Arcuate, annular, and polycyclic inflammatory and infectious lesions

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Abstract

Common shapes encountered in dermatologic diseases include linear, nummular, annular, polycyclic, and arciform. The last three have a relatively restricted differential, which must be entirely explored. It is not uncommon for a single disease to present in annular, arciform or polycyclic configurations; moreover, the lesions may evolve from being arciform to annular and then become polycyclic. Regardless, recognizing the arrangement of the defect will undoubtedly help in making a diagnosis and guiding subsequent management. We explore diseases that often present in annular, arciform, and/or polycyclic forms.

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Introduction

In describing a cutaneous disease, it is important to note the distribution, morphology, and arrangement of the lesion(s). Each of these parameters provides possible differential diagnoses and, taken together, helps include or exclude certain dermatoses. Many skin conditions have a characteristic pattern or shape. The configuration of the lesions thus provides information regarding the possible etiology as well as the involved structures.

Common shapes encountered in dermatologic diseases include linear, nummular, annular, polycyclic, and arciform. The last three have a relatively restricted differential, which must be entirely explored. At the outset, however, it must be noted that diseases that are not ordinarily included within these differential diagnoses may occasionally show these shapes. For example, linear immunoglobulin (Ig) A disease may occasionally show annular or arciform lesions. Although dermatoses with an annular outline are more common than polycyclic or arcuate forms, there is considerable overlap in disease presentation. It is not uncommon for a single disease to present in annular, arciform or polycyclic configurations. The lesions may evolve from being arciform to annular and then become polycyclic. Recognizing the arrangement of the defect will undoubtedly help in making a diagnosis and guiding subsequent management. The following sections explore diseases that often present in annular, arciform, and/or polycyclic forms.

Proposed mechanisms of annular, arciform, and polycyclic lesions

We propose the following, distinct mechanisms as responsible for such shaped lesions:

In the first, the lesions simply form at a site and then spread radially (Figure 1). Sarcoidosis is an example of such spread.
In the second, a disease process spreads along a plane in the skin. Figure 2 shows a process spreading in this manner. An example is tinea, which spreads in the uppermost layer of the skin, the stratum corneum.

In the third, the process extends linearly along blood vessels in the superficial dermis, as depicted in Figure 3.

As the inflammatory process continues, it compromises the normal vascular tissue in front of it, causing the inflammation to spread. In the meantime, the later stages of the inflammation are limiting the process in the opposite direction along the vessel as the front of the inflammation has passed it. Figure 3 shows a theoretical graph of an inflammatory parameter, such as vascular permeability, as the inflammation spreads along the vessel. The entire process is analogous to extension of a nerve impulse along an unmyelinated nerve. Because the vessels are arrayed in a grid-like network, the clinical appearance is a figurate spread, as shown in Figure 4.

If the inflammation extends into the superficial layers of the epidermis, the cells of the epidermis may show changes as they progress to the surface as the cells of the basal layer continue to be involved in the radially extending process, producing a following scale, as shown in Figure 5. This pattern is characteristic of the superficial figurate erythemas, including erythema gyratum repens and some cases of erythema annulare centrifugum (See below).

Alternatively, the process may spread along vessels in both the superficial and deep dermis, again producing a figurate lesion, as shown in Figure 6, but typically without a following scale. The superficial and deep forms of figurate erythemas, which include erythema chronicum migrans and the remaining cases of erythema annulare centrifugum, are examples of the latter pattern. The superficial forms tend to produce complex figures, whereas the superficial and deep forms produce simpler, more regular lesions.

Figurate erythemas (persistent figurate erythemas)

The figurate erythemas comprise the defining members of this group of diseases. They include erythema annulare
centrifugum (EAC) and erythema chronicum migrans (ECM; erythema migrans), which are classified subsequently under annular lesions, and erythema gyratum repens (EGR), which is classified subsequently under polycyclic lesions. The two classifications, annular and polycyclic, may overlap. As noted, figurate erythemas are of two types, superficial and deep, showing different pathologic presentations.

The superficial group, which includes EGR and some cases of EAC, tends to show complex or polycyclic patterns clinically. The lesions are relatively superficial and are characterized by a “following scale” (i.e., a scale on the inner aspect of the figures formed). Histologically, one sees a small focus of spongiosic edema overlain by parakeratosis corresponding to the following scale and a loose, but well-defined cufflike lymphocytic infiltrate around superficial blood vessels only (Figures 7-9). This entity (superficial EAC) was described and illustrated in Robert Willan’s classic textbook (Figure 10).

The deep group, which includes ECM and some cases of EAC, tends to show simple, annular patterns clinically. There is no following scale, and the lesions are firm and easily palpated. Histologically, these lesions show no spongiosis or parakeratosis and a dense cufflike infiltrate of lymphocytes around blood vessels in both the superficial and deep dermis (Figures 11 and 12). These characteristics were originally delineated by Darier (see the subsequent discussions).

**Annular**

Annular lesions, although common, can be misleading. These lesions often appear as circular or ovoid macules or patches with an erythematous periphery and central clearing. Some annular shapes result from the centrifugal extension of an infection, whereas others arise from the spreading of a
neoplastic or inflammatory process.\textsuperscript{1} Dermatophytoses are the most common causes of these ringed lesions in adults, but other more serious causes, such as sarcoidosis, may need to be excluded.

\textbf{Tinea corporis}

Tinea corporis is a dermatophytosis of the skin. It is commonly known as “ringworm,” because it was once thought to be caused by invasive worms. \textit{Trichophyton}, \textit{Microsporum}, and \textit{Epidermophyton} species are the most common causes of tinea corporis.\textsuperscript{2} The fungus spreads by close contact with an infected person, dog, other animal, or soil. Climate and hygiene are major determinants of infection. Warm, moist environments are conducive to fungal growth; thus, public bathing facilities are common places of infection.

Dermatophytes mostly infect nonliving, cornified layers of skin. Although the depth of infection is often limited to the epidermis, some fungi may release keratinases to invade deeper into the stratum corneum.\textsuperscript{3} This is rare due to host defense mechanisms. After a 1- to 3-week incubation period, dermatophytes invade peripherally in a centrifugal pattern.\textsuperscript{3} In response to the infection, the active borders have increased epidermal proliferation, resulting in a raised border and scaling. Patients often present with well-demarcated, annular, erythematous papules or plaques, which may have enlarged over time.

A diagnosis is made with microscopic examination of potassium hydroxide (KOH) prepared slides. The sample
should be obtained from the active border of the lesion because this region provides the highest yield. Branched or septate hyphae, or both, are often visualized. Cultures may also be done if the KOH examination is negative, but clinical suspicion is high; they, however, take 2 to 4 weeks to obtain adequate growth. Treatment consists of topical and systemic antifungal agents. The former is the first-line therapy, except for cases of resistant infection, disabling disease, chronic infection, immunosuppression, or infection of nails, palms or soles.

**Pityriasis rosea**

Pityriasis rosea is a self-limited papulosquamous eruption. It usually affects people between ages 10 and 35 years, but can occur at any age. Women are more likely affected. The eruption appears more during the spring and autumn seasons. Though there is no confirmed etiologic agent, pityriasis rosea may be a viral exanthem.

The initial presenting lesion, the “herald patch,” may appear as an annular lesion with an erythematous, raised border, scale, and central clearing. The herald patch is often found on the trunk and is usually between 2 and 10 cm in diameter. The appearance of the herald patch may be preceded by headaches, arthralgias, chills, vomiting, diarrhea, or malaise. Within 2 weeks, eruption of small, pink, oval macules, exhibiting peripheral scaly collarettes appear. These lesions are dominantly found on the neck, trunk, and proximal extremities and may follow dermatomes, giving rise to the pattern of a Christmas tree.

The diagnosis of pityriasis rosea is often clinical. The condition is self-limiting and does not require treatment. Oral antihistamines and topical corticosteroids may provide relief for associated pruritus, and phototherapy helped in severe cases. The lesions, however, mostly resolve on their own in 6 to 8 weeks and recurrence is rare.

The lesions may mimic secondary syphilis. If no unequivocal record or history of a herald patch is obtained, appropriate serologic or other testing must be done to rule out syphilis.

**Granuloma annulare**

Granuloma annulare is a benign, self-limiting skin condition. It is characterized by smooth, skin-colored annular plaques and papules. The cause of this rash is unknown, but dysfunction of the immune system is thought to play a role. Immune complex vasculitis and an abnormality of tissue monocytes are proposed pathogenic mechanisms.

The lesions may be pearly white, skin-colored, red, or purple. They are often found on the hands, feet, wrists, and ankles. Plantar surfaces are usually spared. The rash begins as a round, firm, smooth bump and then becomes a circular ring with a central clearing. There is no scaling or associated vesicles or pustules.

Granuloma annulare is a clinical diagnosis. Biopsy may be taken, but other laboratory tests are of little benefit. In patients with generalized disease, glucose intolerance is common. Spontaneous resolution is common in localized disease. As this condition is mostly asymptomatic, treatment is unnecessary. If there are cosmetic concerns, then intralesional corticosteroid therapy, ultraviolet light therapy, or electrodesiccation may be used.

**Sarcoidosis**

Sarcoidosis is characterized as an idiopathic, multisystem disease with noncaseating epithelioid granulomas. Though the disease is commonly associated with granuloma formation in the lungs, other manifestations involve the cutaneous, ocular, hepatic cardiac, nervous, musculoskeletal, renal, and endocrine systems. In the United States, the prevalence of sarcoidosis is 1 to 40 cases/100,000, and the incidence of this disease is higher in African Americans than in whites. Sarcoidosis has a bimodal age distribution, with peaks between ages 25 to 35 and 45 to 65 years.

Typical skin changes in sarcoidosis include infiltrated papules and plaques. Erythema nodosum, which presents as tender, erythematous, primarily subcutaneous nodules on the extremities, is a common cutaneous presentation of sarcoidosis. It is a secondary, reactive process. Subcutaneous nodules and infiltration of scar tissue may also be present. Reddish to violaceous, indurated plaques may appear on the face and mucous membranes. These lesions may measure between 1 and 3 cm. Such lesions can coalesce, appearing annular and even polycyclic in form.

Clinical presentation, histology, and radiology help establish the diagnosis of sarcoidosis. If sarcoidosis is suspected, then the search for cutaneous lesions is essential. Biopsy of a cutaneous lesion may eliminate the need for a more extensive workup. Treatment of sarcoidosis is with systemic corticosteroids for symptomatic relief. Topical or intralesional corticosteroids may help resolve skin manifestations. Histologically, one sees “naked” granulomas, consisting of masses of histiocytes lacking a surrounding rim of lymphocytes. Often, however, some of the granulomas have prominent rims of lymphocytes.

**Leprosy**

Leprosy, also known as Hansen disease, is a chronic infection caused by *Mycobacterium leprae*. The disease is rare in the United States, with approximately 150 cases diagnosed each year. There are two main forms: tuberculoid and lepromatous. The tuberculoid form arises from a vigorous cellular immune response against *M leprae*. This results in few skin lesions and limited peripheral nerve involvement. Individuals with lepromatous disease, on the other hand, have a minimal cellular immune response, leading to extensive skin and nerve involvement.
Because *M. leprae* grows best at 96°F, the bacteria preferentially colonizes the cooler parts of the body. Thus, the scalp, groin, and axillae are usually spared. In tuberculoid leprosy, presentation often includes erythematous, demarcated macules or plaques. Scaling, alopecia, and anesthesia may also be present. In lepromatous disease, there is a heavy bacterial load. This results in extensive macules and papules that may coalesce into annular lesions.

Diagnosis of Hansen disease is often made on clinical grounds with biopsy for confirmation. When leprosy is suspected, an acid-fast stain should be ordered to identify the bacillus. Treatment involves a multidrug regimen, usually consisting of dapsone and rifampin. In tuberculoid leprosy, treatment may last for 3 to 5 years. In lepromatous disease, treatment may be for life.

**Erythema annulare centrifugum**

Erythema annulare centrifugum (EAC) was first described by Darier in 1916 as a primarily nonscaling, annular, erythematous eruption. EAC affects the trunk, buttock, thighs, and legs. The hands, feet, and face are spared. The etiology of this disease is unknown, but EAC may be a manifestation of an underlying infection or malignancy. Darier described two distinct forms, a superficial form, in which a following scale is seen histologically and clinically (Figures 7-9) and a deep form, with firmer lesions and no scale (Figure 11). The former is associated histologically with a loose, edematous cuffing infiltrate of lymphocytes around superficial blood vessels (Figure 9), the latter with a dense, well-demarcated lymphocytic infiltrate around superficial as well as deep dermal blood vessels (Figure 12). The mechanisms by which both these forms give rise to annular lesions were discussed in previous sections.

Diagnosis is mostly made on clinical grounds. The disease course is variable. It may last for several decades; most cases, however, resolve in 9 months. Treatment of underlying conditions hastens disease resolution. Corticosteroids or antihistamine may improve related pruritus.

**Erythema chronicum migrans (erythema migrans)**

Lyme disease is a systemic infection caused by *Borrelia burgdorferi*. The spirochete is spread by ticks of the genus *Ixodes*. From 1992 to 2006, approximately 250,000 cases of Lyme disease were reported to the Centers for Disease Control and Prevention. The incidence of disease is relatively high in Connecticut and Massachusetts; whereas there are few cases in states like Colorado and Montana. This disparity is due to the prevalence of vectors for disease, such as mice and deer that host the ticks. Mice are much more important hosts. The manifestation of disease may be caused directly by the spirochete or by immunopathogenetic mechanisms.

The classical skin manifestation of Lyme disease is erythema migrans (erythema chronicum migrans, Figure 13). This lesion begins as an erythematous macule or papule at the site of the tick bite. The eruption expands centrifugally over days to weeks, growing a few centimeters a day. At presentation, some lesions can be up to 70 cm in diameter, but most are smaller than 20 cm. The defects are red and have a classical central clearing, leading to the typical bulls-eye or target pattern. Fatigue, headache, myalgias, arthralgias, and low grade fever are sometimes present in early disease.

Diagnosis of Lyme disease is made on clinical grounds, especially in the presence of erythema migrans. Skin biopsy, culture or polymerase chain reaction (PCR) of the rash may obtain additional diagnostic information, especially in complicated cases. Histologically, a superficial and deep infiltrate is seen, resembling the deep form of erythema annulare centrifugum. No scale is seen.

With erythema migrans, empiric antibiotic therapy, consisting of amoxicillin or doxycycline, is reasonable. Cutaneous manifestations of Lyme disease respond promptly to proper antibiotic therapy. The duration of recommended therapy for solitary erythema migrans is 10 to 30 days.

**Polycyclic**

Polycyclic lesions present as configurations arranged in more than one ring. Some annular lesions may coalesce into a polycyclic shape; alternatively, the appearance of such lesions may be independent. For example, the annular lesions of tinea corporis can combine to create a polycyclic lesion. In the case of urticaria, the wheals may present as either annular or polycyclic. When there is a new onset polycyclic rash, it is important to investigate possible underlying malignancy, especially in the presence of erythema gyratum repens.

**Urticaria**

Urticaria is characterized by pruritic, evanescent wheals with erythematous raised borders and blanched centers. The
wheals can assume papular, plaquelike, annular, or polycyclic forms. In acute forms of the disease, prompt medical attention is advised. The course of acute urticaria may last from a few hours to 6 weeks. Urticaria that is present for more than 6 weeks is classified as chronic urticaria. Urticaria is a relatively common phenomenon, affecting between 15% and 20% of the population at some point in their lives.21

The appearance of urticaria is due to the release of histamine and other chemical mediators from mast cells and basophils. The activation of mast cells is caused by the binding of an antigen-immunoglobulin E complex to cells’ FeER1 receptor (type I hypersensitivity).22 Pollens, foods, parasitic infection, fungi, and medications are common instigators of this allergic form of urticaria. In transfusion reactions, urticaria results from an antibody-dependent, cell-mediated cytotoxicity (type II hypersensitivity). In serum sickness, urticaria arises from the formation of antigen-antibody complexes and the subsequent activation of the complement system (type III hypersensitivity). Urticaria may also result from physical, thermal, cholinergic, or solar stresses.22

Urticaria is diagnosed by history and physical examination. The cutaneous findings of urticaria may be confused with those of erythema multiforme. In the latter, however, the lesions are stationary and progress to a dusky color with bulla formation. Early histologic findings in urticaria may demonstrate intravascular margination of neutrophils, which later show diapedesis of neutrophils. Increased eosinophils are often present. Because collagen fibers, present in the dermis, contract during formalin fixation, edema in the dermis is often difficult to appreciate histologically.

Urticaria often spontaneously resolves, but symptomatic relief should be provided. Therapy consists of H1 antihistamines and other medications that block histamine action.22 If possible, the inciting etiologic agent should be identified and the patient educated to avoid it.

Erythema gyratum repens

Erythema gyratum repens (EGR) is a rare figurate erythema with only a handful of cases reported. It is characterized by concentric, mildly scaling erythematous bands, which gives it a polycyclic, wood-grain appearance.23 The rash is often found on the trunk and extremities and is accompanied by intense pruritus.

EGR is thought to be a paraneoplastic condition and is associated with malignancy in approximately 80% of patients.24 This condition is associated most with lung cancer, followed by breast, bladder, uterus, gastrointestinal tract, and prostate cancers.25 Detection of the tumor can occur at the time the EGR appears or up to 6 years after. There are three hypotheses for the pathogenesis of EGR:

1. The tumor antigens cross-react with skin antigens.
2. Tumor products alter skin antigens, making the latter susceptible to autoimmune reaction.

3. Tumor antigens form immune complexes which are deposited into the skin.

EGR can also occur in tuberculosis, lupus erythematosus, CREST syndrome (calcinosis, Raynaud syndrome, esophageal dysmotility, sclerodactyly, telangiectasia), psoriasis, and pityriasis rubra pilaris.

EGR usually mirrors the course of the underlying illness. Resolution of the underlying malignancy is accompanied by the clearance of the lesions and relief of the pruritus. Histologic examination of EGR demonstrates mild spongiosis, focal parakeratosis, and a superficial perivascular lymphohistiocytic infiltrate, resembling the superficial form of EAC. Patients presenting with EGR should be evaluated for the detection of a clinically relevant malignancy.26 To relieve the intense pruritus, corticosteroid injection may be beneficial; the steroids, however, do not resolve the cutaneous findings.

Necrolytic migratory erythema

Necrolytic migratory erythema is strongly associated with glucagonoma syndrome. The rash is characterized as an intensely pruritic eruption of erythematous patches that become superficial vesicles and bullae. Healing begins after central crust formation and the edges progress to form a well-demarcated polycyclic pattern. The eruption may be localized or generalized. The rash may develop on the abdomen, on the groins, circumorally, and in intertriginous regions. Glucagonoma syndrome peaks in incidence in patients aged 45 to 65 years and is more common in women.27 The pathogenesis of this necrolytic migratory erythema is poorly understood aside from its association with elevated glucagon levels.

Patients presenting with necrolytic migratory erythema should be evaluated for an underlying tumor of the glucagon-secreting cells in the pancreas. Surgical cure of the tumor leads to resolution of the cutaneous lesions; however, 60% to 80% of glucagonomas producing the syndrome are malignant at the time of diagnosis.27 Liver metastases are common, making surgical resection difficult and prompting the use of chemotherapy. Isolated treatment of necrolytic migratory erythema is difficult. Topical and systemic steroids, ultraviolet light therapy, dapsone, and tar preparations have been used with limited success.27 Intravenous infusion of octreotide has been found to help in some patients, alleviating the cutaneous eruption by reducing the formation of glucagon. Most patients live for 2 to 10 years with this slow-growing cancer, with the majority living more than 5 years.27

Subacute cutaneous lupus erythematosus

Subacute cutaneous lupus erythematosus (SCLE) is a distinct dermatosis in the spectrum of lupus erythematosus skin disease. SCLE can present as a polycyclic, annular, or papulosquamous rash.28 This skin condition may be drug-
induced, but can also occur in patients with systemic lupus erythematosus (SLE), Sjögren syndrome, or deficiency of the second component of the complement immune system. Most patients with SCLE fulfill four or more of the American Rheumatism Association’s criteria for SLE. The primary lesion is an erythematous papule or a small plaque with scaling. These lesions may expand and merge, forming a polycyclic or annular shape. The typical figurate eruption of SCLE has a photodistribution and may be accompanied with arthritis, pleuritis, or pericarditis.

Serologic testing is positive for the Sjögren syndrome antigen/Ro antibody in 90% of patients with annular or polycyclic SCLE. The histologic examination may show vacuolar alteration of the basal layer and perivascular, perappendiceal, and/or subepidermal lymphocytic infiltrate. Therapy for SCLE includes sun-protection, topical corticosteroids, intralesional corticosteroids, and antimalarials. Antimalarial therapy is found to be quite effective and is associated with lower rates of disease progression and thrombovascular disease.

Erysipelas

Erysipelas is a bacterial infection, mostly by Streptococci, that extends into the skin’s lymphatics. Trauma to the skin allows for bacterial inoculation, leading to the development of erysipelas (Figure 14). Infection may be promoted by lymphedema, venous insufficiency, inflammatory conditions, dermatophyte infection, and stasis ulcerations. Infection may be preceded by malaise, chills, and fever. Diagnosis is made from clinical findings and laboratory examinations are unnecessary. Treatment with penicillin is sufficient for most cases of erysipelas, but the drug regimen may need to be changed due to bacterial resistance or the patient’s allergies. Hospitalization is required for infants, elderly, and immunocompromised patients as well as in severe cases.

Mycosis fungoides

Mycosis fungoides is the most common type of T-cell lymphoma. Well known as a dermatologic masquerader, this malignancy can mimic more than 50 different clinical entities. The neoplastic T cells invade the skin, producing patches, plaques, tumors, or erythroderma. In early stages of the disease, the lesions mostly present as erythematous macules or papules, which may resemble eczema with defined borders. As the infiltrate grows, scaling may appear on the borders. Mycosis fungoides may take on an arciform, annular, or polycyclic form. At times, more than one configuration can be found on a single patient. The involved skin surface may be atrophic and have orange, red, livid, or brown components.

Diagnosis of mycosis fungoides is formulated on clinical, histopathologic, and genetic evidence. With histologic examination, mycoses fungoides lesions contain large atypical lymphocytes and a lymphocytic infiltrate in the papillary dermis. It is difficult, however, to delineate early disease from inflammatory conditions. There are a variety of treatments for mycosis fungoides, and selection depends

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**Fig. 14** Erysipelas after placement of an intravenous device.

**Fig. 15** Erysipelas of the face after a dental procedure.
on the stage and therapy history. In general, stage I patients receive topical therapies, whereas those in stage IIB or higher receive systemic therapies. Mycoses fungoides is an incurable condition in most patients, with the exception of stage 1A disease. In patients with stage IIB disease, survival after diagnosis is close to 3 years in patients with stage IIB disease with a median of 4 to 6 years after diagnosis of stage III disease. In stage IV disease, survival is usually less than 1.5 years. In early-stage disease, the goal of treatment is to slow the progression of the disorder so that the patient dies of an unrelated disease long before the lymphoma becomes a significant health hazard. We advise patients that our goal is for them to die of the disease when they are well over 100 years old.

**Arciform**

Arciform refers to an arc-like configuration. Often, the arcuate shape arises from the clearing of part of an annular lesion. Erythema multiforme may present as or evolve into an arciform rash. Other diseases with an arcuate appearance are not as common, especially in developed countries. With early diagnosis and treatment, erythema marginatum and secondary syphilis have markedly decreased in incidence.

**Erythema multiforme**

Erythema multiforme (EM) is an acute, self-limited hypersensitivity reaction to infections, drugs, and other triggers. EM often appears as a target lesion (Figure 16), which then may evolve into arcuate forms. EM minor is a localized skin eruption with minimal mucosal involvement. EM major and Steven-Johnson syndrome (SJS) are more severe conditions and can lead to death. The definitions and basis for delineation of EM minor and EM major/SJS are controversial, but there seems to be a consensus that EM minor and EM major/SJS are two separate disorders based on their different cutaneous patterns.

The characteristic target lesion may arise because a circulating toxin gains access to the epidermis, causing epidermal necrosis, seen clinically as a gray spot a few millimeters in diameter. This then causes additional blood flow to the skin, seen as a red spot that displaces the gray area outward. This then introduces more toxin to the epidermis, causing an additional wave of epidermal necrosis, seen as a new central gray spot displacing the former lesions outward, followed again by additional blood flow, seen as a new red center. This process repeats itself, giving rise to the target lesion.

Herpes simplex virus is the most commonly associated etiologic agent for EM. Recurrent EM is sometimes associated with reactivation of the herpes simplex virus. *Mycoplasma pneumonia*, fungal infections, barbiturates, hydantoins, nonsteroidal anti-inflammatory drugs, penicillins, phenothiazines, and sulfonamides are other known triggers. Although its pathophysiology is not completely understood, EM associated with herpes simplex virus (always always EM minor) appears to be a type IV hypersensitivity reaction. Lesions in drug-associated EM (usually EM major) are positive for tumor necrosis factor-α, suggesting a different mechanism for disease.

EM may begin within 3 days of prodromal symptoms. In EM minor, an upper respiratory tract involvement is common. In EM major, fever, sore throat, vomiting, and diarrhea are common prodromal findings. The skin lesions usually appear on the distal extremities and progress proximally. Lesions on the dorsal hands and extensor aspects of the extremities are characteristic. Initial lesions consist of red macules that expand over 48 hours to a maximal diameter of 2 cm. A small papule, vesicle, or bulla develops in the centers and then flattens and may clear. The periphery becomes cyanotic, forming the characteristic target lesion. Depending on the healing of the lesions, they then may appear to be arcuate or polycyclic in form.

EM is diagnosed clinically. A skin biopsy specimen, which may have nonspecific findings, is not always necessary when the clinical picture is clear. Usually, however, skin biopsy is diagnostic, showing necrotic keratinocytes. When the diagnosis is unclear, skin biopsy may also help rule out other diseases. Laboratory testing for herpes simplex virus may confirm suspected infection. Treatment of an underlying infection or discontinuing possible instigating drugs may be necessary. Therapy is otherwise symptomatic with antihistamines, analgesics, and mouthwash. Topical corticosteroids may be used, but the use of systemic corticosteroids is controversial. Skin care is necessary to prevent infection of the lesions and surrounding areas.

In EM minor, lesions subside within 3 weeks, usually without consequence, whereas in EM major the lesions subside within 6 weeks and the mortality rate can reach 50%. EM due to a drug reaction may continue to worsen for a week or more after the drug has been withdrawn.
Palpable migratory arciform erythema

Palpable migratory arciform erythema (PMAE) is a rare disease with few report. It is controversial whether PMAE is a pseudolymphoma or an atypical manifestation of lymphocytic infiltration of the skin. PMAE is described to a have characteristic blue-violet nodules with raised, sharp arcuate borders. The lesions have a predilection to present on the trunk; there is no facial involvement. The clinical manifestation changes within days to weeks due to the migratory nature of the disease.

Histologically, PMAE has a dense perivascular and periadnexal inflammatory infiltrate with no mucin or plasma cells. Because the disease does not have a definitive cause, treatment is difficult. Antibiotics have helped in relieving skin lesions. Topical steroids have also been tried, but with variable success. It necessary to exclude other cutaneous malignancy, especially lymphoma, before a diagnosis of PMAE is made.22

Erythema marginatum

Erythema marginatum is one of the major diagnostic criteria for rheumatic fever. This arciform rash occurs in less than 10% of acute rheumatic fever patients; more so in children than in adults. Given the decrease in incidence of rheumatic fever, erythema marginatum has become less common. The mechanisms producing the rash are unknown, but it is thought that there is an abnormal immunologic response to group A β-hemolytic streptococci.43

Erythema marginatum presents with erythematous macules, which spread peripherally becoming patches or plaques. The lesions migrate as quickly as 1 mm/hr for the first 12 hours and may be arciform or polycyclic in arrangement. There is a predilection for the rash to appear on the trunk, axillae, and proximal extremities. The lesions appear predominantly during the active phase of rheumatic fever, preceding arthritic symptoms of rheumatic fever.

Erythema marginatum usually persists for a few hours to a couple of days. It may recur if the rheumatic fever persists. Diagnosis is clinical and is aided by other manifestations of rheumatic fever. Biopsy is not necessary, but the specimen reveals a neutrophilic-dominant interstitial and perivascular infiltrate. Treatment is nonspecific, and lesions usually resolve spontaneously. Efforts should be made to treat the underlying rheumatic fever and minimize heart valve damage.

Secondary syphilis

In secondary relapsing syphilis, the lesions tend to be arciform and asymmetric. The secondary stage of disease results from dissemination of treponemes through the blood vessels and lymphatic circulation.44 The most common clinical presentation of the secondary stage of syphilis is a generalized, nonpruritic papulosquamous eruption. The arciform lesions may be accompanied by moth-eaten scalp alopecia in the occipital area. Lesions may also be present on the mucosal surfaces of the mouth, throat, and cervix. Secondary syphilis may have concomitant lymphadenopathy and, at times, splenomegaly.

The diagnosis of secondary syphilis is best made with darkfield microscopy, where available, and a reactive serologic test for syphilis. A skin biopsy specimen may be diagnostic, but more often is suggestive. Failure to find Treponema pallidum in cutaneous or mucosal lesions does not rule out disease. Local antiseptics, soaps, and drying of lesions may result in negative findings. Without therapy, the lesions may spontaneously resolve within a couple of months of their appearance. Once diagnosis is made, the proper state services may need to be notified. Penicillin G remains the treatment of choice; a single intramuscular dose of 2.4 million units is considered sufficient.45

Conclusions

The shape of a cutaneous lesion and the pattern by which surrounding lesions are arranged may provide recognizable clues leading to rapid visual diagnosis. Whether guttate (drop shaped), nummular (coin shaped), polygonal (with several sides), serpiginous (wavy, snakelike), arciform (arclike), annular (ringlike), or polycyclic (merged circles), recognizing the configuration of cutaneous disease provides an initial, usually reliable differential diagnosis; moreover, it alerts the physician to search for possible underlying malignancies (eg, in erythema gyratum repens) or systemic infection (eg, in secondary syphilis). In arciform, annular, or polycyclic lesions, the overlapping disease manifestations must be kept in mind. An annular lesion may heal into an arcuate lesion or individual lesions merge to become polycyclic. By surveying the entire body, the examiner may be able to distinguish pure polycyclic and arciform lesions from annular lesions in the process of healing. By taking a lesion’s shape into consideration and combining it with other aspects of the clinical examination, one can usually effectively include and exclude certain diseases, order suitable diagnostic tests, and pursue a proper course of management.

References


