is low. Consultation with a pediatric infectious disease specialist is recommended.

- Intermediate penicillin resistance is an increasing concern (especially in travelers from areas where penicillin resistance has been reported); as a result, some recommend using ceftriaxone, cefotaxime, or chloramphenicol until susceptibilities are available.
- A quadrivalent conjugate meningococcal vaccine immunization is now routinely recommended beginning at age 11 years. It can be used to prevent infection in *high-risk* groups from age 2 months to 55 years.
- A meningococcal B vaccine is routinely recommended for ages 16–18 years [2]. For *high-risk* groups (such as sickle cell anemia patients), individuals should receive the vaccine starting at age 10 years.

- In addition, supportive therapy (such as vasoactive agents and fluids) may be necessary.
- Close contacts within 5–7 days prior to the onset of illness (e.g., household, child care, slept, or ate in the same dwelling) should receive chemoprophylaxis.

Prognosis

Invasive meningococcemia has a mortality of approximately 15%. Morbidities include limb or digit amputations, skin scarring, neurologic sequelae, and hearing loss.

References

1. Red Book. Report of the committee on infectious diseases 2024–2027. 33rd ed. Itasca, IL: Published by the American Academy of Pediatrics; 2024. p. 585–600.
2. Parikh SR, et al. The everchanging epidemiology of meningococcal disease worldwide and the potential for prevention through vaccination. J Infect. 2020;81:488–98.

Chapter 4
Herpesvirus Infections

Patricia Treadwell

Introduction

The viruses in the herpesvirus group include Herpes Simplex, Varicella- Zoster, Epstein Barr, and Cytomegalovirus. This chapter will address both Herpes Simplex and Varicella-Zoster viruses. Herpes Simplex cutaneous lesions are caused by the herpes simplex virus. Herpes zoster and varicella are caused by the virus Varicella-Zoster.

Epidemiology

The lesions of cutaneous Herpes Simplex virus can occur at any age. Herpetic gingivostomatitis is more common in younger children. Reactivation of the lesions can occur following the initial simplex infection. Genital herpes lesions are generally more common in adolescents and adults than in young children. Herpes zoster is seen at any age; however, it is more common in adolescents than in young children and most common in adults.

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_4

Clinical Findings

Herpes Simplex—Cutaneous lesions consist of grouped vesicular lesions with an erythematous surround. Lesions may be located anywhere, including mucous membranes. When the vesicles rupture, the ulcers are deep-seated (occurring at the subepidermal level). Crusting can be noted (Fig. 4.1). Some vesicles may become pustular. Regional lymphadenopathy is often noted.

Reactivation of the lesions may be accompanied by a prodrome of burning, itching, or stinging.

Herpes Zoster is characterized by lesions similar to those in Herpes Simplex; however, they are arranged in a dermatomal pattern involving usually 1–3 dermatomes (Fig. 4.2). Pain, burning, or itching may occur prior to the onset of visible lesions. Occasionally, systemic viremia may be present.

Laboratory

Herpes Simplex and Herpes Zoster lesions can be diagnosed by the clinical appearance. If confirmation is needed, the virus can be identified from a specimen retrieved from the base after de-roofing an intact vesicle. The specimen can be submitted for PCR, fluorescent antibody, or culture (Herpes Simplex virus grows more reliably

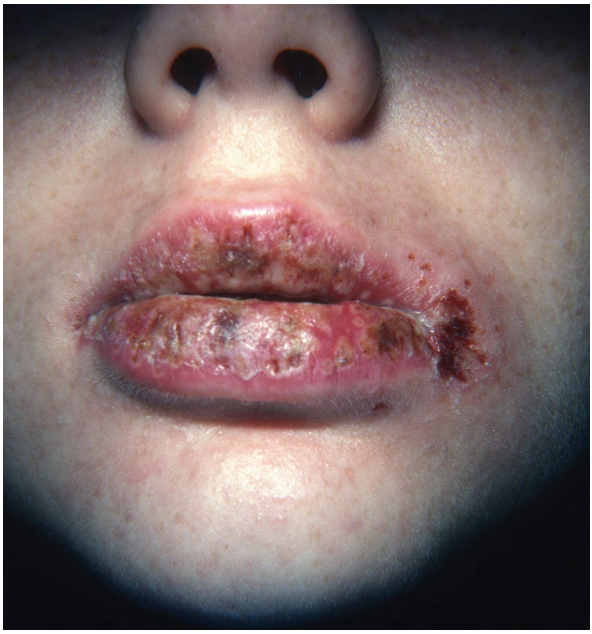

Fig. 4.1 Herpes simplex of the lips. Ulcers and crusting are noted

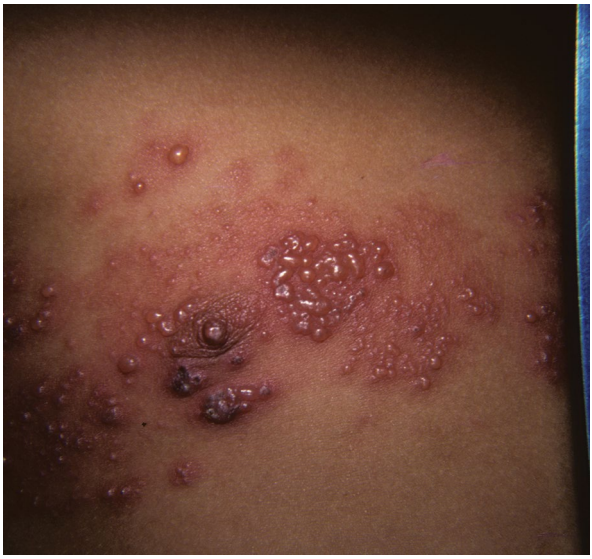

Fig. 4.2 Herpes zoster of the right chest

than Varicella Zoster virus). Both viruses are present in the lesions and are contagious to susceptible individuals.

Treatment

Herpes Simplex virus-cutaneous lesions are acutely treated with analgesics and prevention of secondary bacterial infection. Recurrent lesions can be treated with topical docosanal or topical penciclovir (both used at home as soon as the prodrome begins). If recurrent lesions are occurring more often than every 4–6 weeks or are occurring in an immunosuppressed patient, consider systemic antiviral treatment either episodically or on a suppressive basis.

Varicella Zoster lesions can be treated with analgesics and prevention of secondary bacterial infections. If the lesions continue to enlarge and spread even after several days or are occurring in an immunosuppressed patient, systemic antivirals may be necessary. Post-herpetic neuralgia is less common in children and adolescents than in older adults; thus, systemic steroids are not prescribed as often as with older adults.

Prognosis

In immunocompetent individuals, recurrent Herpes Simplex lesions tend to be confined to a specific area. Recognizing triggers (e.g. fever, illness, menses, sun exposure, etc.) can be useful—triggers can sometimes be avoided to decrease the frequency of recurrences. In immunocompromised individuals, the lesions can be widespread and cause systemic issues. In addition, any active lesions can be contagious for susceptible individuals.

In adolescents, Herpes Zoster can sometimes be a signal of an immune issue. Considering this, further work-up should be initiated as indicated. As mentioned above, post-herpetic neuralgia is less common in children and adolescents versus adults. Scarring may be noted (Fig. 4.3). Most typically, Herpes Zoster occurs only once. In addition, any active lesions can be contagious to susceptible individuals.

Suggested Readings

1. Chang JY, et al. A narrative review of alternative symptomatic treatments for herpes simplex virus. Viruses. 2023;15(6):1314.
2. Tayyar R, et al. Herpes simplex virus and varicella zoster virus infections in cancer patients. Viruses. 2023;15(2):439.

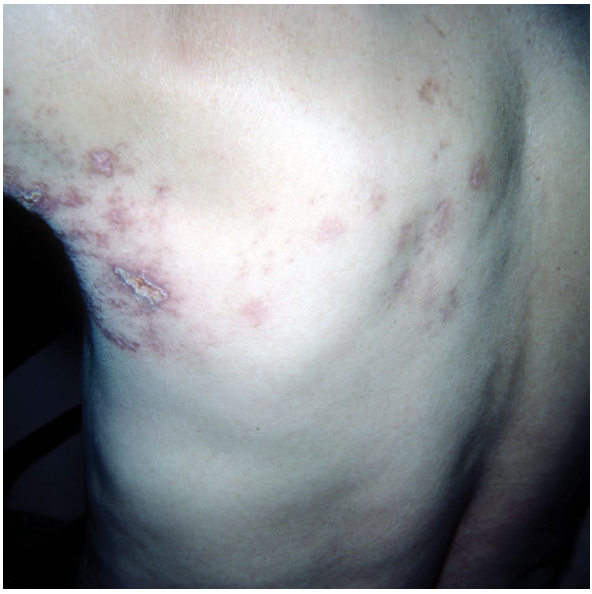

Fig. 4.3 Scarring following herpes zoster

Chapter 5
Tinea Infections

Patricia Treadwell

Introduction

Infections caused by dermatophytes are known as tinea infections. They are specifically named by the areas in which they are located. In adolescents, some of the most frequent infections are tinea corporis (including tinea faciei), tinea pedis, and tinea cruris.

Epidemiology

Tinea corporis can affect any age group. It is most often found on exposed skin, but not exclusively. Typical etiologic organisms are trichophyton species and microsporum species. Tinea pedis (dermatophytes affecting the feet) is seen most commonly in adolescents and adults, and it tends to be uncommon in prepubertal children. Typical etiologic agents are *Trichophyton rubrum, Trichophyton interdigitalis, Trichophyton mentagrophytes*, and *Epidermophyton floccosum*. Tinea cruris often has an onset in adolescence.

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_5

Clinical Findings

Tinea corporis: erythematous circular or annular lesions that have accompanying scale. (Fig. 5.1) May be itchy. Less commonly, the lesions may become vesicular or pustular (Fig. 5.2).

Tinea pedis: scaly patches of the feet, especially noted in the toe web spaces (Fig. 5.3); sometimes develops vesicles. A secondary bacterial infection may become superimposed.

Tinea cruris: Scaly patches of the inguinal areas. Often is itchy.

Laboratory

The diagnosis is usually made based on the clinical findings. However, a positive potassium hydroxide preparation and/or positive culture can confirm the diagnosis.

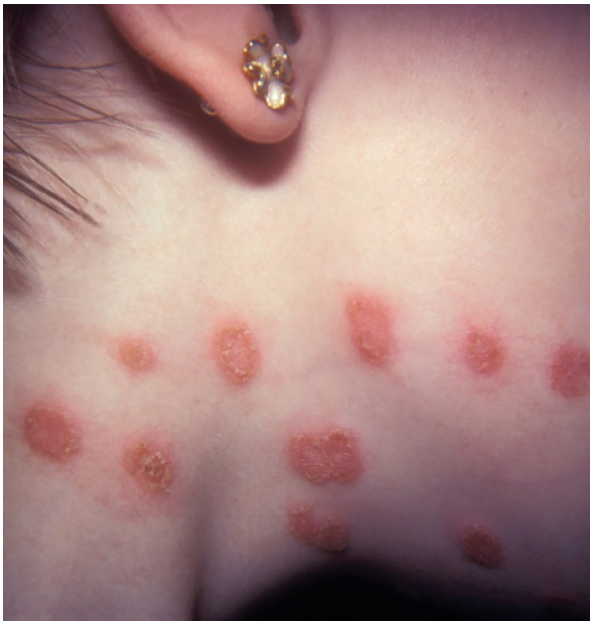

Fig. 5.1 Multiple tinea corporis lesions of the right neck and mandibular areas due to contact with an affected kitten

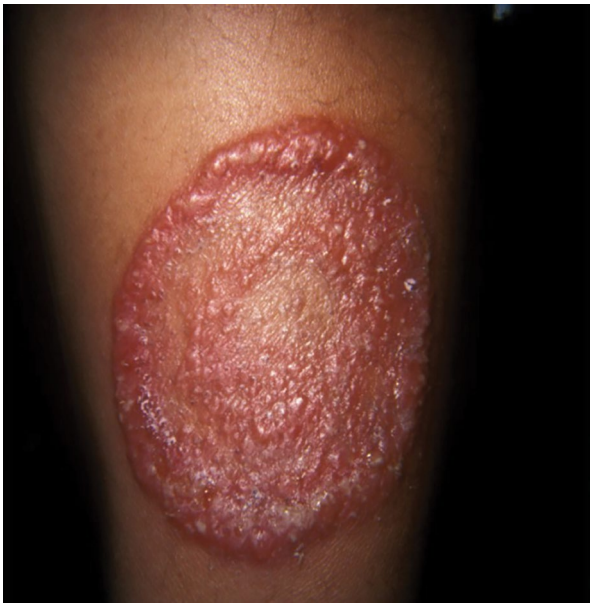

Fig. 5.2 Vesicular tinea corporis lesion of the arm

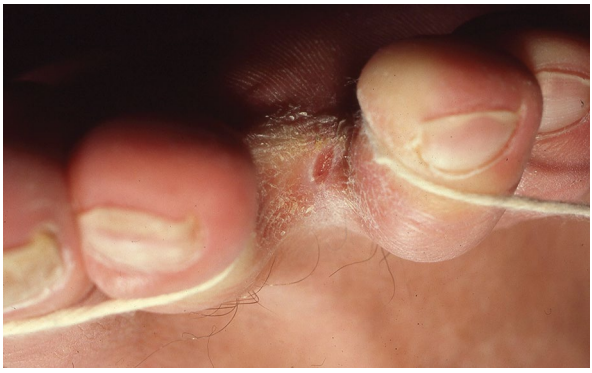

Fig. 5.3 Scaly patch of toe web in tinea pedis

Treatment

Topical antifungals (e.g., terbinafine, clotrimazole, miconazole, etc.) are applied bid for 10–14 days. If the lesions are widespread, involve a hair-bearing area, or have significant inflammation, oral medications are prescribed. Griseofulvin-2–3 week course; Fluconazole-1–2 week course; Itraconazole-1–2 week course; Terbinafine-1–2 week course.

Treating any hyperhidrosis accompanying tinea pedis may be useful as adjunctive therapy.

Prognosis

Prognosis is good with treatment; however, with tinea corporis dyspigmentation may occur in skin of color.

Suggested Readings

1. Kovitwanichkanont T, et al. Superficial fungal infections. Aust J Gen Pract. 2019;48:706–11.
2. Pei Y, et al. Tinea Faciei. J Pediatr. 2022;250:108–9.
3. Woo TE, et al. Diagnosis and management of cutaneous Tinea infections. Adv Skin Wound Care. 2019;32:350–7.

Chapter 6
Tinea Versicolor

Patricia Treadwell

Introduction

Tinea versicolor (aka pityriasis versicolor) is a superficial fungal infection caused by *Malassezia* species (formerly *Pityrosporum* species). The yeast forms reside in healthy skin. The clinical lesions of tinea versicolor result from the conversion of the yeast forms to the mycelial forms [1].

Epidemiology

Tinea versicolor can occur in any age group but is primarily seen in adolescents and adults.

An increased incidence is noted in the summer and also in patients with diabetes, pregnancy, and immunosuppression.

Clinical Findings

The lesions are slightly raised, scaly macules and patches that can be hypopigmented [3], reddish-brown color (Fig. 6.1), and/or hyperpigmented (Fig. 6.2). The most typical locations for tinea versicolor lesions are the upper back, chest, neck, and/or proximal arms. These areas have the highest sebum concentration.

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_6

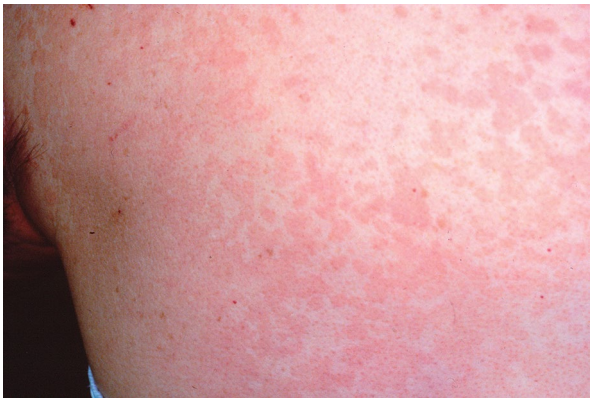

Fig. 6.1 Reddish-brown fairly flat lesions of the back with thin scale

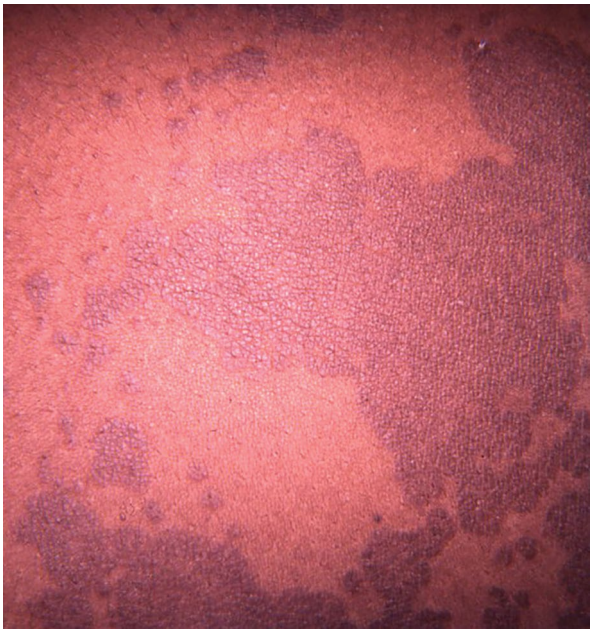

Fig. 6.2 Hyperpigmented tinea versicolor lesions in skin of color. Some areas have become confluent

Laboratory

Diagnosis is most often made with a potassium hydroxide plus India ink preparation of the scale. Microscopic examination shows spores and short hyphae ("spaghetti and meatballs"). Wood's light examination can show yellow-orange fluorescence.

Treatment

A variety of treatment regimens have been recommended for tinea versicolor.
Topical:

1. Topical anti-fungal products
2. Ketoconazole shampoo or selenium sulfide lotion/shampoo
3. Keratolytics—e.g. benzoyl peroxide

Oral: varied regimens in literature—see ref. [2]

1. Ketoconazole
2. Fluconazole
3. Itraconazole

Prognosis

Recurrences are common, and retreatment may be necessary each spring/summer. Occasionally the pigmentary changes will persist despite resolution of the scale following treatment.

References

1. Brandi N, et al. Tinea versicolor of the neck as a side effect of topical steroids for Alopecia Areata. J Dermatolog Treat. 2019;30:757–9.
2. Leung AKC, et al. Tinea versicolor: an updated review. Drugs Context. 2022;11:2022-9-2.
3. Ricles VR, et al. Tinea versicolor in an infant. J Pediatr. 2024;267:113920.

Chapter 7
Lice

Julie Prendiville

Introduction

Lice infestation in children and adolescents is mainly caused by the head louse (*Pediculus humanus capitis*). The pubic or crab louse (*Phthirus pubis*) is transmitted by sexual contact or by co-sleeping. Infestation with the clothing louse (*P. humanus humanus*) occurs in homeless populations.

Epidemiology

Head louse infestation (pediculosis capitis) is common in school-aged populations worldwide. All socioeconomic groups are affected. It is spread primarily by head-to-head transmission and possibly also from fomites (sharing of combs, hairbrushes, hats, pillows). There is an apparent increased prevalence in girls. Head lice may be associated with secondary infection of the scalp by *Staphylococcus aureus* and/or Group A streptococcus.

Clinical Findings

Pruritus is the main symptom but can be absent initially. Head lice may also present with impetigo of the scalp and occipital/cervical lymphadenopathy. Eggs, or nits (white empty egg cases) (Fig. 7.1), are found cemented to the hair shafts, often at

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_7

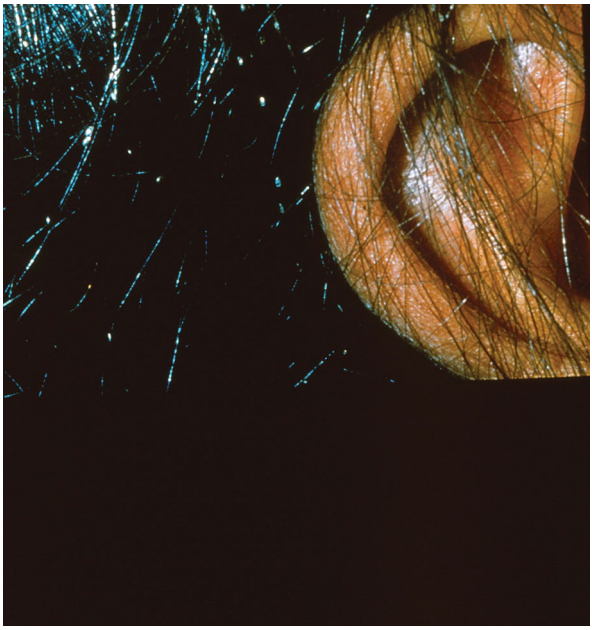

Fig. 7.1 Nits noted involving scalp hairs

the nape of the neck and behind the ears. Live lice may sometimes be observed. Nits in the pubic or axillary hair and on the eyelashes are an indication of crab lice (Fig. 7.2).

Laboratory

Lice found in the hair or after combing may be identified by simple magnification or by light microscopy. Nits can be distinguished from hair casts or "pseudonits" by dermoscopy or light microscopy.

Treatment

Over-the-counter treatments for head lice include pyrethrin shampoo, 1% permethrin lotion, ivermectin 0.5% lotion, isopropyl myristate/cyclomethicone, and dimethicone lotion. Topical medications available by prescription include spinosad 0.9% suspension and malathion 0.5% lotion. The instructions for use must be followed carefully for maximal effect. Nit combing every 2–3 days after treatment is often recommended to prevent or identify reinfestation.

For cases of head lice recalcitrant to topical treatments, oral ivermectin 200 micrograms/kg in two single doses 7–10 days apart may be considered.

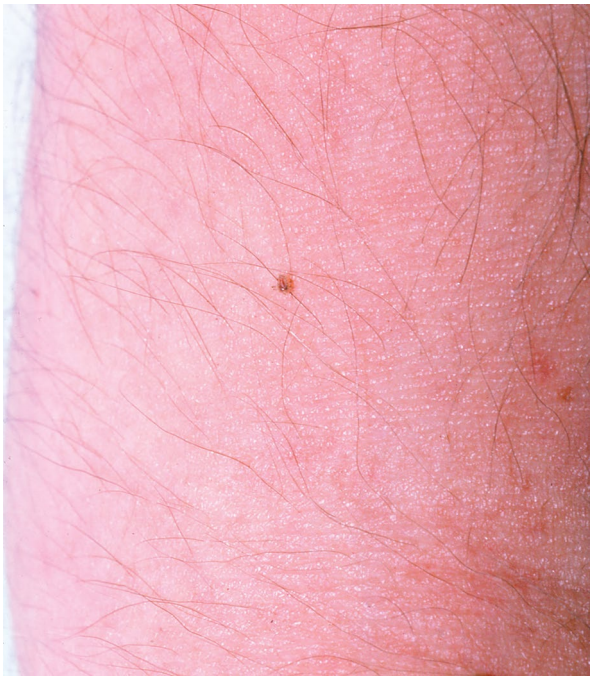

Fig. 7.2 Pubic lice (crab lice)

Pubic or crab lice are treated with 1% permethrin or 0.5% ivermectin lotion. Petrolatum may be applied to the eyelashes, if involved.

Prognosis

Failure to respond to treatment may be caused by inadequate treatment or medication-resistant lice. Reinfestation may occur due to repeated exposure.

Suggested Readings

1. Coates SJ, Thomas C, Chosidow O, et al. Ectoparasites: pediculosis and tungiasis. J Am Acad Dermatol. 2020;82:551–69.
2. Nolt D, Moore S, Yan AC, Melnick L; AAP Committee on Infectious Diseases, Committee on Practice and Ambulatory Medicine, Section on Dermatology. Pediatrics. 2022;150:e2022059282.

Chapter 8
Scabies

Julie Prendiville

Introduction

Scabies is an infestation of the skin by the *Sarcoptes scabiei* var. *hominis* mite.

Epidemiology

Scabies is transmitted by close human physical contact or by sharing of beds. It is prevalent worldwide and affects all age groups. The female scabies mite burrows into the epidermis and lays eggs. The itchy inflammatory eruption is a response to the presence of mites and their products in the skin. Clinical signs and symptoms develop approximately 4 weeks after the first infestation.

Clinical Findings

The presenting complaint is usually pruritus, particularly during the night. It is of variable severity and may be absent in crusted scabies. The pathognomonic burrows (Fig. 8.1) are found on the hands, typically in the web spaces, and on the volar wrists, feet, axillary folds, and male genitalia. A variable generalized papular or eczematous dermatitis (Fig. 8.2) occurs on the trunk and limbs. Scabies nodules and vesicles are more common in young children than adolescents. Excoriations can

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_8

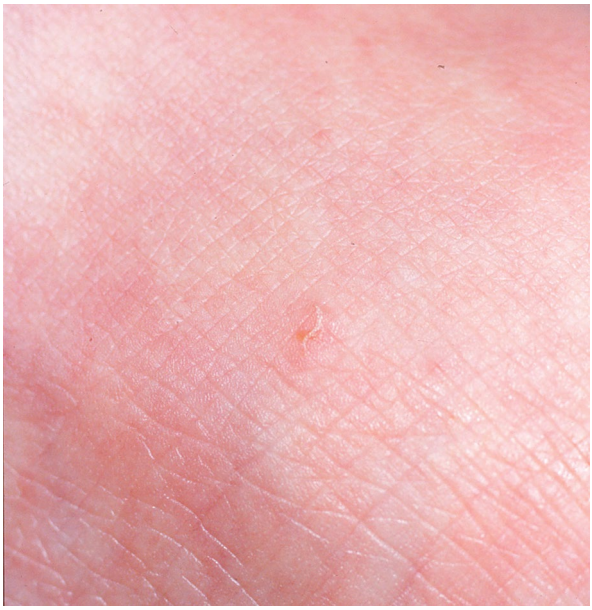

Fig. 8.1 Characteristic scabies burrow

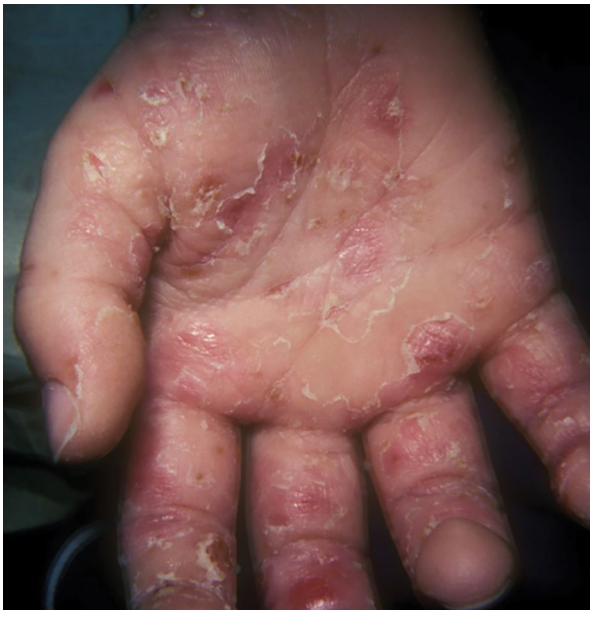

Fig. 8.2 Eczematous dermatitis of the palm in a patient with scabies

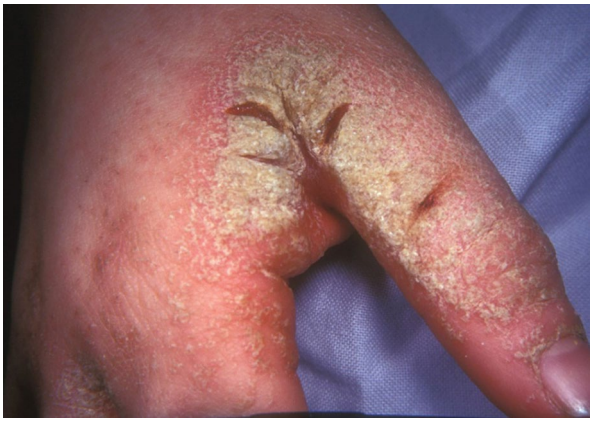

Fig. 8.3 Crusted scabies on the hand

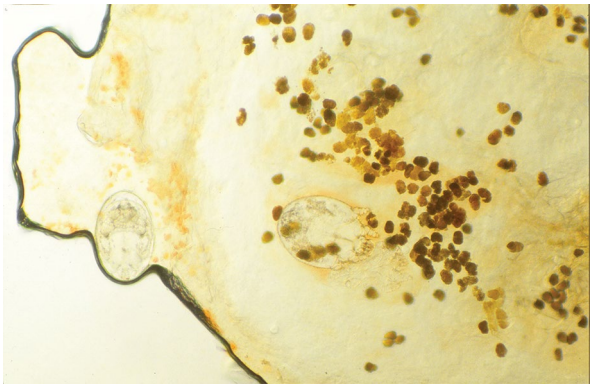

Fig. 8.4 Microscopic examination of a scraping from a patient with scabies showing mite, egg, and scybala

lead to secondary impetigo with *Staphylococcus aureus* and Group A Streptococcus. Crusted scabies is a rare and highly contagious variant (Fig. 8.3).

Laboratory

A diagnosis of scabies is confirmed by microscopic identification of the mite, eggs, or scybala (Fig. 8.4). The mite may be visualized within a burrow by dermoscopy.

Treatment

Standard treatment is the application of a topical scabicide such as permethrin 5% cream or lotion for 10–12 hours, repeated in one week. The scabicide must cover the entire body, including the neck and behind the ears. All household members and

close contacts should be treated concurrently, whether symptomatic or not. Antibiotics may be required for secondary infection.

Bed linen and clothing worn recently should be laundered. Clothing that cannot be washed may be dry cleaned or stored in a sealed plastic bag for at least 3 days. Vacuuming is advised for mattresses, carpets, and soft furnishings.

Oral ivermectin, 200 micrograms/kg in a single dose repeated in 9–10 days, is indicated for institutional outbreaks and for teenagers who are unable to apply or are noncompliant with topical therapy.

Crusted scabies requires a more complex treatment protocol with oral ivermectin in addition to topical therapy.

Prognosis

The prognosis is good if affected individuals and all close contacts and household members are treated appropriately and concurrently. Skin inflammation may persist for 1–4 weeks and require treatment with a topical steroid. Scabies nodules may persist for several months.

Suggested Reading

1. Thomas C, Coates SJ, Engelman D, et al. Ectoparasites: scabies. J Am Acad Dermatol. 2020;82:533–48.

Chapter 9
Rocky Mountain Spotted Fever

Patricia Treadwell

Introduction

Rocky Mountain Spotted Fever (RMSF) is a tick-borne illness caused by *Rickettsia rickettsii* (an intracellular gram-negative coccobacillus bacterium). The incubation following the tick bite is 2–14 days.

Epidemiology

RMSF (despite the name) has been noted in almost every contiguous state of the United States. Most of the cases occur between April and October. This coincides with peaks of tick activity and time spent outdoors. The species of tick responsible varies by region [1, 2]. Patients or families will often report tick exposure or bites. Adolescents are affected; however, it is most common in adults who work outside.

Clinical Findings

Cutaneous findings are common in RMSF. Approximately 90% of affected patients will have an exanthem. The exanthem initially begins as erythematous macules and papules; however, later the lesions become petechial or purpuric. The lesions are initially located on the wrists, palms, ankles, and soles and later spread centrally (Fig. 9.1).

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_9

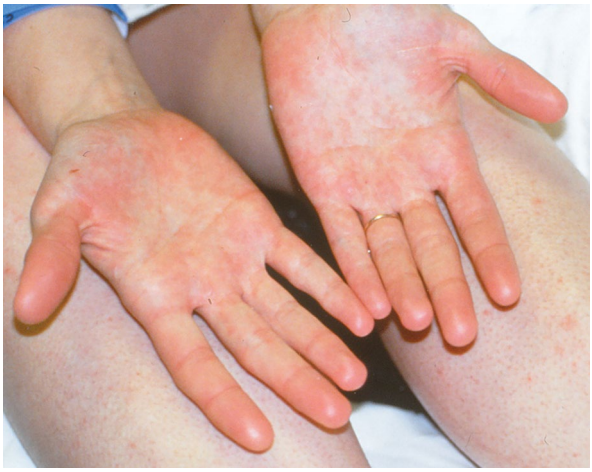

Fig. 9.1 Erythematous lesions of the palm in RMSF

Other findings include: fever, severe headache, confusion, nausea and vomiting, and photophobia.

Laboratory

The gold standard test for diagnosis is the indirect immunofluorescence antibody (IFA) test. IgG and IgM antibodies may increase in 7–10 days after symptoms begin.

Treatment

Supportive therapy may be needed if the vasculitis is widespread. Doxycycline is the treatment of choice for any age group. The dose is 2.2 mg/kg/dose given bid for 5–7 days. The doxycycline can be given empirically since delayed treatment is associated with increased mortality.

Prognosis

The mortality rate for RMSF is noted to be 5–10% of patients. Severe RMSF is associated with sequelae—particularly neurologic sequelae, e.g., hearing loss, blindness, movement disorders, speech disorders, etc.

References

1. Redbook. 2024–2027 33rd edition: Report of the Committee on Infectious Diseases. Rocky Mountain spotted fever. Itasca: American Academy of Pediatrics; 2024. p. 727–30.
2. Jay R, et al. Clinical characteristics of Rocky Mountain spotted fever in the United States: a literature review. J Vector Borne Dis. 2020;57:114–20.

Chapter 10
Cutaneous Larva Migrans

Michael Lee Smith

Introduction

Cutaneous larva migrans, also known as creeping eruption, is a common parasitic infestation of the skin caused by percutaneous penetration and migration by dog and cat hookworms. The most common organisms are *Ancylostoma braziliense* and *Ancylostoma caninum*, but other nematodes may cause this condition as well. These organisms are not capable of completing their life cycles in human hosts, so they migrate through the skin and cause an inflammatory response that is usually quite pruritic.

Epidemiology

Although ubiquitous in distribution worldwide, warmer tropical and subtropical climates are the most common areas of infestation. Warm, moist, sandy soil contaminated with eggs and larvae of the animal hookworms are the prime locations for infestation. Direct skin contact with the soil allows the larvae to enter the skin.

Clinical Findings

The incubation period is typically 1–5 days. Tracks often appear first on the feet, since that is the most common point of contact with the contaminated soil. The typical features include serpiginous erythematous tracks representing the migration

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_10

route of the larval hookworm. The tracks may be macular or raised and palpable (Fig. 10.1a, b). Vesiculobullous lesions may be seen in up to 15% of cases. There is usually intense pruritus, so secondary excoriations are common. The distribution of lesions is dependent upon the areas of skin in direct contact with the contaminated soil. However, the larvae may survive for several days and migrate a significant distance from the point of initial contact.

Laboratory

Laboratory testing is not necessary. This is only cutaneous, so peripheral blood eosinophilia is unlikely. The organism does not reach the gastrointestinal tract, so testing for ova and parasites will be negative.

Treatment

Oral ivermectin or oral albendazole are the treatments of choice. Topical thiabendazole, ivermectin, and albendazole have been reported to be efficacious as well.

Prognosis

The prognosis for cutaneous larva migrans is generally excellent, since most cases resolve spontaneously in a few weeks. Treatment may speed the resolution as well, with symptoms usually resolved within 7–10 days of treatment.

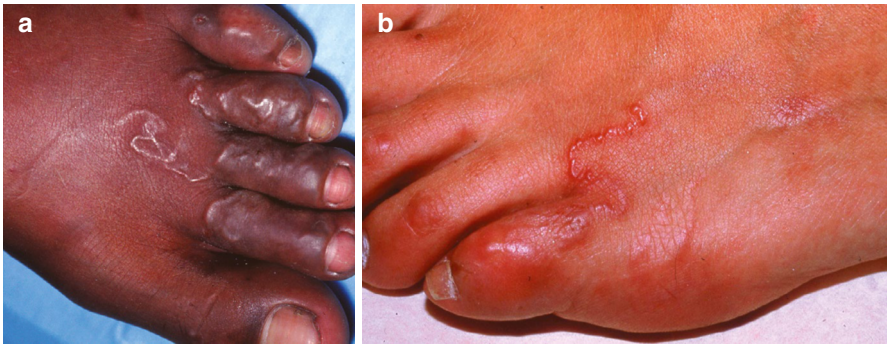

Fig. 10.1 (**a**, **b**) Serpiginous tracks on the dorsal feet in cutaneous larva migrans

Suggested Readings

1. Eldin C, Gautret P. Multiple itchy lesions after recent travel. BMJ. 2021;372:n231.
2. Shareef S, Ashack K. Cutaneous larva migrans: the unexpected souvenir. Int J Dermatol. 2023;62:e460–1.

Part III
Eczematous and Papulosquamous

Chapter 11
Atopic Dermatitis

Julie Prendiville

Introduction

Atopic dermatitis (AD) or eczema is a common inflammatory skin condition of variable severity. It is characterized by pruritus and a chronic relapsing course. AD is associated with a familial predisposition to other atopic disorders, such as asthma, food allergy, and allergic rhinoconjunctivitis. Moderate to severe AD adversely affects the health, quality of life, and emotional well-being of adolescents.

Epidemiology

Childhood AD may persist or reemerge in adolescence. The prevalence of AD in the 13–17 year age group in the United States is estimated to range from 7% to 8.6%, with up to one third suffering from moderate to severe disease. It affects all ethnic groups.

Clinical Findings

Atopic dermatitis may be categorized as *acute*, with weeping, inflamed eczema (Fig 11.1); *chronic*, with lichenification, excoriation, and pigmentary change (Fig. 11.2); or *subacute*, an intermediate appearance. The skin is often dry and may show follicular prominence or, in some cases, features of ichthyosis vulgaris. Mild

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_11

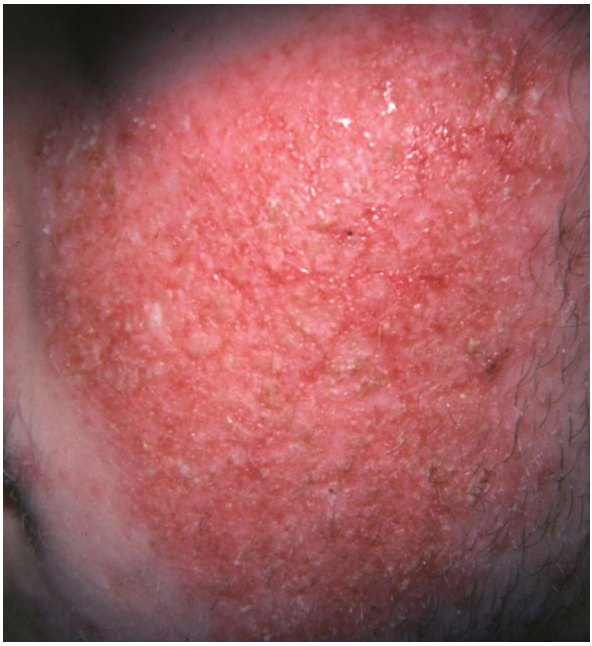

Fig. 11.1 Inflamed eczema with weeping and crusting

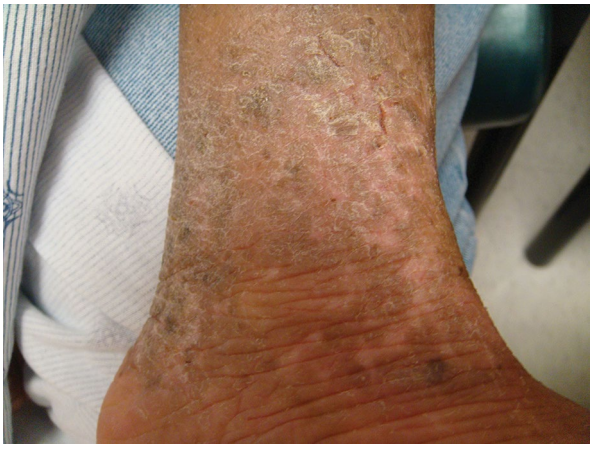

Fig. 11.2 Chronic atopic dermatitis with lichenification and dyspigmentation

eczema in adolescents is usually localized, with a predilection for the limb flexures and neck (Fig. 11.3). Moderate to severe AD is more extensive with acute, subacute, and/or chronic eczematous change, and sometimes a nodular morphology called prurigo. Pruritus is the predominant symptom. Pain or severe discomfort occurs in the presence of secondary infection. Organisms causing infection in AD are *Staphylococcus aureus* (either methicillin-sensitive MSSA or methicillin-resistant MRSA), *Streptococcus pyogenes*, and the herpes simplex virus (Fig. 11.4). Viral warts and molluscum papules may also be more prevalent.

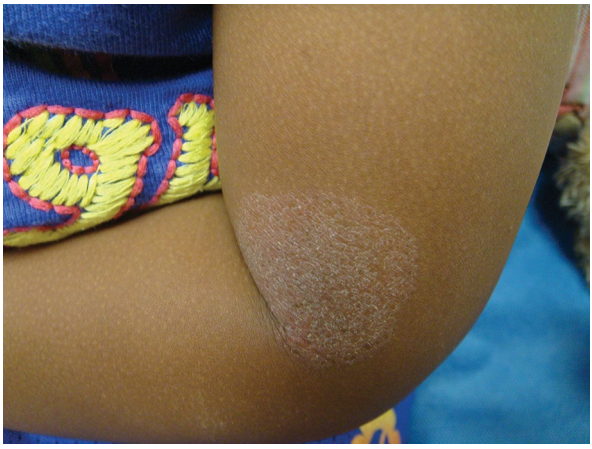

Fig. 11.3 Circular eczema with mild erythema and scale

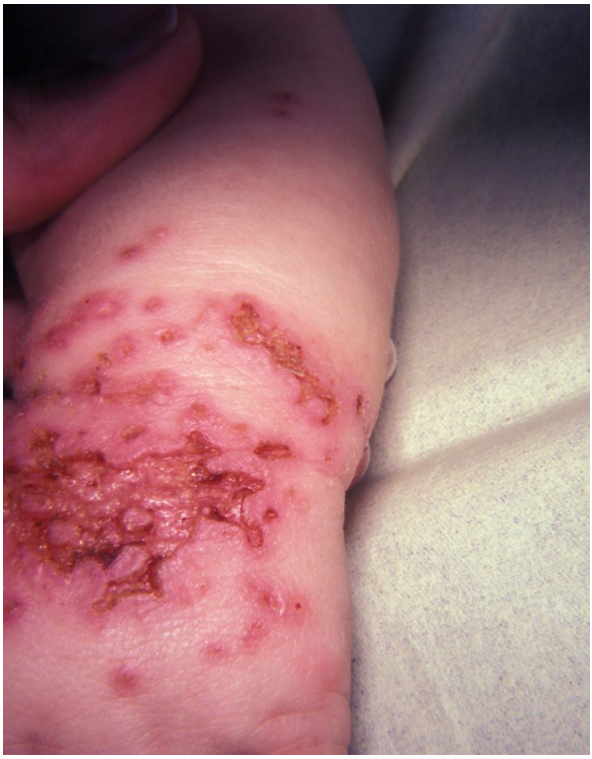

Fig. 11.4 Eczema herpeticum: Infection of eczema with herpes simplex

Laboratory

Atopic dermatitis is a clinical diagnosis based on the history and clinical findings. Laboratory investigation may be indicated to rule out other conditions (e.g., allergic

contact dermatitis, scabies, tinea corporis) or to investigate comorbidities. Skin cultures and viral studies are needed to identify secondary infections.

Treatment

Basic treatment of AD consists of avoidance of irritants, hydration of the skin by bathing and application of emollients, and judicious use of topical steroids. Topical calcineurin inhibitors may also be helpful. Secondary infection requires treatment with an oral antibiotic or antiviral medication.

Whereas treatment of mild AD is usually straightforward, moderate or severe AD in adolescents can be challenging to manage. Teenagers are particularly susceptible to developing striae from the application of topical steroids. Adherence to a topical treatment regimen may be difficult, and discouragement and emotional distress are common. Psychosocial support is important.

Advanced therapies include phototherapy and systemic anti-inflammatory and immunomodulatory drugs. Systemic steroids are only rarely justified for short-term treatment of an acute generalized AD flare. Medications such as low-dose methotrexate, cyclosporine, and azathioprine have been prescribed off-label for severe disease with modest benefit. Dupilumab, a human monoclonal antibody targeting the IL-4 receptor alpha subunit, significantly improves AD signs and symptoms and quality of life and is now widely used for severe AD in adolescents. Oral Janus kinase (JAK) inhibitors are also very effective and can be considered as a second-line treatment in some cases.

Prognosis

The prognosis is very variable. Many patients show improvement with time. Others continue to suffer from eczema into adult life.

Suggested Readings

1. Choo ZY, et al. Update in atopic dermatitis for the primary care physician: a review of advances in the understanding and treatment of atopic dermatitis. Dis Mon. 2024;70(4):101687.
2. Yim HJ, et al. Recent advances in immunomodulators for atopic dermatitis. Curr Opin Pediatr. 2023;35:671–679S.

Chapter 12
Allergic Contact Dermatitis

Michael Lee Smith

Introduction

Allergic contact dermatitis (ACD) is a type IV hypersensitivity reaction to any of a variety of substances after repeated skin contact. It has two phases: a sensitization phase during which the allergen(s) have sufficient and generally recurrent contact with skin, followed by an elicitation phase in which subsequent allergen contact induces an inflammatory response.

The most common allergic contact dermatitis seen in adolescents is due to plants of the *Toxicodendron* genus, including poison ivy, oak, and sumac. This is also known as rhus dermatitis. Numerous other allergens are problematic for teens as well. The most common non-plant allergen causing ACD in teens is nickel, followed by neomycin, fragrances, preservatives (formaldehyde, methylisothiazolinone, methylchloroisothiazolinone), and lanolin [1–3].

Exposure history is often the key to the diagnosis of ACD. Outdoor plant exposure history is helpful. Among teens, other exposures should be queried, including hair dyes, perfumes, nail polish, henna, sports equipment, and work history.

Epidemiology

Prevalence estimates of ACD are over 15% and increasing. Nickel allergy is the most common ACD worldwide with prevalence estimates 5–10%. In one study referenced below, among patients with ACD, over 75% are adolescents or adults [2].

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_12

There is a female predominance of almost 3:1, especially with nickel ACD (Ni-ACD).

Clinical Findings

Once sensitized, ACD appears soon after exposure to the offending allergen, often starting with patchy erythema. Papules and/or vesiculobullous lesions follow, often accompanied by underlying edema. Vesiculobullous lesions may develop serous crust. Chronic ACD often consists of lichenified plaques with excoriations and hyperpigmentation. Intense pruritus is a consistent feature.

Rhus dermatitis lesions are most often noted in exposed areas. The patients will often present with a history of exposure and are noted to have erythematous (sometimes linear or patterned) papulovesicular lesions (Fig. 12.1). Itching is typical.

Nickel ACD will be noted in specific areas of exposure to nickel. The lesions are plaque-like and may develop crusting. Common areas are earlobes (Fig. 12.2), posterior neck, and the lower abdomen (Fig. 12.3). Other sites of exposure may be noted as well (Fig. 12.4a, b). Widespread "id" reaction may be noted (Fig. 12.5).

Chronic hand dermatitis is a frequent presentation of ACD in adolescents. It may have features of dyshidrotic eczema with microvesicles (Fig. 12.6), desquamation, or chronic erythema and scale. Similarly, isolated hand and foot eczema in older children and adolescents may require evaluation for ACD [1].

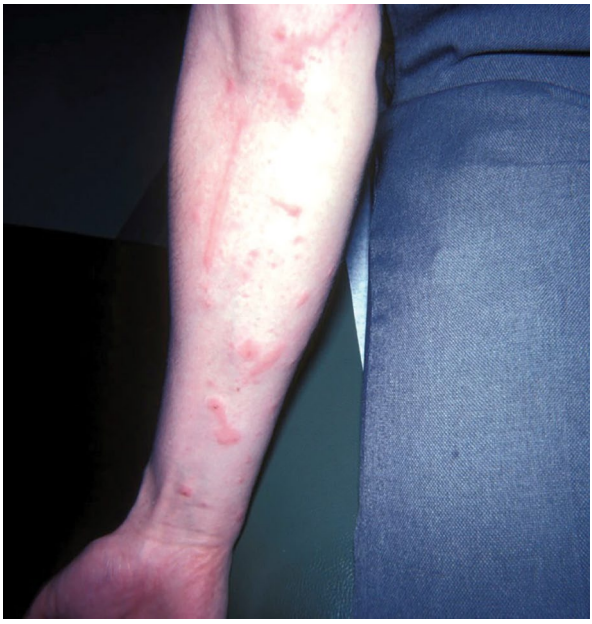

Fig. 12.1 *Rhus* dermatitis with linear lesions

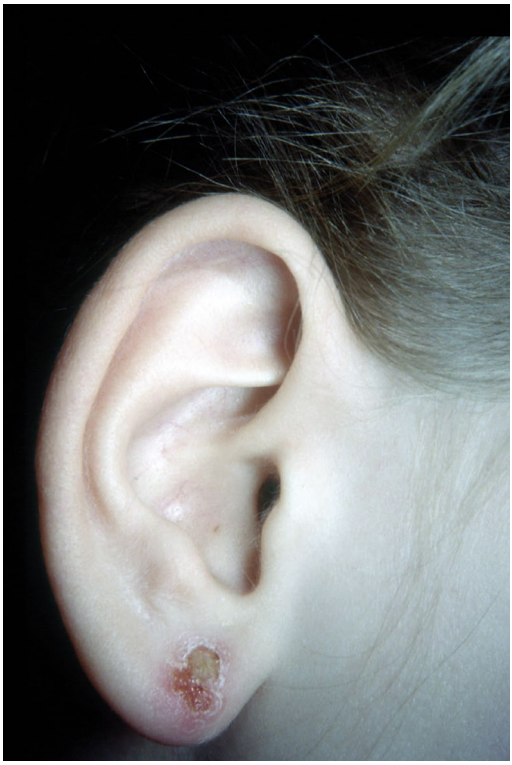

Fig. 12.2 Earlobe dermatitis from nickel exposure associated with piercing

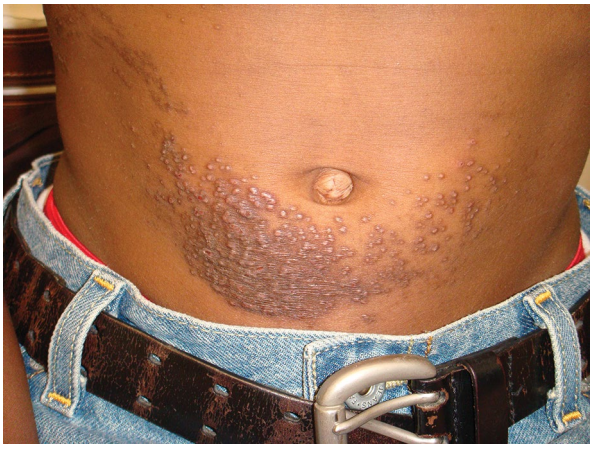

Fig. 12.3 Ni –ACD of lower abdomen from belt buckle

Laboratory

Skin biopsy is not usually necessary, but if obtained, it should show spongiotic dermatitis with numerous eosinophils. Allergens other than plants can be assessed by

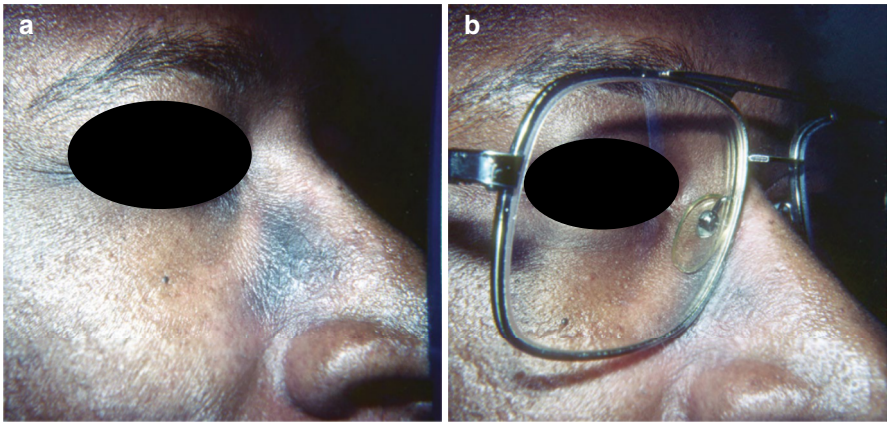

Fig. 12.4 (**a** & **b**) Ni-ACD from glasses

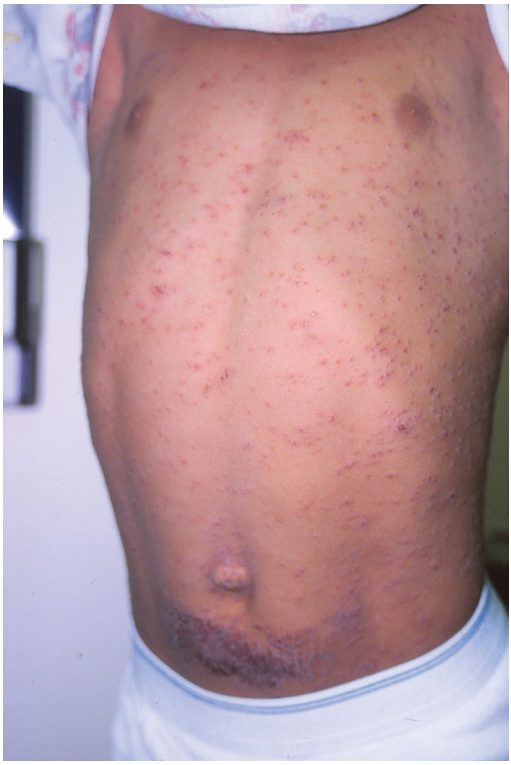

Fig. 12.5 Id reaction associated with Ni-ACD on abdomen

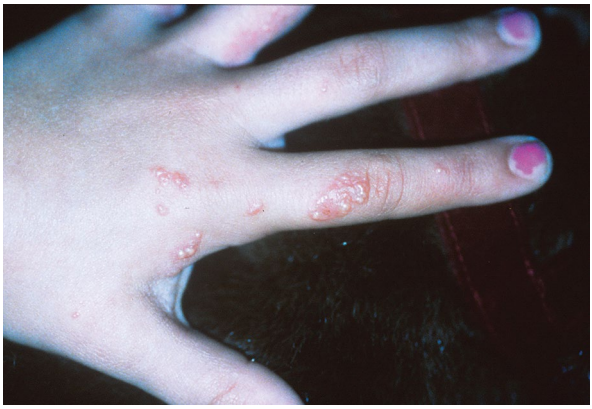

Fig. 12.6 Microvesicles in a teen with ACD

patch testing [2, 3]. The presence of released nickel from metal accessories can be assessed with a dimethylglyoxime test.

Treatment

Treatment consists of avoiding offending allergens. Rhus dermatitis can be minimized by using clothing to cover the skin and cleansing skin as soon as possible after exposure has occurred.

Topical or systemic corticosteroids are useful for calming inflammation. Topical calcineurin inhibitors can help reduce the inflammation and may delay or prevent the elicitation phase.

Prognosis

The sensitization tends to be permanent, so patients should attempt allergen avoidance.

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Chapter 13
Seborrheic Dermatitis

Patricia Treadwell

Introduction

Seborrheic dermatitis is an irritant dermatitis that affects approximately 3% of the population. The sebaceous gland secretions are altered by *Malassezia* (previously *Pityrosporum*) species (part of the normal flora), and an irritant dermatitis develops.

Epidemiology

This chapter will focus on the adolescent and adult populations (not the infantile type). The dermatitis tends to be more widespread in the setting of immunosuppression (e.g., HIV/AIDS and hepatitis C) and underlying neurologic diseases (e.g., Parkinson's disease). Reference [1, 2]

Clinical Findings

Seborrheic dermatitis tends to be noted in areas with more concentrated sebaceous glands including the paranasal and glabellar areas, the eyebrows, and the scalp. Seborrheic dermatitis is characterized by lesions with erythema and yellowish scale – which has been described as "potato chip" scale on an oily background (Fig. 13.1). Itching may be present. Scalp involvement in seborrheic dermatitis consists of diffuse whitish scale (Fig. 13.2).

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_13

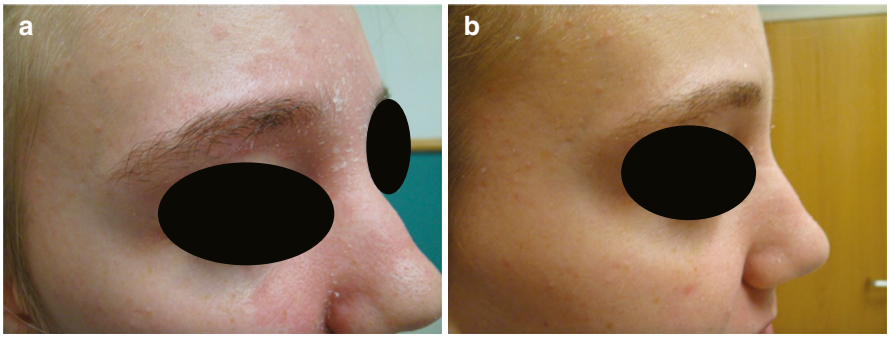

Fig. 13.1 (**a**) Seborrheic dermatitis in an adolescent with erythema and scale. (**b**) Seborrheic dermatitis shows significant improvement in the patient in 1A with ketoconazole cream for 2 weeks

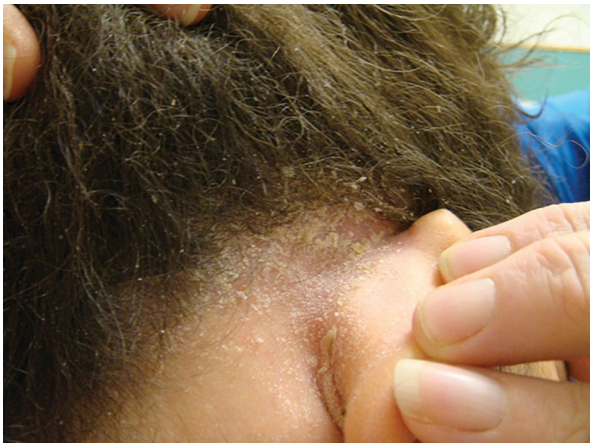

Fig. 13.2 Whitish scale of the retroauricular area and scalp from seborrheic dermatitis

Laboratory

The diagnosis is most often made based on clinical findings.

Treatment

Topical antifungals such as azole preparations (e.g., ketoconazole) and shampoos have been noted to be effective in seborrheic dermatitis. Figure 13.1b Topical corticosteroids or topical calcineurin inhibitors can be a useful adjunct when inflammation is moderate to severe. Phosphodiesterase (PDE)-4 inhibitors have been used off-label. Reference [3] Systemic antifungal therapy is not FDA-approved and may have adverse side effects.

Although selenium sulfide and zinc pyrithione shampoos have been prescribed for many years, limited evidence exists for their efficacy.

Prognosis

Seborrheic dermatitis tends to be chronic but does not have significant long-term effects.

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Chapter 14
Pityriasis Rosea

Patricia Treadwell

Introduction

Pityriasis rosea (PR) is a benign papulosquamous disorder which can be seen in adolescents.

Epidemiology

The most common age group affected is 10–35-old individuals.

There has been speculation that PR is associated with a viral illness. There is a seasonal variation in the number of cases, and Human Herpes Virus (HHV) 6 and 7 have been specifically implicated.

Clinical Findings

The initial lesion in 88% of patients is termed a "herald patch." The "herald patch" is most often an erythematous somewhat raised lesion with overlying scale. The "herald patch" typically measures 2–5 cm in diameter and can occur anywhere on the body. After 10–14 days, multiple smaller (5–10 mm), oval-shaped lesions are noted primarily on the trunk and proximal extremities oriented parallel to Langer lines (Fig. 14.1). In skin of color, the lesions may be more papular and/or crusted. The lesions are usually present for 6–8 weeks.

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https://doi.org/10.1007/978-3-031-86177-2_14

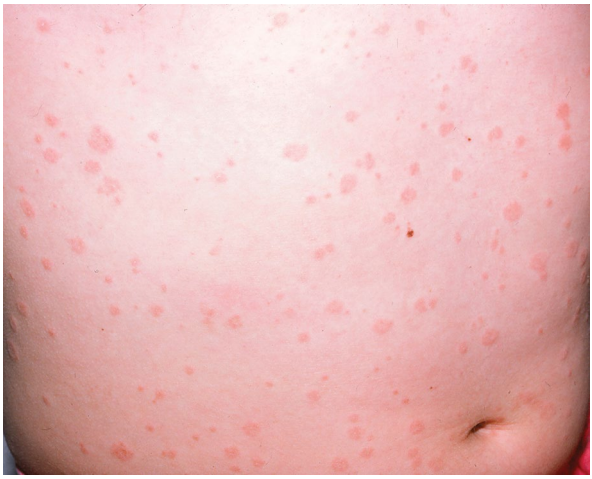

Fig. 14.1 Pityriasis rosea lesions with erythema and scale, the lesions (especially on the flank) follow Langer lines

Laboratory

No laboratory testing is diagnostic. Serologic tests for syphilis are recommended if suspected by medical history and/or if lesions are noted on the palms and soles.

Treatment

Since the disorder resolves on its own, treatment is often not necessary. Topical or systemic antipruritics may be prescribed if the itching is significant. A Cochrane review of interventions for PR found only acyclovir as resulting in some improvement. Reference [1]

Prognosis

As mentioned above, the disorder resolves spontaneously. In skin of color, post-inflammatory dyspigmentation can be seen.

References

1. Contreras-Ruiz J, et al. Interventions for pityriasis rosea. Cochrane Database Sys Rev. 2019;2019(10):CD005068.

Part IV
Autoimmune and Rheumatologic

Chapter 15
Systemic Lupus Erythematosus

Patricia Treadwell

Introduction

Systemic lupus erythematosus (SLE) is a chronic inflammatory condition that can affect various organ systems. The version of this disorder that occurs in childhood is labeled as childhood-onset systemic lupus erythematous (cSLE).

Epidemiology

Childhood-onset SLE (cSLE) has its peak incidence in early adolescence. The average age of presentation of cSLE is 12–14 years. The M:F ratio is 1:5 prior to puberty. Following puberty, the ratio is 1:9. Although no registry exists, estimates of the incidence are typically quoted as 3.3–8.8 per 100,000 individuals. It has been noted that cSLE affects children of Black, Hispanic, and Asian ancestry more often, and they have a worse prognosis.

Clinical Findings

The most common cutaneous findings are photosensitivity, malar erythema (Fig. 15.1), discoid LE lesions (Fig. 15.2), nailfold telangiectasias Fig. 15.3 and oral ulcers. Subacute cutaneous lupus (SCLE) is characterized by scaly erythematous lesions in sun-exposed areas such as the back. Figure 15.4a, b Other findings include

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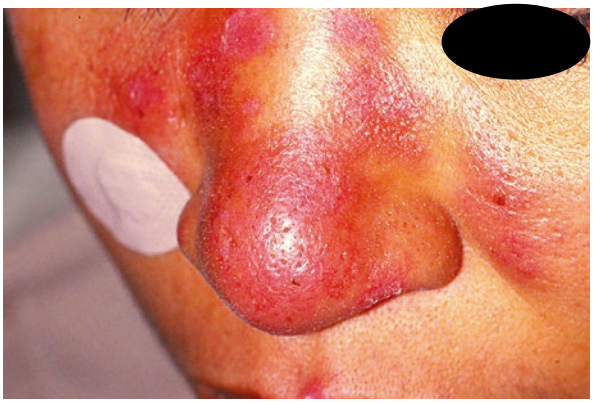

Fig. 15.1 SLE-Erythema of the malar areas and nose

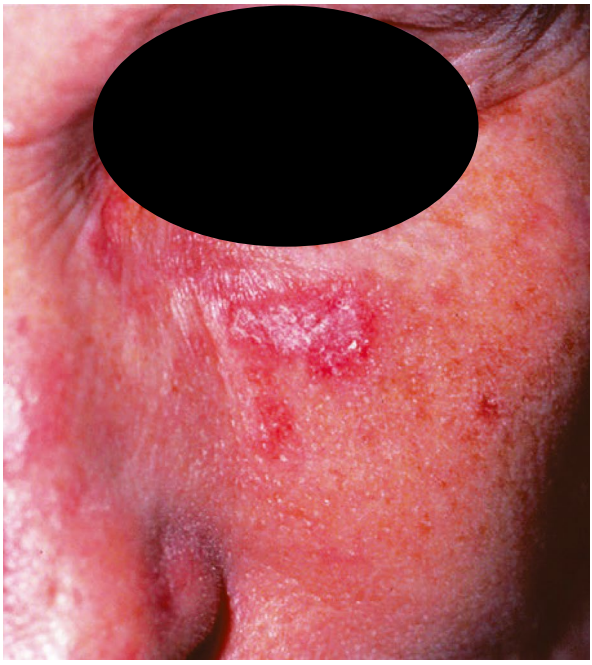

Fig. 15.2 DLE Somewhat annular lesion with scale, hypopigmentation, and atrophy

alopecia, livedo reticularis, Raynaud phenomenon, and panniculitis. Extracutaneous features include fever, arthralgias, arthritis, fatigue, and CNS involvement.

Laboratory

In 99% of children with cSLE, the antinuclear antibody (ANA) is positive. If the titer is greater than 1:1280 or higher, cSLE is a likely diagnosis.

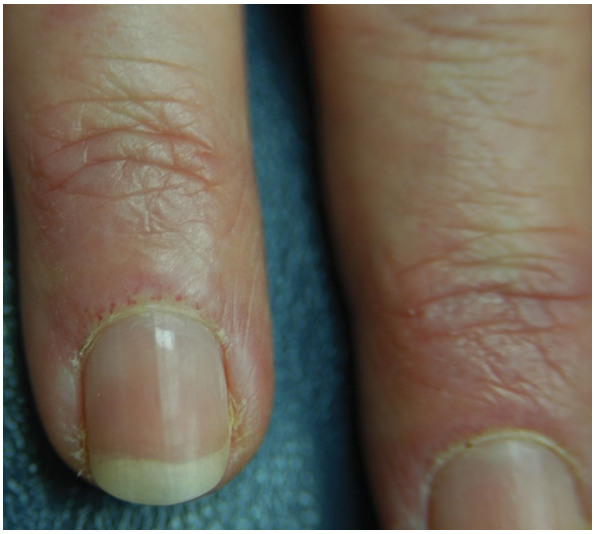

Fig. 15.3 cuticles show dilated capillaries

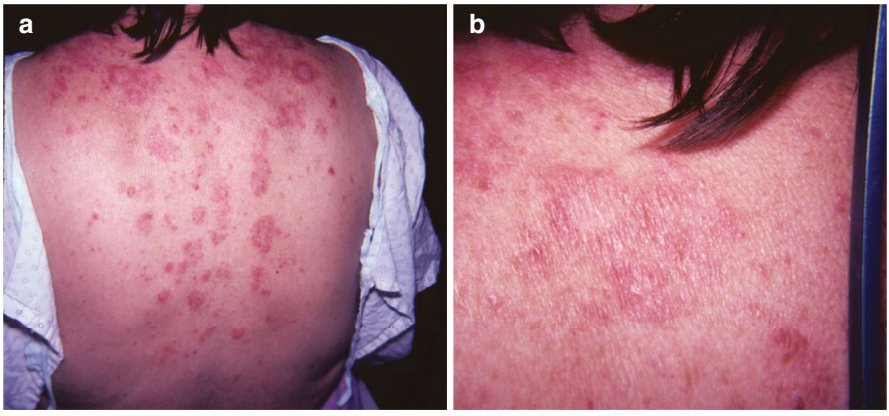

Fig. 15.4 (**a**) SCLE Erythematous scaly patches of the back. (**b**) Scaly patch in close up

Anti-dsDNA antibodies can be used to monitor disease activity. The anti-dsDNA and cardiolipin antibodies are more frequent in cSLE as compared to aSLE (adult-onset SLE); however, a positive rheumatoid factor is less common.

Urinalysis is an important tool to evaluate any renal involvement that may develop.

Treatment

Treatment options for cSLE include oral hydroxychloroquine, glucocorticosteroids, and other immunosuppressants, including biologics. The cutaneous lesions may be treated with topical or intralesional steroids or with topical calcineurin inhibitors in addition to sun protection.

Prognosis

Adolescents with cSLE have a good prognosis when there is no renal involvement. Those adolescents with renal involvement require more aggressive therapies and close follow-up.

Suggested Readings

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3. Trindade VC, et al. An update on management of childhood-onset systemic lupus erythematosus. Pediatr Drugs. 2021;23:331–47.

Chapter 16
Juvenile Dermatomyositis

Patricia Treadwell

Introduction

Juvenile Dermatomyositis (JDMS) is a rare idiopathic inflammatory disorder affecting skin and muscle.

Epidemiology

JDMS can begin in adolescence; however, peak incidence is 5–15 years of age, occurring 3–4 cases per million children each year. It is rare, however, JDMS is the most common myopathy of childhood [1]. The male-to-female ratio is 1:2.

Clinical Findings

Characteristic violaceous discoloration of the eyelids (heliotrope) (Fig. 16.1), erythematous scaly papules of the dorsal hands overlying the MCP and PIP joints (Gottron's papules) (Fig. 16.2) and other parts of the body (Fig. 16.3). In some cases, calcinosis may develop. Capillary dilatation is noted in the periungual areas with some capillary dropout. Symmetrical proximal muscle weakness is also noted. Parents may report photosensitivity and/or worsening of the cutaneous findings with sun exposure.

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_16

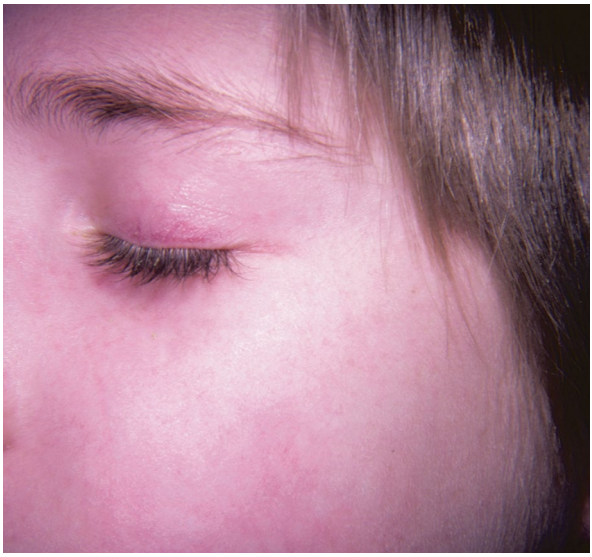

Fig. 16.1 Heliotrope-violaceous discoloration of the upper eyelid

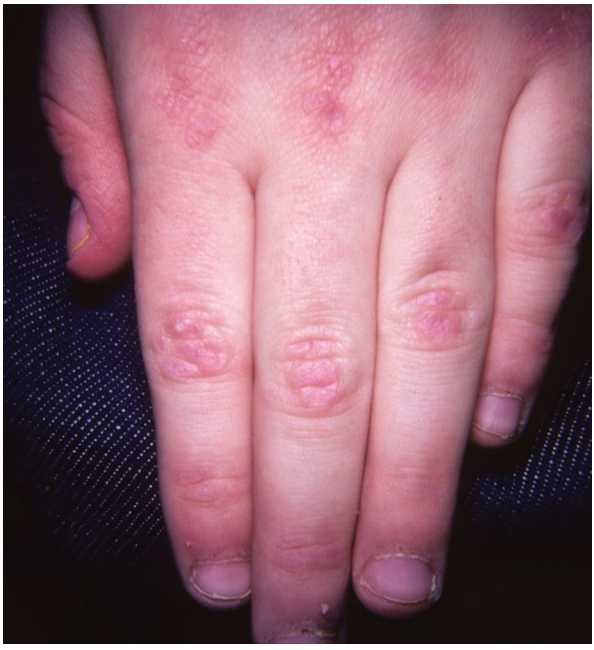

Fig. 16.2 Gottron's papules—noted on the dorsal hands overlying the joints

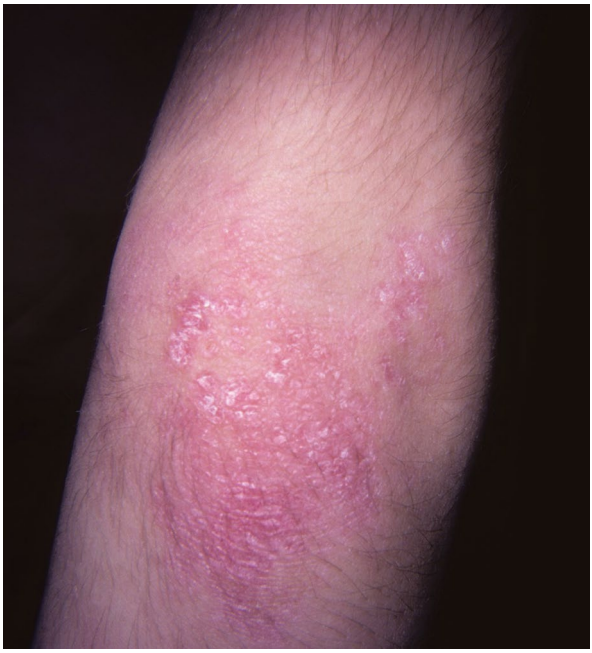

Fig. 16.3 JDMS patch with calcinosis of the elbow

Other non-cutaneous features may include pulmonary, cardiac, and gastrointestinal findings.

Laboratory

Laboratory evaluation shows elevated muscle enzymes (serum aldolase, AST, CPK, and LDH). Skin biopsies are nonspecific with perivascular inflammation.

MRI has been used for diagnosis and for monitoring disease activity. The T2, fc-T2 (fat-corrected T2) and FF (fat fraction) measurements are helpful in JDMS to distinguish those MRI findings that may also be seen in other myopathies. EMG and muscle biopsy are less often used because of the invasiveness. Muscle biopsies, when they are performed, can be guided by MRI since the muscle involvement can be unevenly distributed.

Myositis-Specific Antibodies (MSAs), when they are available, can be helpful in facilitating diagnosis, help predict prognosis, and guide management.

Distinguishing features of some of the MSA's (Ref. [2]):

Mi-2	4–10% of JDMS patients
	Benign prognosis
	Good response to conventional therapy
TIF1	18–32% of JDMS patients
	Younger at onset; chronic course
	Severe refractory skin disease
MDA5	7–33% of JDMS patients
	Milder muscle involvement
	Ulcerative cutaneous lesions
	Interstitial lung disease (ILD)
	Require intensive immunosuppressive therapy
NXP2	15–23% of JDMS patients
	Calcinosis cutis
	Disabling myopathy
	Lower remission rate
SAE	1% of JDMS patients
	Severe cutaneous disease
	Minimal muscle disease
SRP	1.6% of JDMS patients
	Immune-mediated necrotizing myositis
	Severe muscle disease
	Cardiac involvement

Table adapted from Ref. [2]

Myositis-Associated Antibodies (MAAs) are seen in JDMS patients who have an JDMS overlapping with other autoimmune connective tissue diseases. Reference [2]

Treatment

The medications used most often for JDMS are systemic corticosteroids, antimalarials, methotrexate, mycophenolate mofetil, and rituximab. Other immunosuppressive agents, intravenous immunoglobulin, cyclophosphamide, and calcineurin inhibitors have also been used in more refractory cases. Emerging treatments include other biologics and disease-modifying anti-rheumatic drugs (DMARDs), which have been used in some ongoing trials. Sun protection and physical therapy are also recommended.

Prognosis

The mortality rate of JDMS is less than 2%. JDMS is generally not associated with malignancy (in contrast to adult DM), and an occult malignancy work-up is not recommended.

References

1. DeWane ME, et al. Dermatomyositis: clinical features and pathogenesis. J Am Acad Dermatol. 2020;82:267–81.
2. Kwiatkowska D, et al. The significance of autoantibodies in Juvenile Dermatomyositis. Biomed Res Int. 2021:5513544. https://doi.org/10.1155/2021/5513544.

Part V
Reactions to External Causes

Chapter 17
Fixed Drug Eruption

Patricia Treadwell

Introduction

Fixed drug eruption (FDE) is a unique reaction to a variety of drugs. The drug reaction is categorized as a delayed-type hypersensitivity reaction mediated by CD8+ memory T cells [1].

Variants include FDE, non-pigmented FDE (NPFDE), and generalized bullous fixed drug eruption (GBFDE)

Epidemiology

This particular drug eruption recurs in the same site(s) in response to exposure to the offending agent. The medications most often responsible are: (1) non-steroidal anti-inflammatory drugs (NSAIDs); (2) other analgesics; (3) antibiotics [2]; (4) laxatives; (5) antiepileptics; (6) and other drugs. Foods have also been implicated.

Clinical Findings

The lesions are typically oval or round with "dusky" (brownish or violaceous) centers [3]. Figure 17.1 On occasion, a bulla may form. They can occur anywhere on the body, including mucosal surfaces. Hyperpigmentation is a common feature; however, non-pigmented FDE (NPFDE) has been reported [1].

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_17

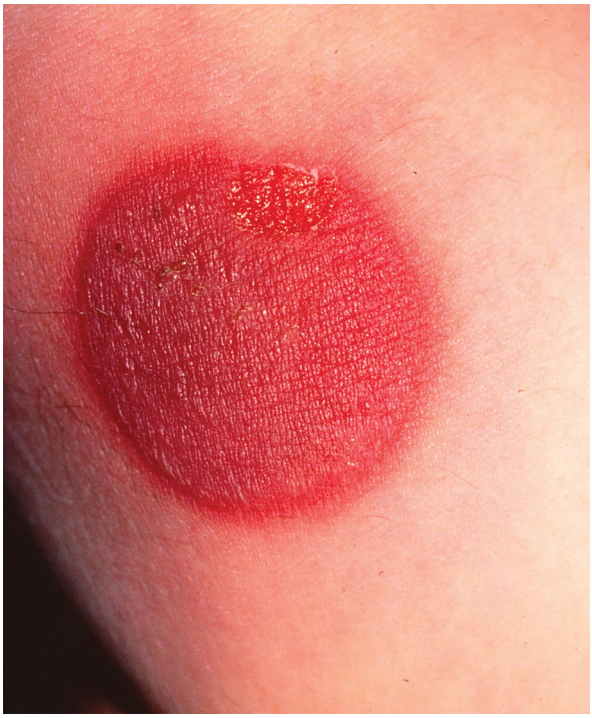

Fig. 17.1 FDE from tetracycline

Laboratory

Histopathology shows a lichenoid infiltrate with necrotic keratinocytes and spongiosis. Oral drug challenge tests (also known as oral provocation tests) are considered the gold standard. However, patch tests can also be utilized to confirm the causative agent, as can lymphocyte transformation tests.

Treatment

Avoid the offending drug. Topical and systemic corticosteroids have been effective treatments.

Prognosis

Avoidance of the drug trigger can minimize recurrences. Residual pigmentation can be noted in some cases.

References

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2. Barbier O, et al. Ecstasy-induced fixed drug eruption. Contact Derm. 2022;87:280–1.
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Chapter 18
Phytophotodermatitis

Patricia Treadwell

Introduction

Phytophotodermatitis is a dermatitis that occurs following various furocoumarins coming in contact with the skin and subsequent ultraviolet exposure.

Epidemiology

Phytophotodermatitis can occur at any age. The furocoumarins associated with the dermatitis are found in citrus fruits (especially limes and lemons), figs, wild dill and parsley, celery, fennel, and parsnip [1–3]. The hyperpigmentation may be more evident in skin of color.

Clinical Findings

In milder cases, the cutaneous findings may consist of hyperpigmentation in an unusual pattern. In more severe cases, erythema and tense bullae may develop (Fig. 18.1). The tense bullae may be painful. Itching may be present.

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https://doi.org/10.1007/978-3-031-86177-2_18

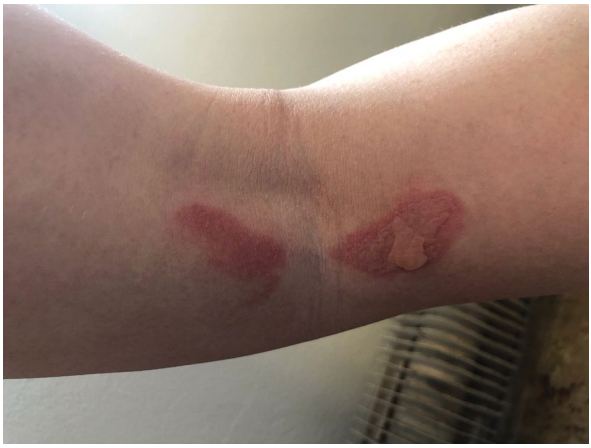

Fig. 18.1 Phytophotodermatitis with erythematous lesions, one with a bulla on the back of the leg after hiking

Laboratory

No laboratory testing is diagnostic or necessary.

Treatment

If the lesions are symptomatic, topical corticosteroids or antipruritics can be prescribed. More severe involvement may require admission to a burn unit.

Prognosis

The prognosis is generally good, since the hyperpigmentation will fade over time. Unfortunately, some cases have been misdiagnosed as child abuse based on the patterns of the lesions [4].

Avoiding the offending agents and ultraviolet exposure can minimize future occurrences.

References

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2. Hamann CR, et al. Paediatric phytophotodermatitis ‘by Proxy’ from parental transfer of lime. Contact Derm. 2022;87:98–9.
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Chapter 19
Outdoor Exposures: Arthropods and Sun

Michael Lee Smith

Introduction

Arthropod bites and stings are ubiquitous. The majority of exposures are related to outdoor activities. Such outdoor exposures occur at all ages, with no unique predilection for adolescents. Reactions may range from a minor nuisance to severe and life-threatening conditions. Photosensitivity reactions are far less common and often difficult to diagnose. Outdoor activities predispose teens to a variety of cutaneous maladies, including three specific photosensitivity disorders that are often seen in adolescents and young adults: polymorphous light eruption, actinic prurigo, and hydroa vacciniforme. Many other genetic and inflammatory conditions cause photosensitivity but are beyond the scope of this chapter.

Epidemiology

In the U.S. alone, arthropod bites and stings account for almost a million emergency department visits annually. Injuries range from simple bites to painful stings, sometimes resulting in serious envenomation, anaphylactic reactions, or as vectors of systemic disease. Polymorphous light eruption (PMLE) may appear in the second decade of life, but may be more common in the third decade. There is a female predominance. PMLE is seen more often in patients with skin of color than in fair skin. Actinic prurigo is seen almost exclusively in Native Americans or individuals of combined Indigenous and European descent. Hydroa vacciniforme does not appear to have skin color or ethnic predilection [1–3].

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Clinical Findings

Arthropod injuries may be papular (Fig. 19.1), papulovesicular (Fig. 19.2), bullous (Fig. 19.3), indurated nodules or plaques, and/or urticarial wheals. Fire ant stings are often multiple around a pivot point where the ant grasps the skin with mandibular chelicerae and then rotates the abdomen to deliver several stings (Fig. 19.4a–c). Within a day the stings become pustules on an erythematous base. Symptoms range from itching to pain from stings, to tender indurated nodules.

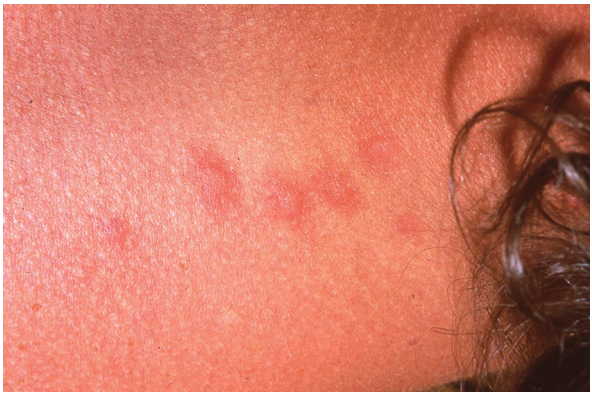

Fig. 19.1 Flea bites in a 10-year-old

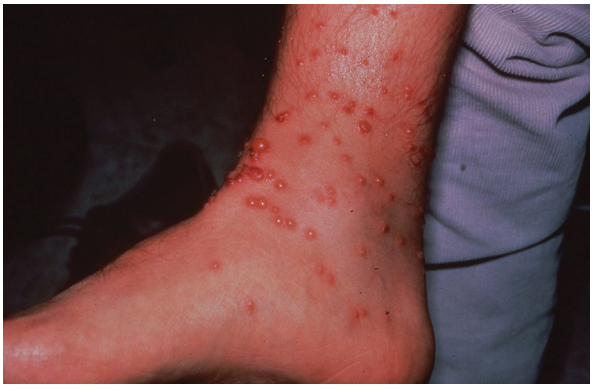

Fig. 19.2 Chigger bites in a college student

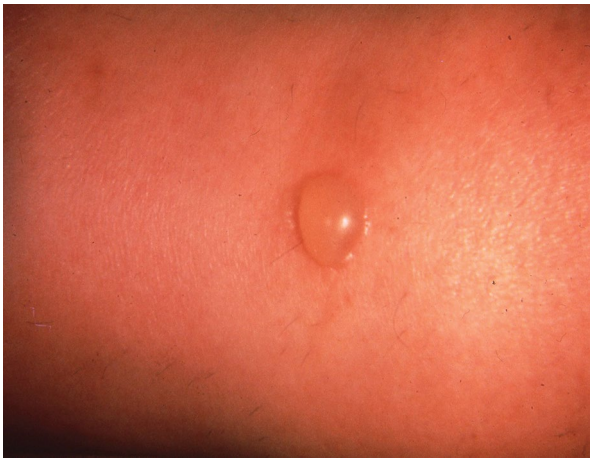

Fig. 19.3 Bullous arthropod bite in a teen

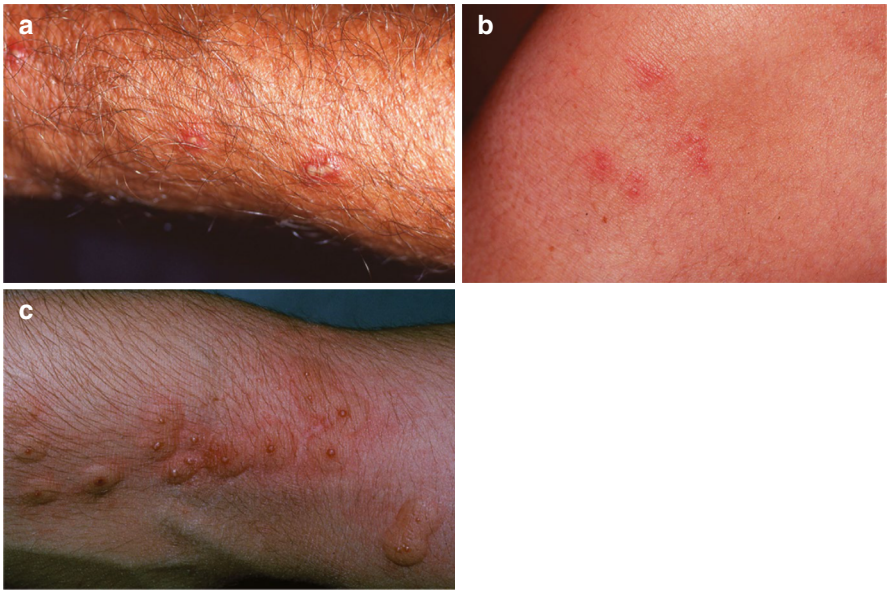

Fig. 19.4 (**a**) College student with fire ant stings showing an arcuate cluster of tiny pustules (**b**) Fire ant stings in early papular stage (**c**) Fire ant stings in a young adult showing tiny pustules on erythematous edematous wheals

PMLE is intermittent and pruritic. It may appear hours after sun exposure. It tends to be worse in the spring and early summer, with reduced symptoms in late summer due to "hardening" with ongoing sun exposure. In darker skin, it is seen commonly on the neck, with tiny erythematous papules that may coalesce into thin plaques. In fair skin, the extremities are more often involved, with clusters of papules or papulovesicles (Fig. 19.5).

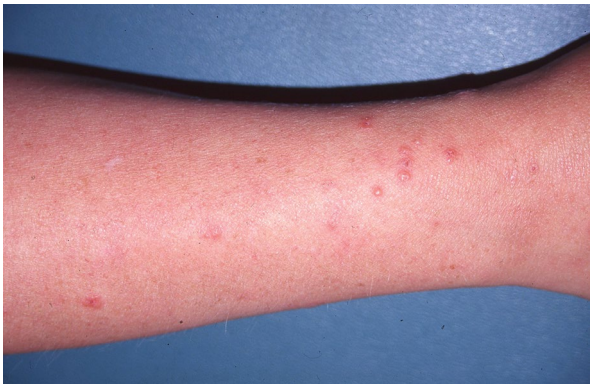

Fig. 19.5 PMLE in a young adult female with discrete erythematous papules

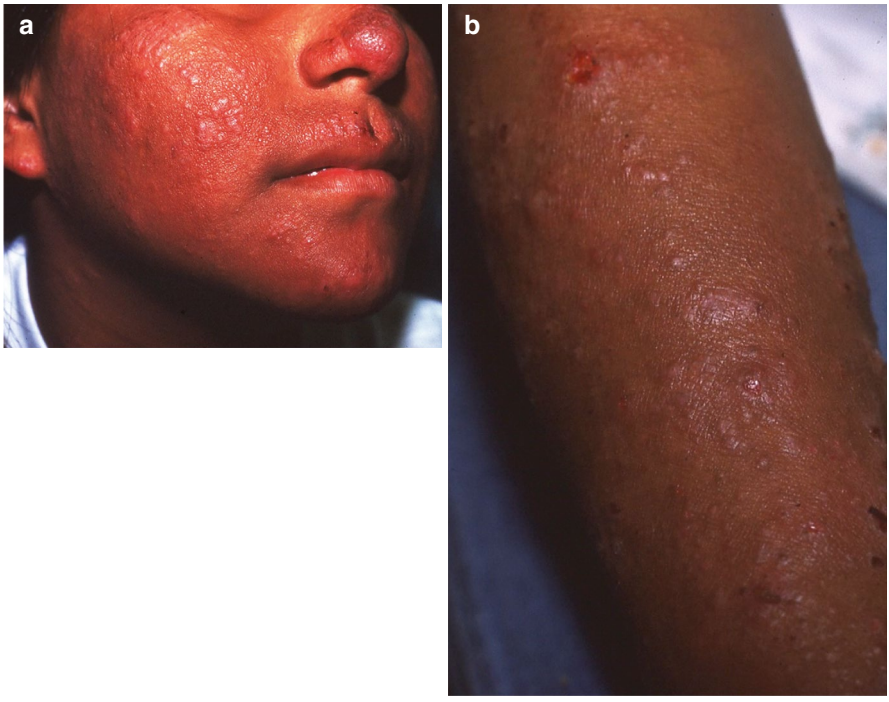

Fig. 19.6 (**a**) Actinic prurigo in a 14-year-old Arapahoe girl (**b**) Actinic prurigo in a 14-year-old Arapahoe girl. Clusters of erythematous to hyperpigmented papules and plaques, as well as nasal tip hyperpigmentation are noted

Actinic prurigo may start hours to days after exposure, with pruritus that may become intense. Papules and nodules frequently are deeply excoriated, leading to significant scarring. Face, arms and legs are most often involved (Fig. 19.6a, b).

Hydroa vacciniforme develops hours to days after sun exposure, with pruritus and stinging as the initial symptoms. Hemorrhagic papulovesicles are common, often followed by bullae and crusting. Scarring is severe (Fig. 19.7a–c).

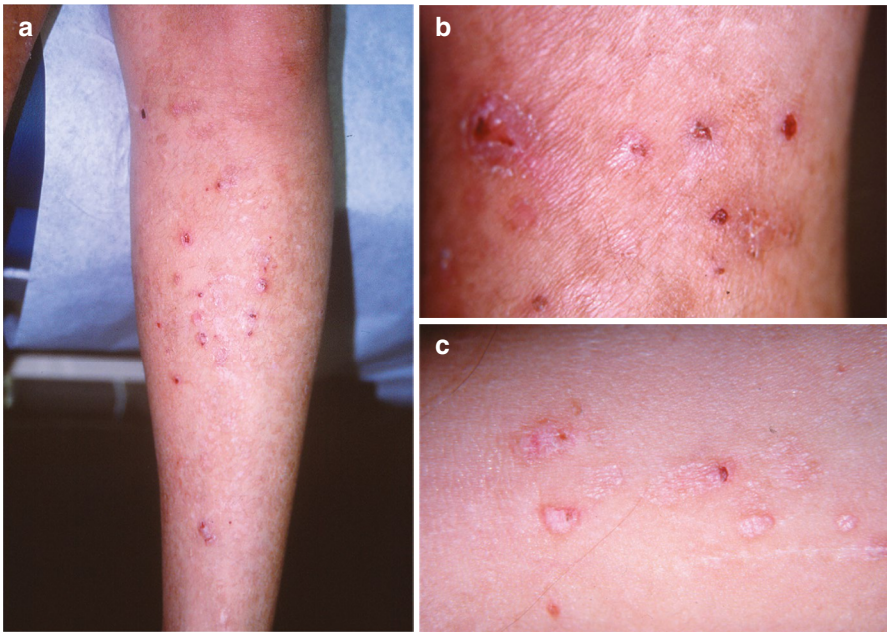

Fig. 19.7 (**a**) Hydroa vacciniforme in a 16-year-old girl showing excoriated erythematous papules. (**b**) Hydroa vacciniforme in a 16-year-old girl (**c**) Scarring in Hydroa vacciniforme

Laboratory

Laboratory testing is not often helpful. The diagnoses are made clinically, with classic presentations in typical photo distribution. Skin biopsy may help in uncertain cases.

Treatment

Arthropod bites and stings can be avoided much of the time with proper clothing, the use of age-appropriate repellents, and avoidance of locations and times of day when the arthropods are more active. Symptoms may be relieved by the use of cold packs, topical corticosteroids, menthol- or pramoxine-based anti-itch products, or even oral corticosteroids if necessary. See Herness et al. [2].

Strict photoprotection is of paramount importance in managing photosensitive disorders. Symptoms may be ameliorated with the use of topical corticosteroids, topical calcineurin inhibitors, or hydroxychloroquine. PMLE may be reduced in intensity with ultraviolet phototherapy "hardening" before spring outdoor exposure. Shah et al. [4] reported significant improvement in actinic prurigo with the use of dupilumab.

Prognosis

The prognosis for uncomplicated arthropod bites and stings is excellent. As vectors for infectious diseases, causes of severe allergic reactions, and delivery of toxins or venoms, the prognosis varies and is beyond the scope of this atlas.

The prognosis for individuals with the above photosensitivity disorders is generally very good, but the scarring in both actinic prurigo and hydroa vacciniforme is permanent.

References

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Chapter 20
Body Art: Tattoos and Piercings

Michael Lee Smith

Introduction

Tattoos and body piercings have been performed since ancient times. They have been used for cultural, social, decorative, and personal reasons, such as the preservation of Polynesian history with tattoos. The popularity of body art has increased substantially over the past 50 years, particularly among teens.

Epidemiology

Estimates of tattoo prevalence range from 5% to 40% in adults and 4% to 13% in adolescents, with the prevalence increasing among teens. Piercings, however, are much more common. The prevalence of piercings among adolescents ranges from 27% to 50%, with ear piercings being most prevalent. Among teens with piercings, earlobe is the most common, followed by ear cartilage, navel (especially in females), and oral and genital regions.

The frequency of complications with body art is very high, with pierced ear cartilage infection seen in 41% and earlobe infection in 30% of adolescent females. Cirks et al. surveyed Emergency Department (ED) visits for piercing complications. They found that 85% of patients presenting to an ED with a piercing complication were female, with adolescents accounting for 55% (median age 13.7 years). Ear piercing accounted for 71% of the ED visits. The median age of patients with infections at the piercing site was 17.4 years [1–4].

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_20

Clinical Findings

Complications of tattooing include skin infections, often due to contaminated equipment or inks, improper sterile technique, and itching with scratching leading to contamination. Bacterial infection has been reported most often, with the majority due to *Staphylococcus aureus* (Fig. 20.1) and *Streptococcus pyogenes*. Atypical mycobacterial infections are seen as well. Cutaneous viral infections are common, with human papillomavirus, herpes simplex virus, and molluscum contagiosum the most frequent. Systemic infections are beyond the scope of this chapter, but readers are directed to the references below. Other tattoo complications include eczematous reactions such as allergic contact due to red cinnabar (mercury sulfide) (Fig. 20.2), lichenoid dermatitis (Fig. 20.3), plaques, or nodules. The latter lesion types may become ulcerated or necrotic. Granulomas (Fig. 20.4), pseudolymphoma, and Koebnerization of pre-existing dermatoses (e.g., psoriasis, lichen planus) have been noted as well.

The most common complications of piercings are infections. These are typically localized skin and cartilage infections. The most common organisms responsible are *Staphylococcus aureus*, *Streptococcus pyogenes*, and *Pseudomonas* sp. Non-infectious complications include keloid formation (Fig. 20.5), hematomas, allergic reactions (most commonly to nickel), and tearing of the skin (Fig. 20.6). Tears occur most often with ear, nose, mouth, nipple, and navel piercings. Oral piercings have a significant potential for complications including dental injury, increased salivation, gingival inflammation, and/or recession, eating difficulty, and speech problems. Genital piercings frequently become infected and/or painful [2–5].

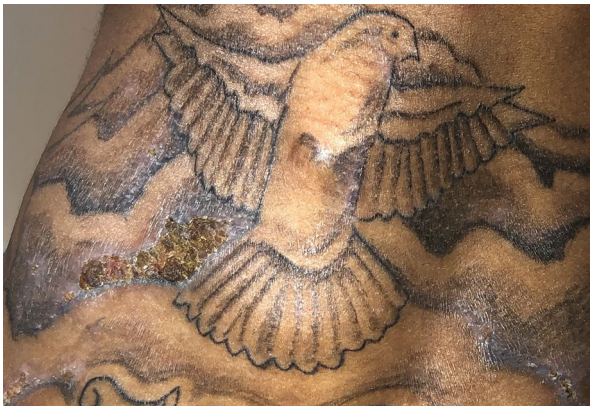

Fig. 20.1 Staphylococcal infection in a tattoo showing erythematous patches with crusting. (Photograph courtesy of Neerav Desai, MD)

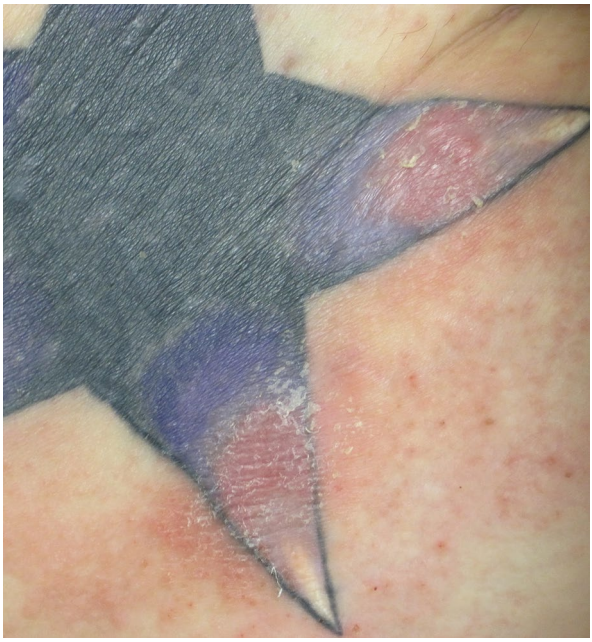

Fig. 20.2 Allergic reaction to cinnabar (red pigment) in 20-year-old. The red areas are scaly edematous plaques

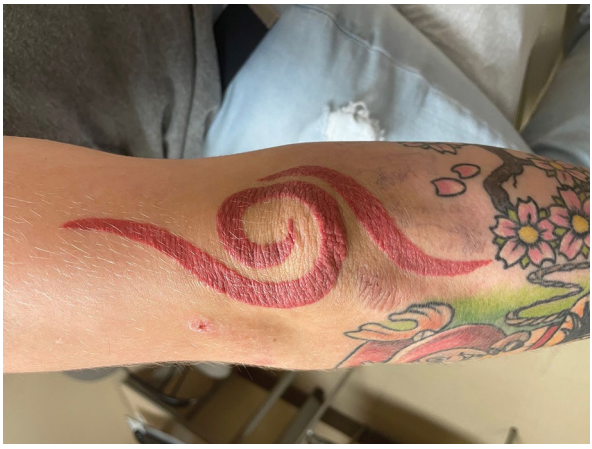

Fig. 20.3 Lichenoid dermatitis in a tattoo with the entire red area as an edematous plaque. Surrounding areas exhibit papular and plaque-like erythema, indicating the extension of the lichenoid dermatitis. (Photograph courtesy of Neerav Desai, MD)

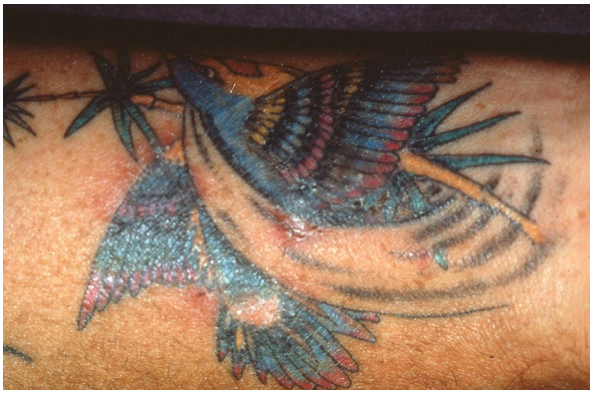

Fig. 20.4 Granulomatous reaction

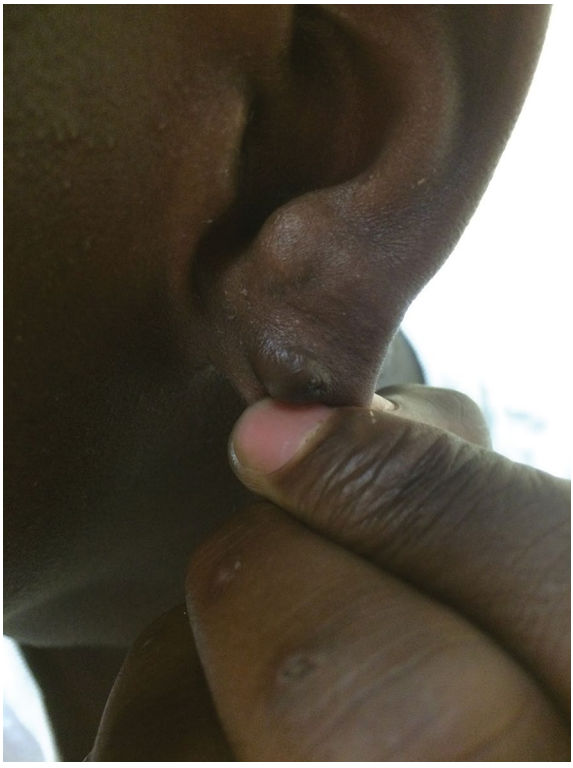

Fig. 20.5 Earlobe keloid in an adolescent secondary to piercing. The nodule is firm and mildly tender, with extension through the lobe. (Photograph courtesy of Neerav Desai, MD)

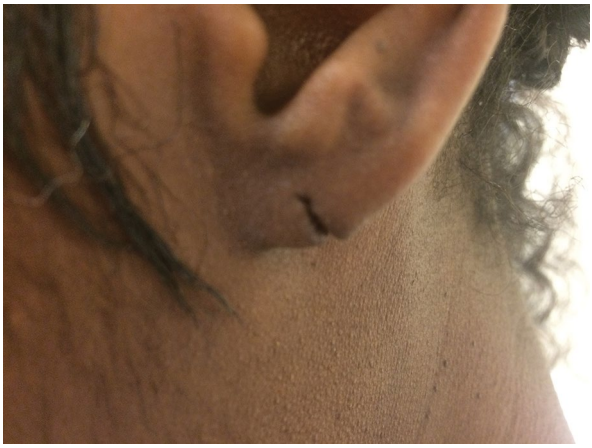

Fig. 20.6 Split earlobe from hoop earring. (Photograph courtesy of Neerav Desai, MD)

Laboratory

If bacterial infection is suspected, culture and sensitivity should be obtained. Atypical mycobacterial infections are common in tattoos, especially with granulomatous lesions. Appropriate cultures should be obtained for this as well. If viral infections are suspected, viral cultures or PCR studies may be helpful.

Treatment

Infected tattoos should be treated with appropriate topical or oral antimicrobial agents. With infected piercings, aftercare solutions should be discontinued, and hardware/jewelry should be removed. Empiric antimicrobial treatment should be administered [3–5].

Prognosis

Prognosis is generally very good for complications of body art. However, there may be permanent sequelae such as scars, disfigurement from torn tissue, or inability to use adornments such as certain jewelry if a nickel allergy develops.

References

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2. Conte S, et al. Complications of body piercings: a systematic review. Cutis. 2023;112:139–45.
3. Desai N, et al. Tattoos and piercings in female adolescents and young adults. J Pediatr Adolesc Gynecol. 2023;36(1):14–7.
4. Gualdi G, et al. Tattoo: ancient art and modern problems. J Cosmet Dermatol. 2021;20:602–4.
5. Kim MM, et al. Ear-piercing complications in children and adolescents. Can Fam Physician. 2022;68:661–3.

Part VI
Genodermatoses and Genetic Conditions

Chapter 21
Neurofibromatosis

Patricia Treadwell

Introduction

Neurocutaneous syndromes consist of neurologic issues in addition to cutaneous findings. One of the most common disorders that may be identified in the adolescent age group is neurofibromatosis. Adolescents with neurofibromatosis are predisposed to both benign and malignant tumors. Neurofibromatosis is an example of a RASopathy. RASopathies are a group of genetic conditions caused by changes in the genes that control the Ras/mitogen-activated protein kinase (Ras/MAPK) pathway.

Epidemiology

Neurofibromatosis 1 (NF1) occurs in approximately 1:2500–1:3000 individuals. NF1 occurs as a result of a germline mutation in one of the 2 alleles of the tumor suppressor gene NF1 on chromosome 17q11.2 (Ly KI et al). Neurofibromatosis 2 (NF2) is less common than NF1, occurring in 1:25,000 individuals. The mutation associated with NF2 occurs in neurofibromin-2 on chromosome 22q12. Patients with NF2 have acoustic neuromas- most typically bilateral. The inheritance of both NF1 and NF2 is considered to be autosomal dominant; however, the rate of spontaneous mutations for NF1 is approximately 42%.

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_21

Clinical Findings

Cutaneous manifestations are significant in NF1. They represent the majority of the diagnostic criteria. The cutaneous findings with NF2 are less prominent. The most common cutaneous finding in NF1 is a café-au-lait macule (CALM) (Fig. 21.1). In skin of color, the café-au-lait macules will be darker in color (Fig. 21.2). Other cutaneous findings that are part of the diagnostic criteria are neurofibromas, freckling of skinfolds (Fig. 21.3), Lisch nodules, and plexiform neuromas. Plexiform neuromas have an increased risk of transformation into a Malignant Peripheral Nerve Sheath Tumor (MPNST). Neurofibromas grow more rapidly during puberty.

Other tumors noted are optic pathway gliomas, rhabdomyosarcomas, and increased breast cancer in women <50 years of age.

Other associated findings with NF1 are pruritus, macrocephaly, learning disabilities, hypertension, skeletal abnormalities (osteopenia, scoliosis, and sphenoid wing dysplasia), and choroidal abnormalities.

As mentioned above, NF2 has less prominent cutaneous findings when compared to NF1. Associated findings with NF2 are meningiomas of the brain and schwannomas of the dorsal roots of the spinal cord.

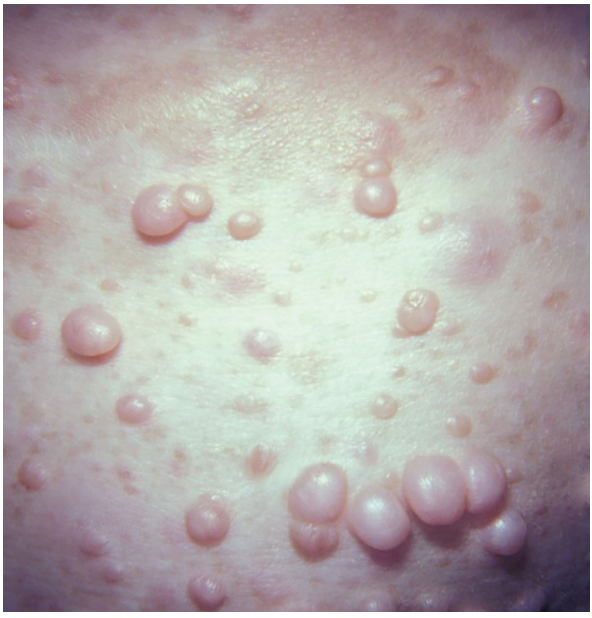

Fig. 21.1 Multiple neurofibromas with a CALM in the upper part of the photograph in a patient with NF1

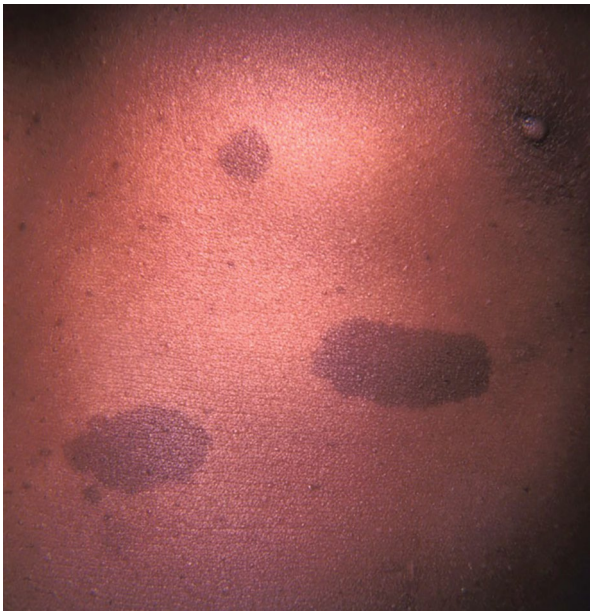

Fig. 21.2 CALM which is a more brown in skin of color

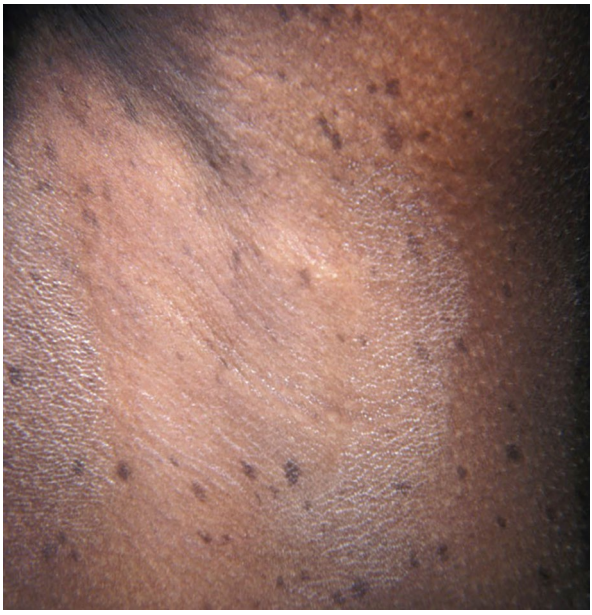

Fig. 21.3 Axillary freckling in a patient with NF1

Laboratory

Genetic testing may be indicated when the diagnostic criteria are not confirmatory or mosaicism is suspected. Tumor surveillance is an ongoing issue.

Treatment

In light of the predisposition for tumors, health supervision includes tumor surveillance, especially if specific findings are noted. Tumor-specific treatment may include surgical excision and/or chemotherapy. Radiation therapy is used with caution because of the secondary malignancy risk.

Prognosis

Life expectancy is shortened due to malignancy and has been estimated to be >10 years less than the unaffected population.

Suggested Readings

Kehrer-Sawatzki H, et al. Challenges in the diagnosis of neurofibromatosis type 1(NF1) in young children facilitated by means of revised diagnostic criteria including genetic testing for pathogenic NF1 gene variants. Hum Genet. 2022;141:177–91.

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Ly KI, et al. The diagnosis and management of neurofibromatosis type 1. Med Clin North Am. 2019;103:1035–54.

Chapter 22
Tuberous Sclerosis Complex

Patricia Treadwell

Introduction

Tuberous sclerosis complex (TSC) is a neurocutaneous disorder characterized by mutations in the TSC1 gene (encoding of hamartin) or TSC2 gene (encoding of tuberin). The mutations result in dysregulation of cellular hyperplasia.

Epidemiology

TSC occurs at a rate of 1:6000 to 1:10,000 live births. TSC1 is located on chromosome 9q34 and encodes hamartin. TSC2 is located on chromosome 16p13 and encodes tuberin. Inheritance of TSC is considered to be autosomal dominant; however, the rate of spontaneous mutations is approximately 70%.

Clinical Findings

Cutaneous findings include hypomelanotic macules (ash leaf spots), facial angiofibromas (Fig. 22.1), periungual fibromas (Fig. 22.2), and connective tissue nevi—fibrous cephalic plaque (on the head and neck) (Fig. 22.3) or shagreen patch (often located on the lower back).

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_22

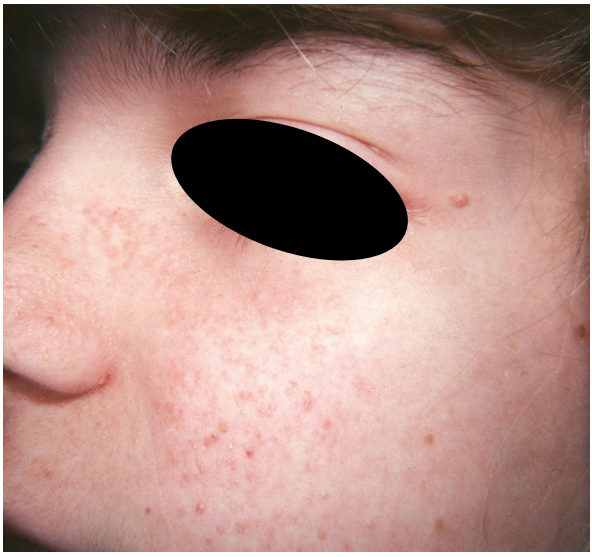

Fig. 22.1 Facial angiofibromas

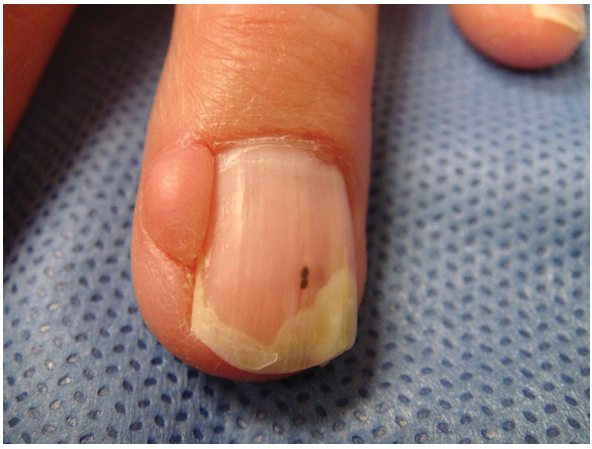

Fig. 22.2 Periungual fibroma

Other associated findings are TSC-Associated Neuropsychiatric Disorders (TAND), including epilepsy, learning difficulties, autism, and attention deficit disorders, among others.

Hamartomas that may develop are retinal hamartomas, subependymal giant cell tumors (SGCT), cardiac rhabdomyomas, renal and non-renal angiomyolipomas (AML), and pulmonary lymphangiomyomatosis (LAM).

Dental enamel pits and intraoral fibromas may be noted.

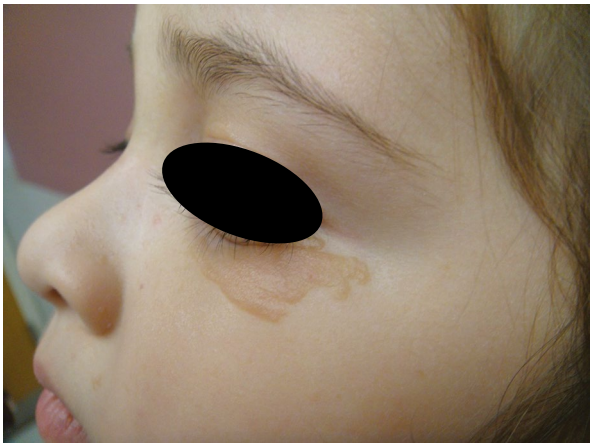

Fig. 22.3 Fibrous cephalic plaque

Laboratory

Imaging may show "cortical tubers" (occurring in about 80% of TSC patients), which are composed of abnormal neurons and glia.

Treatment

Specific guidelines for surveillance assist in the management of the hamartomas and associated findings. Consultation with neurology can be helpful to treat epilepsy. Topical or systemic mTOR inhibitors have been useful for specific complications.

Prognosis

Although the hamartomas are typically benign, they may cause a mass effect.

Suggested Readings

1. Northrup H, et al. Updated international tuberous sclerosis complex diagnostic criteria and surveillance and management recommendations. Pediatr Neurol. 2021;123:50–66.
2. Randle SC. Tuberous sclerosis complex: a review. Pediatr Ann. 2017;46:e166–71.

Chapter 23
Erythropoietic Protoporphyria

Julie Prendiville

Introduction

Erythropoietic protoporphyria (EPP) is a rare genetic disorder of heme biosynthesis characterized clinically by acute pain on sun exposure.

Epidemiology

EPP is the most common porphyria of childhood. It may present or be first diagnosed in adolescence.

Inheritance is autosomal recessive. It is caused by a combination of loss of function mutations and low expression allelic variants in the *FECH* gene. Deficiency of the enzyme ferrochelatase leads to the accumulation of protoporphyrin IX in circulating erythrocytes. Following exposure to sunlight, these protoporphyrins cause acute cutaneous phototoxicity. Accumulation of protoporphyrins in the liver may result in hepatotoxicity and cholestasis.

X-linked protoporphyria, caused by mutations in *ALAS2,* has a similar clinical presentation to EPP.

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_23

Clinical Findings

Symptoms occur shortly after sun exposure, most commonly on the face, dorsal hands, arms, and feet. Objective clinical findings are often absent despite excruciating pain. Variable erythema and edema may be observed. Figure 23.1 Erosions and crusting of the skin are uncommon. Chronic atrophic changes on the face or dorsal hands may result from severe phototoxic reactions or recurrent sun exposure.

Laboratory

Laboratory investigation shows elevated levels of free erythrocyte protoporphyrin. The diagnosis may be confirmed by sequencing of the *FECH* (or *ALAS2*) gene. Patients require monitoring for liver dysfunction, anemia, and vitamin D deficiency.

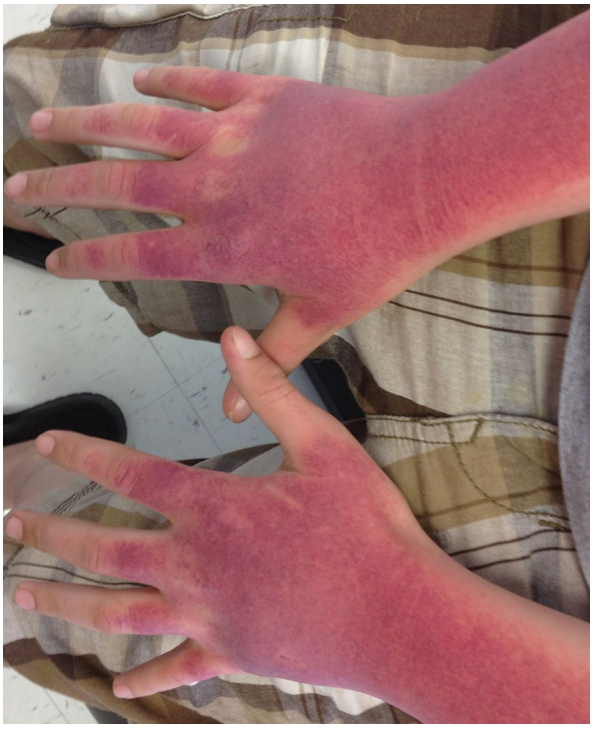

Fig. 23.1 Erythema of dorsal hands in EPP

Treatment

Sun avoidance is the mainstay of treatment. A UV meter may be helpful to indicate the times and months when protective clothing, hats, gloves, umbrellas, and tinted windows are required. Topical sunscreens are of limited benefit. Oral beta-carotene and alfamelanotide have been reported to improve sun tolerance.

Prognosis

Social isolation and lifestyle changes may significantly impair quality of life. A small subset of patients has severe progressive liver disease.

Suggested Readings

1. Balwani M. Erythropoietic protoporphyria and X-linked protoporphyria: pathophysiology, genetics, clinical manifestations, and management. Mol Genet Metabol. 2019;128:298–303.
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Chapter 24
Nevus Sebaceus

Julie Prendiville

Introduction

A nevus sebaceus is a congenital hamartoma of cutaneous adnexal structures, specifically the hair follicles and sebaceous glands. It is commonly located on the scalp or face. The appearance of a nevus sebaceus may change around puberty and can present as a concern in adolescence.

Epidemiology

An isolated nevus sebaceus occurs in approximately 0.3% of newborns. It is caused by a somatic mosaic activating mutation in the *HRAS*, or less commonly *KRAS*, gene. An extensive or multifocal nevus sebaceus may be associated with other developmental anomalies, as in the rare Schimmelpenning syndrome.

Clinical Findings

The typical nevus sebaceus is a yellow-orange plaque with a pebbly or velvety surface located on the scalp or face (Fig. 24.1). It varies in size from one to several centimeters and can be round, oval, or linear in shape. Lesions on the scalp present as a congenital area of circumscribed alopecia. A thickened or verrucous/warty

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https://doi.org/10.1007/978-3-031-86177-2_24

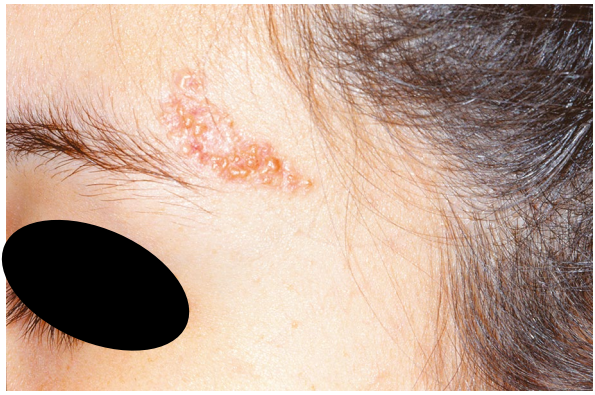

Fig. 24.1 Nevus sebaceus on the left temple

appearance may develop in adolescence when the sebaceous glands enlarge with puberty.

Laboratory

Histopathology following excision or biopsy shows immature hair follicles, numerous hyperplastic sebaceous and apocrine glands, and overlying epidermal hyperplasia.

Treatment

Treatment is by surgical excision. Continued observation is an option as the risk of malignancy is considered low.

Prognosis

Secondary tumors may develop within a nevus sebaceus, usually in adolescence or adult life. Most are benign adnexal growths (e.g., trichoblastoma, syringocystadenoma papilliferum), but malignant tumors can occur.

Suggested Readings

1. Aslam A, et al. Nevus sebaceus: a mosaic RASopathy. Clin Exp Dermatol. 2014;39:1–6.
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Chapter 25
Nevus of Ota and Ito

Patricia Treadwell

Introduction

Nevus of Ota (Nevus fuscoceruleus ophthalmomaxillaris) and Ito (Nevus fuscoceruleus acromiodeltoideus) are lesions that are examples of dermal melanocytosis. Dermal melanocytosis results from abnormal migration of neural crest cells with subsequent pigmentary changes from the ectopic melanocytes in the dermis.

Epidemiology

Nevus of Ota and Ito are more common in patients with African or Asian ancestry, although they can occur in all races [1]. Females are affected more often than males. There are two peaks of presentation, one at birth, with a second peak near puberty.

Clinical Finding

The distribution of pigment in the Nevus of Ota typically corresponds to the ophthalmic and maxillary branches of the trigeminal nerve, less commonly corresponding to the mandibular branch. Two-thirds of patients will have ocular pigmentation (oculodermal melanocytosis). Figure 25.1 In Nevus of Ito, the distribution involves the posterior supraclavicular area and lateral neck. In both entities, dermal

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https://doi.org/10.1007/978-3-031-86177-2_25

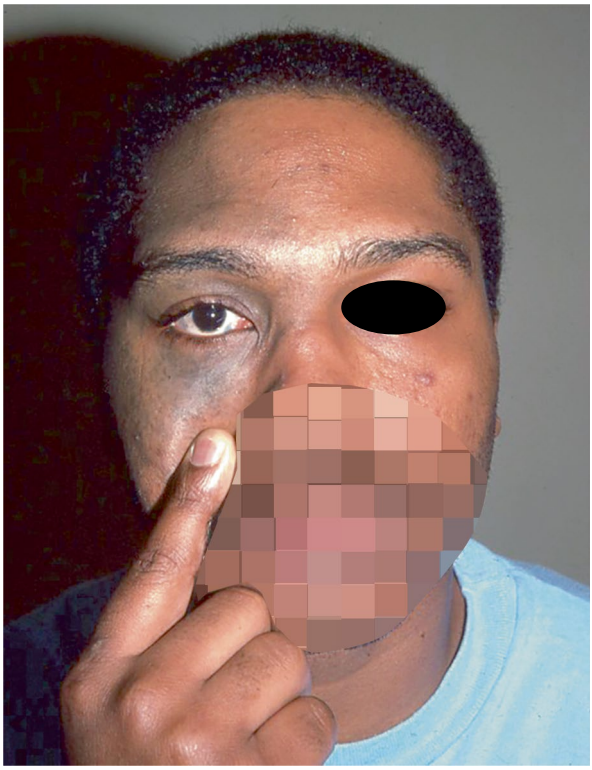

Fig. 25.1 Patient with Nevus of Ota. Also has ocular pigmentation

melanocytosis is responsible for the clinical appearance, with distribution being the main difference between them.

Laboratory

Histopathology shows pigmented dendritic melanocytes dissecting dermal collagen bundles in the dermis.

Treatment

A variety of lasers have been used to treat Nevus of Ota and Ito. Because of the increased risk of glaucoma and a rare occurrence of uveal melanoma in eyes with oculodermal melanocytosis, ophthalmologic exams (including dilated fundoscopic exam) are recommended every 6 months [2].

Prognosis

Adolescents without complications such as glaucoma have a good prognosis. As mentioned above, malignant melanoma occurring in the skin, meninges, and/or eye associated with these lesions is quite rare.

References

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2. Berelli S, et al. Nevus of Ota in 8-year-old male. Pan Afr Med J. 2023;45(81):10.11604/pamj.2023.45.81.40463.

Part VII
Tumors and Nodular Lesions

Chapter 26
Pilomatrixoma

Patricia Treadwell

Introduction

Pilomatrixoma is a benign adnexal tumor. It is also known as a calcifying epithelioma of Malherbe. It consists of benign hyperplasia of the hair follicle matrix cells.

Epidemiology

Pilomatrixomas may occur at any age; nonetheless, they are most often noted prior to age 20. Generally, these lesions do not have a known genetic predisposition; however, multiple lesions have been associated with myotonic dystrophy, Gardner syndrome, Rubenstein-Taybi syndrome, and trisomy 9 (among others).

Pilomatrixoma carcinoma is very rare and has been noted in middle-aged and elderly adults. Mutations in Wnt (wingless and Int-1) signaling pathways have been identified in pilomatrixoma and pilomatrixoma carcinomas. These mutations and the downstream molecules have the potential to be used as diagnostic and prognostic markers [1].

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https://doi.org/10.1007/978-3-031-86177-2_26

Clinical Findings

A pilomatrixomas is typically a solitary nodule and noted most often on the face or proximal extremities [2]. The lesions tend to be slow growing, firm lesions with an irregular surface. Figure 26.1 Some of the lesions will have a violaceous discoloration initially. Pilomatrixomas often develop calcium deposits and become very hard.

Laboratory

When a biopsy is performed, the findings are proliferation of basaloid cells, shadow cells, or both [3].

Treatment

The lesions will sometimes resolve spontaneously over a period of several months. If they become repeatedly flared or superinfected, they can be surgically excised.

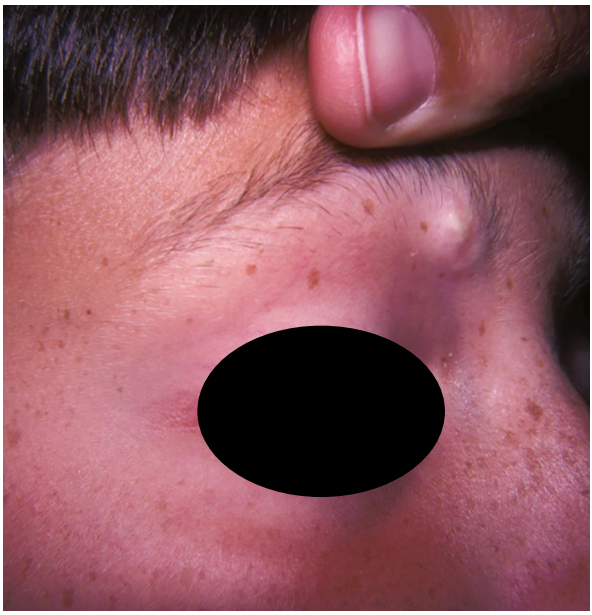

Fig. 26.1 Note the irregular surface of this pilomatrixoma below the medial eyebrow

Prognosis

When the lesions resolve spontaneously, minimal scarring is noted. When they are surgically excised, a healed surgical scar will result.

References

1. Kamil ZS, et al. Early pilomatrixoma carcinoma: a case report with emphasis on molecular pathology and review of literature. Cutis. 2021;108:E24–8.
2. Pelizzari M, et al. Ultrasound findings in 156 children with 169 pilomatricomas. Pediatr Radiol. 2021;51:2038–46.
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Chapter 27
Epidermal Inclusion Cyst

Patricia Treadwell

Introduction

Epidermal inclusion cysts (EICs) (aka epidermoid cysts) are cystic lesions with an epithelial lining that can develop in adolescence and early adulthood. Some may develop following the inflammatory phase of nodular acne lesions.

Epidemiology

In particular, EICs following acne lesions may be noted on the upper back, chest, neck, and face. Other EIC's can be seen anywhere on the body. Most often the lesions are solitary when not associated with a genetic syndrome.

Clinical Findings

An EIC is a slow-growing cystic lesion with components above and below the surface of the skin (Fig. 27.1). Some lesions may have a central punctum, which was previously a follicular opening.

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_27

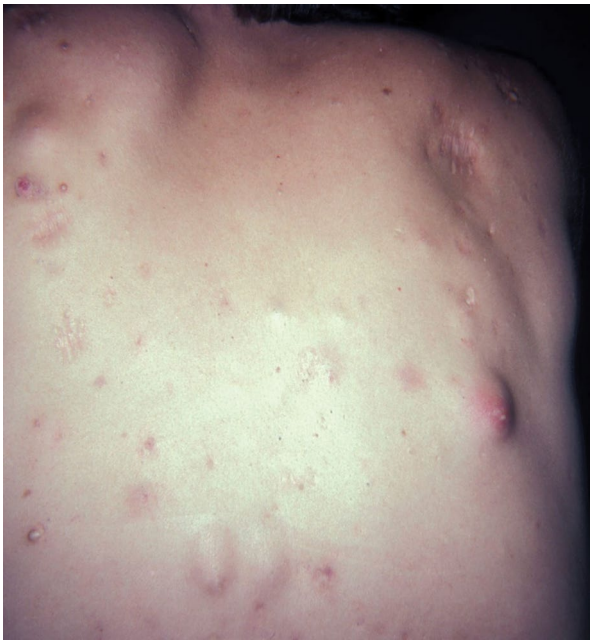

Fig. 27.1 Greater than five EICs in addition to three scarred areas from I&D on the back of a patient with acne

Laboratory

The material expressed from an EIC has been described as "cheesy." The contents of the EICs are generally sterile if cultured. Histologically, an epidermoid cyst is lined by an epithelial cell wall. The epithelium is stratified squamous epithelium resembling epidermis and includes a granular layer and keratin lamellae in the lumen [1].

Treatment

Treatment of asymptomatic lesions may not be necessary. The lesions tend to be slow growing, however. They can exhibit inflammation or develop a secondary bacterial infection. Incision and drainage (I&D) tends not to be an effective treatment, especially if a portion of the lining is still present. Complete surgical excision of the cyst with its lining can result in complete resolution.

Prognosis

The untreated lesions tend to be persistent but may become a cosmetic issue. Scarring may result from I&D or surgical excision.

Reference

1. Hoang VT, et al. Overview of epidermoid cyst. Eur J Radiol Open. 2019;6:291–301.

Chapter 28
Pyogenic Granuloma

Patricia Treadwell

Introduction

The term pyogenic granuloma is unfortunately somewhat of a misnomer. The lesion is not typically pyogenic, nor is it a granuloma. The lesion has also been labeled a lobular capillary hemangioma and is a benign acquired vascular lesion.

Epidemiology

Pyogenic granulomas are most often noted in childhood and adolescence. The etiology is theorized to be neovascularization. This can occur following trauma or can also be seen arising within a Nevus Flammeus [1].

Clinical Findings

The lesion usually is a reddish to violaceous lobulated nodule (Fig. 28.1). The surface may become eroded. In some cases, it can become pedunculated (Fig. 28.2). The history given by the patient and/or caregivers is that it often spontaneously bleeds. They are most often located on the fingers, face, and oral mucosa.

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_28

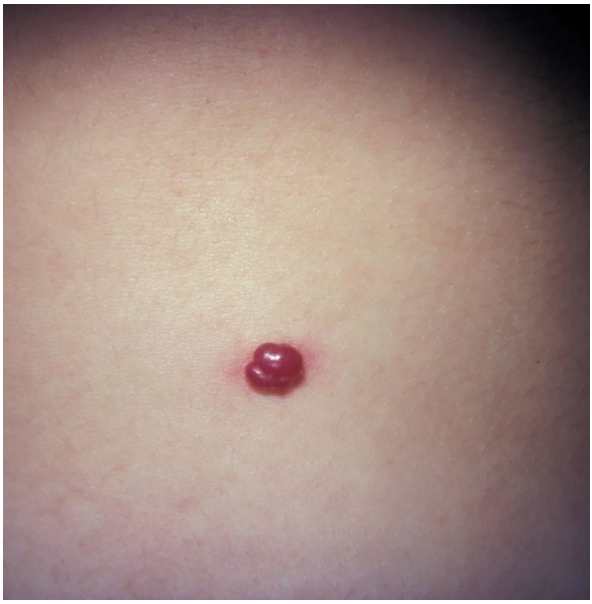

Fig. 28.1 Lobulated pyogenic granuloma

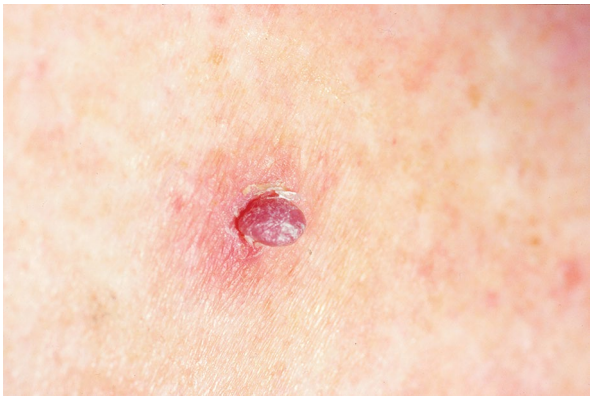

Fig. 28.2 Pedunculated pyogenic granuloma

Laboratory

Histopathology shows a proliferation of capillaries and fibroplasia.

Treatment

Surgical excision with curettage and/or cautery was previously considered the best option; however, recently, topical beta blockers have been noted to be useful [2].

Prognosis

Incomplete excision may result in recurrence.

References

1. Rancan A, et al. Pyogenic granuloma arising within capillary malformations in children: a case report and literature review. Dermatol Reports. 2021;13:9115.
2. Filoni A, et al. Topical beta-blockers in dermatologic therapy. Dermatol Ther. 2021;34:e15016.

Suggested Reading

Gaw C, et al. Management of bleeding pyogenic granulomas in acute care settings. J Emerg Med. 2022;27:374–6.

Chapter 29
Cutaneous Malignancies in Adolescents

Michael Lee Smith

Introduction

Primary cutaneous malignant tumors, while uncommon in adolescents, include basal cell carcinoma (BCC), squamous cell carcinoma (SCC), malignant melanoma (MM) and cutaneous lymphoma or lymphoproliferative conditions. Other rare systemic cancers may have cutaneous lesions, but these are beyond the scope of this chapter.

Epidemiology

Non-melanoma skin cancer (NMSC) generally includes BCC and SCC. While both are rare in adolescents, BCC is the more common of the two. Huang et al. showed that 72% of 124 pediatric and adolescent patients with NMSC were 11 years old or older. Median age at diagnosis was 14 for SCC and 12 for BCC [1].

Genetic conditions are frequent predisposing factors for the development of NMSC. BCC is more often seen in basal cell nevus, Bazex-Christol-Dupre, and Rombo, syndromes. SCC is associated with xeroderma pigmentosum (XP), recessive dystrophic epidermolysis bullosa (RDEB), Bloom syndrome, dyskeratosis congenita, and epidermodysplasia verruciformis. NMSC also are more frequent in primary immunodeficiencies. Other medical conditions that predispose teens to NMSC include prolonged immunosuppression, radiation therapy, chemotherapy, and the use of voriconazole [1, 2].

Most pediatric melanoma presents in adolescence, with almost 80% of patients aged 15–19 years old. Most cases of malignant melanoma (MM) in adolescents are

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https://doi.org/10.1007/978-3-031-86177-2_29

sporadic. Risk factors include increased ultraviolet light exposure, history of sunburns, tanning bed use, and immunosuppression. Those with Fitzpatrick skin phototypes I and II have the highest risk. Other predisposing factors include higher nevus burden, large congenital melanocytic nevus, and history of atypical nevi. Familial atypical mole melanoma syndrome due to CDKN2A mutations confers an estimated 11% risk, which is 28-fold higher than the general population. Fortunately, there has been a favorable declining trend in adolescent MM in recent years, with an average of 11% decrease in prevalence per year from 2003 to 2010 [1, 2].

Primary cutaneous lymphomas (PCL) and cutaneous lymphoproliferative disorders (LPD) are very rare in adolescents, with fewer than one per million per year. The most common PCLs are primary cutaneous CD30+ LPD, with lymphomatoid papulosis (LyP) seen more frequently than anaplastic large cell lymphoma (ALCL). The next most common PCL is mycosis fungoides. These PCL/LPD conditions must be distinguished from secondary skin involvement by systemic lymphomas. Adolescent and pediatric PCL may be seen with increased frequency in Asian, Pacific Islander and African American patients [2, 3].

Clinical Findings

BCC typically appears as a macule, papule, or thin plaque, usually shiny and translucent (Fig. 29.1). There may be surface telangiectasia. The distribution most often involves the face, scalp, and back.

SCC is clinically similar to those seen in adults, with slowly enlarging patches, nodules, or plaques. More advanced lesions may present as ulcerated nodules or plaques. Non-healing wounds should be assessed for proliferative lesions, particularly in teens with any of the above risk factors.

Melanoma in adolescents follows different clinical patterns than in adults. The traditional ABCDE features (asymmetry, border irregularity, color variegation,

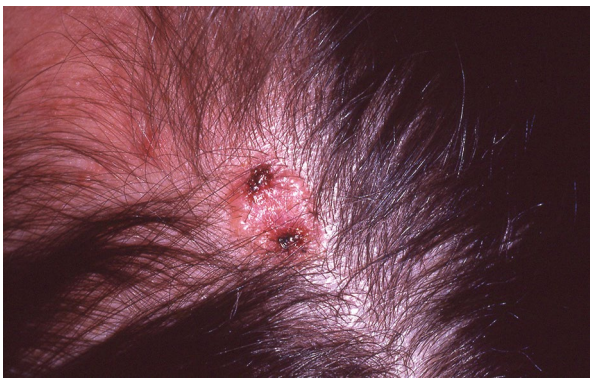

Fig. 29.1 12-year-old with basal cell carcinoma, with no genetic predisposition. The lesion is a pearly translucent plaque with central thinning and mild crusting along the edges. Numerous sunburns along her hair part may have been the trigger

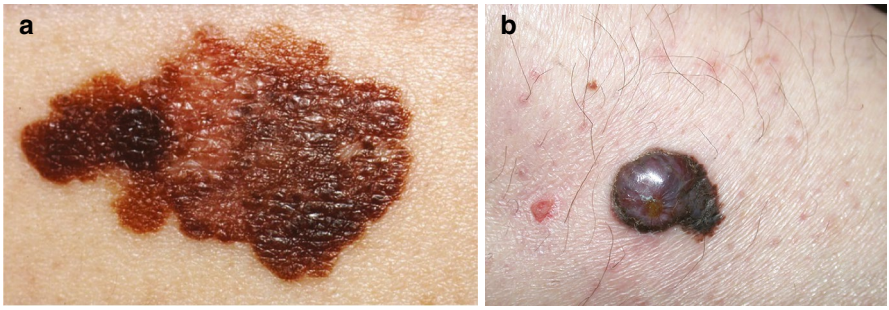

Fig. 29.2 (**a**) Superficial spreading melanoma. (**b**) Nodular melanoma

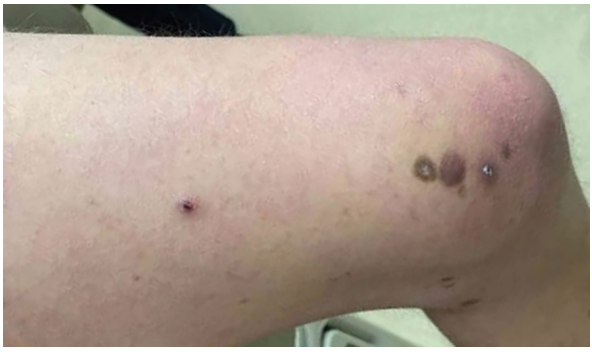

Fig. 29.3 17-year-old male with lymphomatoid papulosis. (Photograph courtesy of John A. Zic, MD)

diameter greater than 7 mm, or evolving rapidly) may be seen in adolescents. However, those criteria pick up only about half of adolescent MM. Modified ABCD criteria have been proposed, including amelanotic, bleeding, bump, color uniformity, de novo, and any diameter (Fig. 29.2a, b) [1, 2]. Please refer to the references below for further elucidation. These modified criteria still miss a significant number but tend to pick up Spitzoid melanoma better than did the older adult criteria. High index of suspicion is important in adolescents, particularly those with any of the above risk factors.

LyP tends to have chronic, recurrent, or self-resolving lesions that may be disseminated or grouped (Fig. 29.3). They range from small papules to large nodules, often with secondary ulceration or a necrotic eschar. Trunk and extremity involvement is most common. ALCL most often present with one nodule or tumor (Fig. 29.4). Mycosis fungoides generally follows a chronic, indolent course. MF starts with scaly patches, plaques, or poikiloderma, most often in non-sun-exposed areas (Fig. 29.5). Hypopigmented MF is more common than in adults (Fig. 29.6a–c).

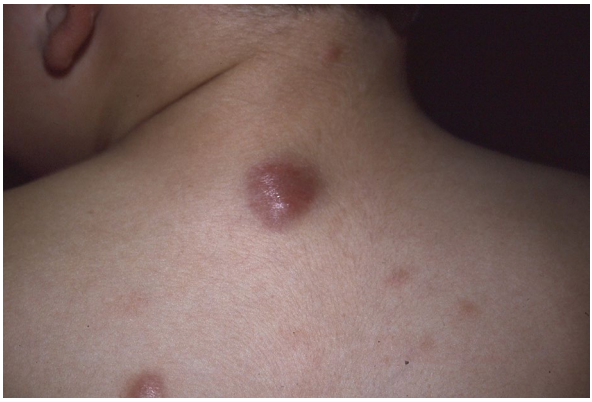

Fig. 29.4 An eleven year-old boy with CD 30+ anaplastic large cell lymphoma, showing firm red indurated nodules

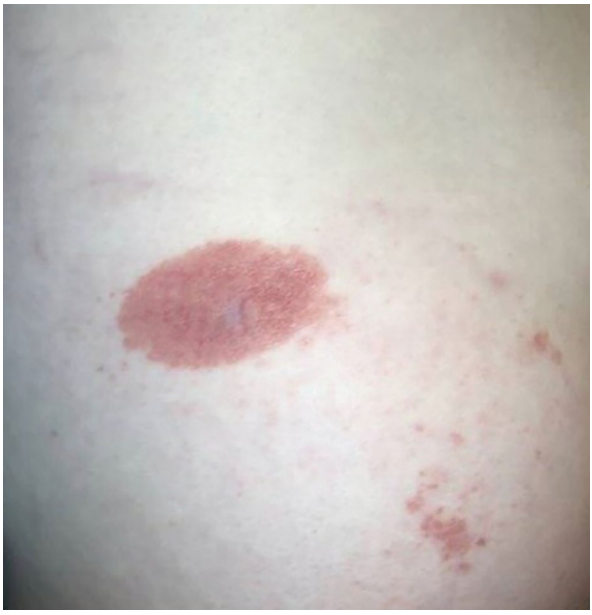

Fig. 29.5 16 year old with pigmented purpuric variant of mycosis fungoides on the hip. (Photograph courtesy of John A. Zic, MD)

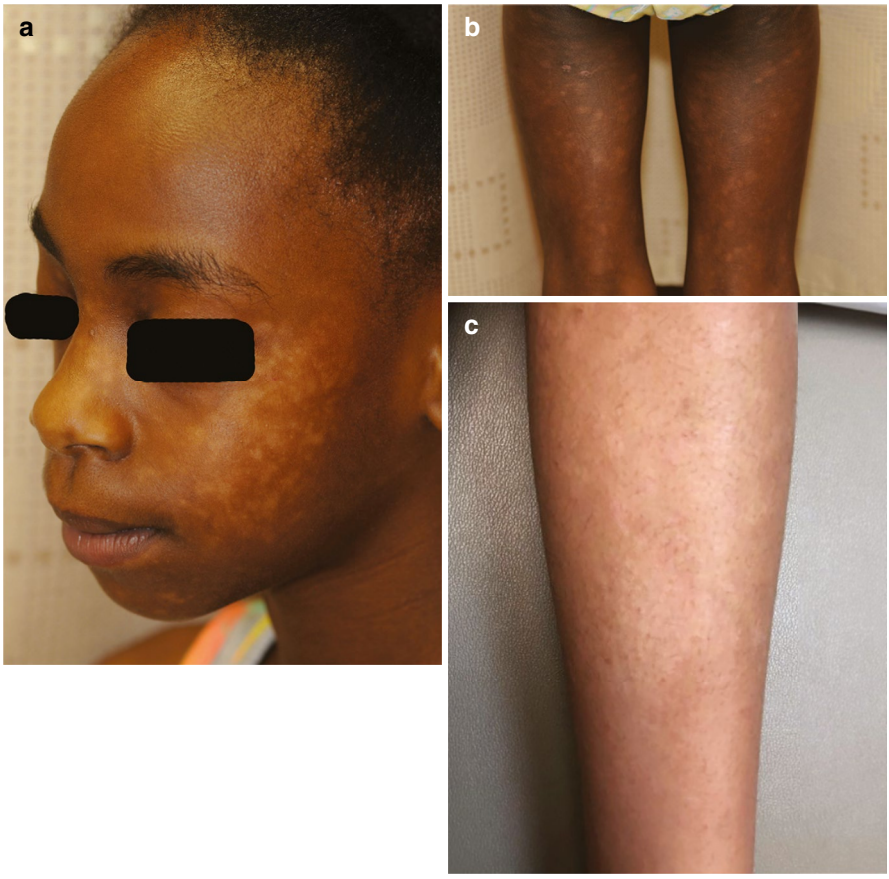

Fig. 29.6 (**a**) 11 year old female with hypopigmented mycosis fungoides. (Photograph courtesy of John A. Zic, MD). (**b**) 11 year old female with hypopigmented mycosis fungoides. (Photograph courtesy of John A. Zic, MD). (**c**) 14 year old male with hypopigmented mycosis fungoides on the shin. (Photograph courtesy of John A. Zic, MD)

Laboratory

Skin biopsy is required for the diagnosis of all cutaneous malignancies in adolescents. Immunohistochemical staining often is used to further delineate the conditions. Primary cutaneous lymphomas often require T-cell receptor gene rearrangement clonality studies, although clonality is less consistently found in adolescents as compared with adults.

Treatment

BCC may be treated with local excision, electrodessication and curettage, 5-fluorouracil, tazarotene, and imiquimod. SSC is most often is treated with local excision. Melanoma is staged by histologic depth (Breslow depth, Clark level) with or without sentinel lymph node assessment. Lower risk (shallow) lesions are treated with wide local excision. PCLs are treated with phototherapy, methotrexate, or brentuximab. One exception is ALCL, which may require excision or radiation therapy.

Prognosis

Prognosis varies widely in NMSC, dependent upon comorbidities. As a general rule, BCC has a very good prognosis. SCC, on the other hand, has a better prognosis if non-syndromic or due to sun exposure. However, in several genetic conditions (e.g. RDEB, XP) it is a frequent cause of mortality.

Prognosis in MM depends upon several factors. Earlier diagnosis and shallow depth tend to be more favorable. Overall, MM tends to be more aggressive in adolescents than in adults.

PCL/LPD prognosis is linked to the specific subtype. LyP and MF tend to have the most favorable prognosis in PCL/LPD.

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2. Moustafa D, et al. Trends in pediatric skin cancer. Curr Opin Pediatr. 2020;32(4):516–23.
3. Stokke J, et al. Diagnosis and management of cutaneous lymphomas and lymphoid proliferations in children, adolescents and young adults. Best Pract Res Clin Haematol. 2023;36(1):101448.

Part VIII
Lymphocytic Disorders

Chapter 30
Pityriasis Lichenoides

Julie Prendiville

Introduction

Pityriasis lichenoides is an inflammatory skin disorder of unknown etiology. There are two variants with overlapping features: (1) pityriasis lichenoides chronica (PLC) and (2) pityriasis lichenoides et varioliformis acuta (PLEVA). Febrile ulceronecrotic Mucha-Haberman disease is a severe form of PLEVA with systemic manifestations.

Epidemiology

The exact prevalence is unknown. Pityriasis lichenoides predominantly affects children and young adults, including adolescents.

Clinical Findings

Widespread erythematous papules with overlying adherent scale are characteristic (Figs. 30.1 and 30.2). PLEVA papules show central hemorrhagic ulceration and crusting and may be mistaken for varicella infection or insect bites. White, or pigmented, macules are seen where previous lesions have resolved. Symptoms are variable and may be absent. The rare febrile ulceronecrotic Mucha-Haberman disease is

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_30

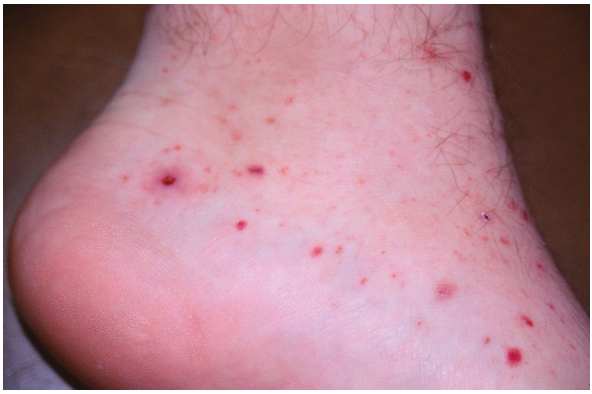

Fig. 30.1 PLEVA lesions of the lateral foot

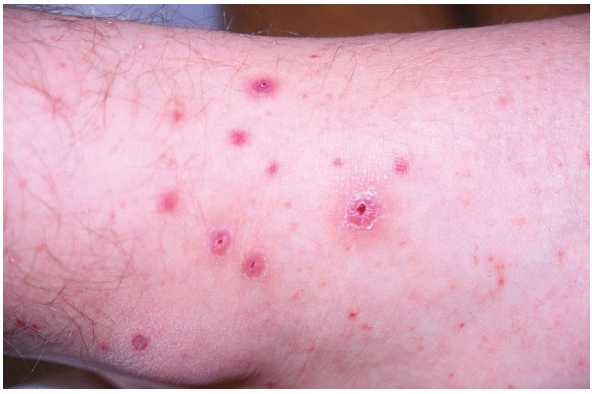

Fig. 30.2 PLEVA lesions of the ankle showing petechiae and scale

characterized by large, painful ulcerating lesions associated with high fever and signs and symptoms of multisystem disease.

Laboratory

The diagnosis is established by skin biopsy. Laboratory markers of inflammation are elevated in febrile ulceronecrotic Mucha-Haberman disease.

Treatment

Prolonged courses of an oral antibiotic (erythromycin or tetracycline) or narrow-band UVB phototherapy are the most commonly prescribed treatments. Systemic anti-inflammatory drugs are reserved for cases of recalcitrant or severe disease.

Prognosis

The course is usually benign and self-limiting with a waxing and waning course over several months or years. Post-inflammatory hypopigmented macules are common in both variants. Ulcerating lesions result in atrophic scarring. There are rare instances of PLC progressing to mycosis fungoides, a type of cutaneous T-cell lymphoma.

Suggested Reading

1. Geller L, et al. Pityriasis lichenoides in childhood: review of clinical presentation and treatment options. Pediatr Dermatol. 2015;32:579–92.

Chapter 31
Pigmented Purpuric Dermatosis

Julie Prendiville

Introduction

Pigmented Purpuric Dermatosis (aka pigmented purpura) refers to a group of benign skin disorders characterized by patches of purpura and petechiae with pigmentation resulting from hemosiderin deposition. The lower extremities are primarily involved. The appearance can be a cosmetic concern for adolescents.

Epidemiology

Pigmented purpura is relatively uncommon. It is seen in all age groups. The cause is unknown.

Clinical Findings

At least five different subtypes are described, but there may be overlap between these disorders. The subtypes are (1) Schamberg disease (progressive pigmentary purpura); (2) Majocchi disease (purpura annularis telangiectoides); (3) Lichen aureus; (4) Gougerot- Blum purpura; (5) Eczematoid-like purpura (eczematoid-like purpura of Doucas and Kapetanakis).

Schamberg disease (progressive pigmentary purpura) is the most common variant in children. It presents with non-palpable reddish-brown areas of purpura and

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P. Treadwell et al. (eds.), *Atlas of Adolescent Dermatology*,
https://doi.org/10.1007/978-3-031-86177-2_31

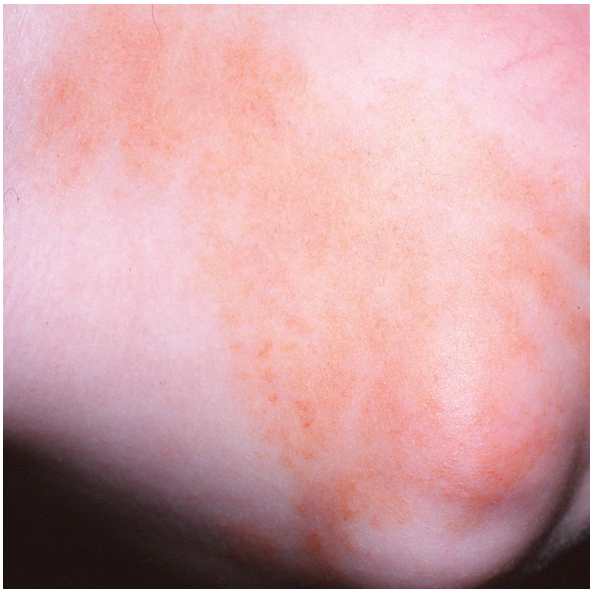

Fig. 31.1 Schamberg disease with reddish-brown areas with petechiae

pigmentation, within which punctate petechiae ("cayenne pepper spots") are visible (Fig. 31.1).

Majocchi disease (purpura annularis telangiectodes) occurs in adolescent patients and is characterized by petechiae, purpura and telangiectases in an annular pattern (Fig. 31.2).

Lichen aureus is a subtype of pigmented purpura with few and localized lesions. The pigmentation typically has an orange-yellow hue.

Less common variants may have a lichenoid morphology or overlying scale and be associated with pruritus. Rarely, mycosis fungoides can present with or mimic a pigmented purpuric dermatosis.

Pigmented purpura primarily affects the lower extremities. It occasionally affects the upper limbs and may rarely be generalized. It is usually asymptomatic.

Laboratory

Histopathology typically shows a perivascular lymphocytic infiltrate with extravasation of red blood cells and hemosiderin deposition in the dermis.

The platelet count and coagulation studies are normal. Laboratory markers of inflammation are not elevated.

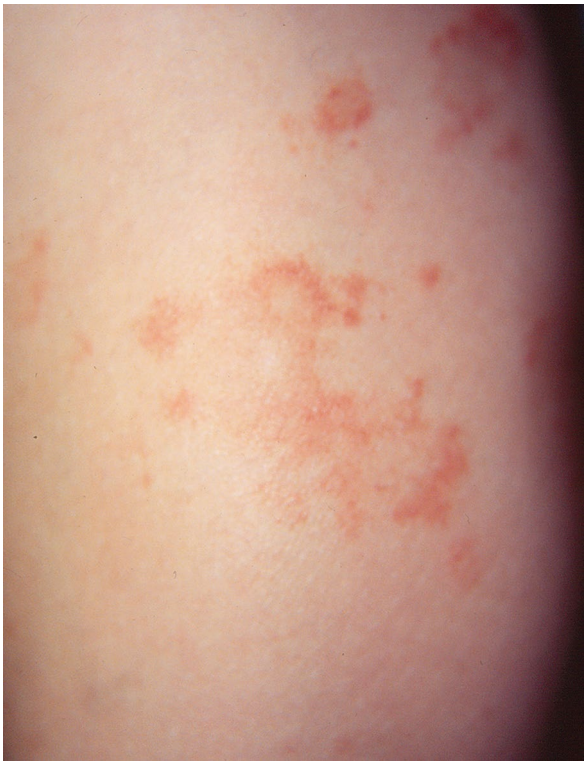

Fig. 31.2 Purpura annularis telangiectodes

Treatment

Treatment is challenging and not necessary for asymptomatic patients unless there is concern about cosmesis. Topical steroids and calcineurin inhibitors are often prescribed. Narrow band UVB phototherapy can be helpful in some cases.

Prognosis

The course is variable. Lesions may persist or recur from several months to many years.

Recommended Readings

1. Kim DH, et al. Characteristics and clinical manifestations of pigmented purpuric dermatosis. Ann Dermatol. 2015;27:404–10.
2. Coulombe J, et al. Pigmented purpuric dermatosis: clinicopathologic characterization in a pediatric series. Pediatr Dermatol. 2015;32:358–62.

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