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Review

Figurate Erythemas

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Abstract

Background: Figurate erythemas are a group of distinct conditions with different underlying causes and clinical presentations. They must be differentiated from a wide variety of dermatological entities including mycoses, urticaria, granuloma annulare, pseudolymphoma. Dermatologists need to be familiar with this set of conditions which include erythema annulare centrifugum, erythema gyratum repens, erythema marginatum, erythema migrans and necrolytic migratory erythema which are all important clues to underlying diseases.

ERYTHEMA ANNULARE CENTRIFUGUM

Erythema annulare centrifugum represents a reaction to a wide variety of triggers [1]. The etiology is unknown in most cases [2].

Triggering Factors in Erythema Annulare Centrifugum [1, 3]:

* ***Infection:*** Chronic dermatophyte infections, intestinal *Candida albicans*, molluscum contagiosum, EBV, genital herpes, Q fever, urinary system infections, tuberculosis, ascariasis ie.
* ***Malignancy:*** Erythema annulare centrifugum can be considered an uncommon but genuine paraneoplastic sign. Bronchial, prostate, nasopharyngeal, ovarian, rectal and hepatic tumors, lymphoma and leukemia are examples.
* ***Food allergies***
* ***Drug reactions:*** Aldactone, amytriptilline, ampicillin, cimetidine, hydrochlorothiazide, penicillin, piroxicam, salicylates, vitamin K ie.
* ***Hematologic conditions:*** Polycythemia vera, myelodysplastic syndrome, hypereosinophilic syndrome, cryoglobulinemia ie.
* ***Endocrinologic conditions:*** Hyperthyroidism, Hashimoto thydroiditis, autoimmune progesterone dermatitis
* ***Other:*** Hepatic disease, after biliary duct surgery

The condition does not affect a particular sex or age group. Erythematous macules or urticarial papules appear first and eventually spread to form annular shapes with central clearing [2]. Vesiculation may be rarely seen [3]. The lesions tend to appear on the body and proximal parts of the extremities. Most of the cases spontaneously recover in weeks [4].

There are two types of erythema annulare centrifugum. It is not known whether these two types are variants of the same pathologic condition [**5**].

In the deep type of erythema annulare centrifugum there is an indurated and apparent border, without desquamation and the lesion rarely itches. In the superficial type, there is desquamation following the advancing border and itching is more frequent [**2**].

Histopathologically; in the deep type: The epidermis is unaffected. There is a "coat sleeve-like" lymphocytic infiltration in the mid and deep dermis. In the superficial type: Epidermal changes such as focal epidermal spongiosis and focal parakeratosis and superficial perivascular lymphohistiocytic infiltrate are present. Endothelial cell edema and erythrocyte extravasation may accompany [**2, 4**]. Eosinophilia may be seen in some cases both histopathologically and in peripheral blood. Sometimes erythema annulare centrifugum may even be an early sign of hypereosinophilia syndrome [**2**].

The lesions of erythema annulare centrifugum may wax and wane and last from months to years. Most cases resolve spontaneously. Topical therapies are usually of no use [**2**]. Antihistamines and/or systemic glucocorticoids can be tried but the lesions recur when treatment is discontinued [**6**]. The empiric use of antibiotics and antifungal agents has been reported to be useful in some cases. The patient may be treated as for chronic urticaria [**2**].

ERYTHEMA GYRATUM REPENS

Erythematous bands spread over the body in waves in this condition [**3**]. These bands have been likened to patterns on wood or the stripes of a zebra [**1**]. There are some clinical differences from erythema annulare centrifugum. These are faster spreading of the lesions (about 1 cm/day) and existence of a more pronounced desquamation and pruritus [**3**]. There is a characteristic collar-like desquamation. The lesions appear on the trunk and extremities [**Figure 1**]. The hands, feet and face are usually not affected [**7**]. Hyperkeratosis of the palms has been reported in about 10% of the patients [**3**].

There is an underlying malignancy in about 80% of the cases of erythema gyratum repens [**3**]. For this reason, the patient must be analyzed carefully for malignancy [**1**].

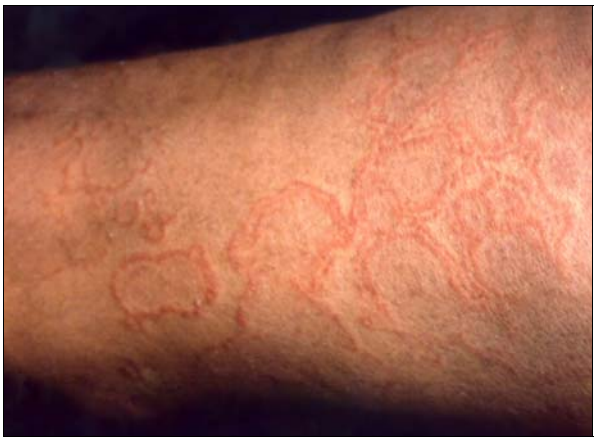

Figure 1. Erythema gyratum repens

The most freqently seen malignancies are lung, breast and esophagus cancers [**4**]. Apart from malignancy, tuberculosis, CREST syndrome, drug hypersensitivity and pregnancy have also been reported. In some cases there may be no underlying cause [**3**].

Although the appearance of the lesions is typical, differential diagnosis from atypical vasculitides, and fungal infections such as tinea imbricata has to be made histopathologically. Lupus erythematosus, pemphigoid, annular psoriasis may also present similarly [**3**]. Histopathologically there is perivascular lymphocytic infiltration resembling erythema gyratum repens but the infiltration is concentrated in the superficial dermis. Additionally epidermal changes are more frequent [**1**]. These changes are acanthosis, spongiosis and parakeratosis [**3**]. Some authors have identified granular C3, C4 or IgG deposition in the sublamina densa region of the basal membrane. This may indicate that the condition has an immunologic basis [**4**].

Treatment of erythema gyratum repens should be directed to the underlying condition. Antihistamins may be used for intense pruritus [**1**].

ERYTHEMA MARGINATUM

(Erythema Circinatum, Erythema Annulare Rheumaticum)

It is a sign seen in about 20% of patients with acute rheumatic fever.[1] It is one of the major Jones' criteria, along with carditis, migratory polyarthritis, chorea and subcutaneous nodules [**3**]. It is thought to be a response to streptococcal antigens [**1**]. It is fre-

quently seen in children[**3**] and patients with active carditis [**1**]. It is more pronounced in fair skinned individuals [**2**]. The trunk is most freqently affected. It usually appears after a fever spike in the afternoon. The lesions are pink, macular or papular and circular. The disappear within hours or in maximum 2-3 days [**1, 3**].

Histopathologically, this condition may be differentiated from other erythematous conditions due to infiltration with polymorphic leukocytes [**3**].

Erythema marginatum has also been reported in conditions such as psittacosis and hereditary angioneurotic edema [**3**].

ERYTHEMA MIGRANS

Erythema migrans is a lesion that forms at the location of the tick bite in Lyme disease [**4**]. Sometimes the patient is not aware of the bite [**2**]. The involved species of Borrelia are *B. burgdorferi sensu lato*, *B. afzelli* or *B. garinii* [**3**].

Classification of Lyme disease [2]:

Early Lyme Borreliosis

Localized infection: Erythema migrans, borrelial lymphocytoma. No signs of disseminated infection. Symptoms such as localized lymphadenopathy and/or malaise may be present.

Early disseminated infection: Multiple erythema migrans-like skin lesions. Neuroborreliosis, arthritis, carditis or other organ symptoms.

Late Lyme Borreliosis

Chronic infection: Acrodermatitis chronica atrophicans. Neurologic, joint or other organ involvement—these should last at least 12 months.

The name erythema “chronicum” migrans is a misnomer [**3**]. Most patients do not have seropositivity against *B. burgdorferi*. Thus, even though the gold standard is culture, diagnosis depends solely on clinical recognition [**8**]. The lesion begins as an erythematous area or red papule 3 to 30 days after the tick bite. It enlarges in a few weeks and the center fades [**Figure 2**]. It reaches a diameter of 25 cm. The duration of the lesion is 4-10 weeks.

On histopathologic examination, perivascular infiltrate containing plasma cells and eosinophils is seen. Spirochetes are seen most frequently in the advancing border of the lesion [**4**].

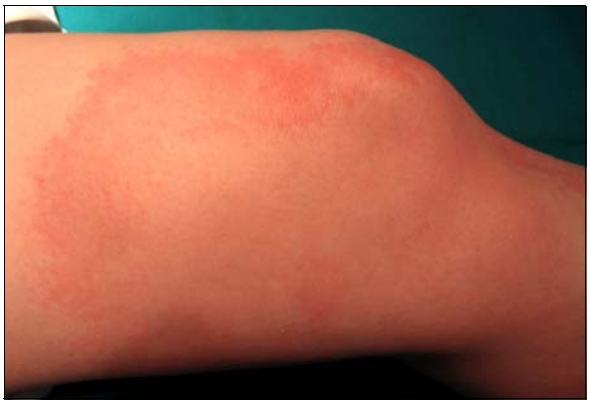

Figure 2. Erythema migrans

Treatment consists of tetracyclin [**5**]. (doxycyclin 2 x 100 mg/day, 2-3 weeks).

ANNULAR ERYTHEMA OF INFANCY

This condition has typically no difference from erythema annulare centrifugum but it is classified as a different condition due to differences in underlying causes. Whereas superficial mycoses and malignancies are rare causes in this age group, lupus erythematosus and infections definitely should be ruled out. No cause can be identified in most cases. *C. albicans* colonization in the intestine and EBV infection have been reported.

Histopathology is the same as erythema annulare centrifugum [**3**].

There are many types of figurate erythemas in infancy. In some cases the lesions are scaly and may resemble T. versicolor. Some types where there is central atrophy are named erythema gyratum repens atrophicans transiens. Due to its histopathological appearance, there is also a condition named neutrophilic figurate erythema of infancy. Treatment consists of a wait and watch policy if no underlying cause is found [**1**].

ERYTHEMA GYRATUM PERSTANS

This condition is named familial annular erythema. The lesions are identical to erythema annulare centrifugum but there is autosomal dominant inheritance. It begins early, sometimes right after birth. Most patients also have dermographism [**1**]. Al-

though the lesions last shorter than erythema annulare centrifugum, the disease itself persists for years [7].

NECROLYTIC MIGRATORY ERYTHEMA

This condition is a paraneoplastic sign seen when there is an underlying glucagon-secreting malignant pancreas alpha-cell tumor. There are rare reports of idiopathic cases or cases due to other gastrointestinal causes (chronic pancreatitis, chronic hepatitis, colon cancer) which is called the pseudoglucagonoma syndrome. Necrolytic migratory erythema is seen frequently in postmenopausal women. Male/femal ratio is 3/1. Glucagon or its metabolites are thought to be responsible. The lesions start as red-brown macules in perioral or inguinal regions and later necrotize and become covered with crusts. Glossitis may accompany. This appearance is similar to *C. albicans* infection. The macules may become vesicular, widespread and may desquamate [1].

Epidermal necrosis, pale basal cells, dyskeratotic cells, acantholysis, subcorneal pustule formation containing neutrophils are seen on histopathology. A perivascular infiltrate composed of lymphocytes and histiocytes are seen in the dermis.

Serum glucagon levels are quite high. Weight loss, malaise, intermittent diarrhea, hypokalemia, resistant diabetes mellitus and anemia are seen. Since symptoms start before the tumor can be identified by radiologic methods, no tumor may be identified.

Zinc deficiency and Hailey-Hailey disease should be included in the differential diagnosis. Pustular psoriasis, subcorneal pustular dermatosis and pemphigus foliaceus should be included as well.

Symptoms reside after resection of the tumor. Recurrence of symptoms is a sensitive indicator of recurrence of the tumor [1].

Keypoints of figurate erythemas are outlined in **Table 1**.

Table 1. Keypoints of Figurate Erythemas [1]

Erythema annulare centrifugum	Slowly advancing lesions, mostly idiopathic
Erythema gyratum repens	Rapidly advancing lesions, mostly indicates malignancy
Erythema migrans	Annular lesions arising at location of tick bite, indicator of Lyme disease
Erythema marginatum	Specific to acute rheumatismal fever, seen right before joint involvement
Necrolytic migratory erythema	Finding of glucagonoma, acral and perioral location
Annular erythema of infancy	It is a group of conditions, underlying causes must be examined
Familial annular erythema	Very rare, autosomal dominant

Diagnostic approach to figurate erythemas [2]

1. Are there any signs or symptoms of malignancy, infection or other systemic disease?
2. Are there other findings of tick bite or Lyme disease?

   There are one or two annular lesions around the tick bite in Lyme disease. Erythema migrans usually transforms into plaque form, which is very rare for erythema annulare centrifugum.

   Lesions are multiple in erythema annulare centrifugum.
3. Are there lesions of urticaria or angioedema? Urticaria lasts shorter and itches more than erythema annulare centrifugum.
4. Are there bullous lesions? Bullous pemphigoid and linear IgA disease also have urticarial phases.
5. Erythema multiforme should be considered if the lesions have an oral and acral distribution.
6. KOH examination should be done.
7. If the lesions are psoriasiform, psoriasis and subacute lupus erythematosus should be considered. Rarely Sjogren syndrome may present with annulare lesions. Ro/La antibodies should be investigated.
8. Are there any other findings of acute rheumatismal fever? Erythema marginatum is the shortest lasting of the figurate erythemas.
9. Are the lesions located orally or in intertriginous locations? Are there any other signs of glucagon excess?

10. Is there family history of similar lesions? Is there anyone in the family with granulomatous disease? Are phagocytic functions normal? Annular lesions may be seen in carrier females with chronic granulomatous disease.

11. Is the patient an infant? Neonatal lupus erythematosus must be ruled out in this age group. Although mycoses are not seen commonly in infants, they should be ruled out.

Differential Diagnosis of Figurate Erythemas [3]

Mycoses, annular urticaria, granuloma annulare, mycosis fungoides, pseudolymphomas (especially erythema arciforme et palpible migrans), bullous pemphigoid, pemphigus, dermatitis herpetiformis, linear IgA disease, erythema multiforme, sarcoidosis, Still disease, annular psoriasis, erythrokeratoderma variabilis, chronic granulomatous disease, pityriasiform seborrheic dermatitis, neutrophilic dermatoses, vasculitides, acute hemorrhagic edema of childhood, lepra, leishmania, trypanosomiasis.

References

1. Burgdorf WHC. Erythema annulare centrifugum and other figurate erythemas. Fitzpatrick's Dermatology in General Medicine. Freedberg IM, Eisen Az, Wolff K et al, 6th edition. USA, McGraw-Hill Companies, Inc, 2003; pp.977-980.
2. Weyers W, Diaz-Cascajo C, Weyers I. Erythema annulare centrifugum: results of a clinicopathologic study of 73 patients. Am J Dermatopathol 2003; 25: 451-462. PMID: 14631185
3. Mobini N, Toussaint S, Kamino H. Noninfectious erythematous, papular and squamous diseases. Lever's Histopathology of the Skin. Elder DE, 9th edition. Philadelphia, Lippincott Williams and Wilkins, 2005; pp.181-183.
4. Braun-Falco O, Plewig G, Wolff HH, Burgdorf WHC. Dermatology. 2nd edition. Berlin, Springer-Verlag, 2000; pp.574-578.
5. Graham RM, Cox NH. Systemic disease and the skin. Rook's Textbook of Dermatology. Burns T, Breathnach S, Cox N, Grifiths C, 7th edition. Massachusetts, Blackwell Publishing Company, 2004; pp.59.70-59.75.
6. Lipsker D, Lieber-Mbomeyo A, Hedelin G. How accurate is a clinical diagnosis of erythema chronicum migrans? Arch Dermatol 2004; 140: 620-621. PMID: 15148115
7. Boyd AS, Neldner KH, Menter A. Erythema gyratum repens: a paraneoplastic eruption. J Am Acad Dermatol 1992: 26: 757-762. PMID: 1583177
8. Kim K, Chang SE, Choi JH et al. Clinicopathologic analysis of 66 cases of erythema annulare centrifugum. J Dermatol 2002; 29: 61-67. PMID: 11890297