Figurative Erythemas

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Figurative Erythemas

- Erythema annulare centrifugum
- Erythema marginatum
- Erythema migrans
- Erythema gyratum repens
- Erythema multiforme
Erythemas

- Erythemas represent a change in the color of the skin that is due to the dilation of blood vessels, especially those in the papillary and reticular dermis.
- The color is blanchable and most last for days to months.
- Figurative erythemas have an annular, arciform or polycyclic appearance.
ERYTHEMA ANNULARE CENTRIFUGUM
ERYTHEMA ANNULARE CENTRIFUGUM

• Pathogenesis: EAC represents a reaction pattern or hypersensitivity to one of many antigens
  – IL-2 and TNF-alpha may have a role
  – Most patients do not have an underlying disease identified
ERYTHEMA ANNULARE CENTRIFUGUM

- Associated with:
  - Infection
    » Dermatophytes and other fungi (*Candida* and *Penicillium* in blue cheese)
    » Viruses: poxvirus, EBV, VZV, HIV
    » Parasites and ectoparasites
  - Drugs: diuretics, antimalarials, gold, NSAIDs, finasteride, amitriptyline, etizolam, Ustekinumab (2012)
ERYTHEMA ANNULARE CENTRIFUGUM

– Foods
– Autoimmune endocrinopathies
– Neoplasms (lymphomas and leukemias)
– Pregnancy
– Hypereosinophilic syndrome
– Lupus (2014)
ERYTHEMA ANNULARE CENTRIFUGUM

http://www.dermaamin.com

Rongioletti, F., Fausti, V., & Parodi, A
ERYTHEMA ANNULARE CENTRIFUGUM

• 2 major forms:
  – Superficial: classic **trailing scale**, may have associated pruritus
  – Deep: **infiltrated borders**, usually no scale, edges are elevated, usually not pruritic
ERYTHEMA ANNULARE CENTRIFUGUM

- Pink papules expand outwards, develop central clearing
- Annular lesions with trailing scale; favor thighs, hips, and trunk
- Desquamation is present at the inner margin = “trailing scale”
  - Lesions persist for weeks to months
  - Rarely involves palms/soles, scalp, mucous membranes
  - As lesions resolved: no scarring.
ERYTHEMA ANNULARE CENTRIFUGUM

• Histology:
  – **Superficial form**: nonspecific, mild spongiosis, microvesiculation, focal parakeratosis, “coat sleeve” superficial perivascular lymphohistiocytic infiltrate
    • Advancing edge is a result of dermal papillary edema
  – **Deep form**: normal epidermis, mononuclear infiltrate with a sharply demarcated perivascular arrangement in the mid and lower dermis
ERYTHEMA ANNULARE CENTRIFUGUM

“coat sleeve” = lymphocytes tightly associated with vessels
ERYTHEMA ANNULARE CENTRIFUGUM

• Treatment:
  – Treat underlying disorder
  – Topical steroids to advancing border, antihistamines, +/- empiric antibiotics and antifungals
  – Systemic treatment is rarely necessary
ERYTHEMA MARGINATUM
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• Introduction:
  – Erythema marginatum is a cutaneous manifestation of acute rheumatic fever
  – Rheumatic fever is characterized by an abnormal immunologic response to a preceding infection with group A β-hemolytic streptococci
  – Triad of fever, arthritis and carditis
ERYTHEMA MARGINATUM

• Epidemiology: 3% of untreated patients develop rheumatic fever, of that 3% the rash is seen in less than 10% of patients

• The peak age-related incidence is between 5 and 15 years
**ERYTHEMA MARGINATUM**

- Major criteria for acute rheumatic fever:
  - Joints (migratory polyarthritis)
  - ♥ (carditis)
  - Nodules (subcutaneous nodules- painless, over bony prominences in long standing disease)
  - Erythema marginatum
  - Sydenham’s chorea

- Minor criteria: fever, arthralgias, elevated ESR, elevated CRP, prolonged PR interval
ERYTHEMA MARGINATUM

- Clinical: migratory annular and polycyclic erythema, 2-5 week latency
  - MC locations: trunk, axillae, proximal extremities
  - New lesions last from a few hours to a few days, most noticeable in the afternoon
  - Lack of scale (helps to r/o EAC and other papulosquamous conditions)
ERYTHEMA MARGINATUM

• Histology: Interstitial and perivascular infiltrate composed mostly of neutrophils w/o vasculitis, extravasated RBCs in later stages, DIF is negative

• Tx: no specific treatment, lesions resolve spontaneously
ERYTHEMA (CHRONICUM) MIGRANS
ERYTHEMA (CHRONICUM) MIGRANS

• **Borrelia burgdorferi** spirochetes by Ixodes tick (may transmit babesiosis, human anaplasmosis)
  – Must be attached >48hrs for transmission

• Seen mostly in US (northeast, midwest, west coast) and Europe

• Natural hosts for Borrelia are white-footed mice and white-tailed deer

• Not all patients who have had tick bites or positive serologic tests for **B. burgdorferi** develop Lyme
ERYTHEMA (CHRONICUM) MIGRANS

• Pathogenesis: *Ixodes* uses tick salivary protein (Salp 15) as a means of enhancing transmission
  – Once in the body it is thought to trigger innate and adaptive immunity
  – 45% of patients with erythema migrans have spirochetemia
  – Spirochetes can be found in the skin for long periods of time after tick bite
ERYTHEMA (CHRONICUM) MIGRANS

- Clinical: erythematous, expanding annular plaque appears on an average of 7-15 days after the tick bite, may have a bull’s eye appearance
  - MC sites for primary erythema migrans is trunk, axilla, groin, popliteal fossa
  - Major organ manifestations of untreated patients: 60% monoarticular or oligoarticular arthritis (usually knees), 10% neurological (MC facial nerve palsy), 5% cardiac (AV block)
ERYTHEMA (CHRONICUM) MIGRANS

• The diagnosis of early Lyme disease can be made solely on clinical grounds when a characteristic erythema migrans lesion is present in patients who live in or have recently traveled to an endemic area.
  – Patients who present with an EM lesion will likely be seronegative, since the lesion often appears prior to the development of a diagnostic immune response. Serologic testing is neither necessary nor recommended in these patients.
ERYTHEMA (CHRONICUM) MIGRANS

http://hdjaguar.mobi/erythema-chronicum-migrans-photo/
ERYTHEMA (CHRONICUM) MIGRANS

• Southern tick-associated rash illness (STARI) has a rash that is indistinguishable from that of Lyme disease
• The etiologic agent is not known
• No serious systemic complications from STARI are currently recognized
• In the Southeast, STARI is much more prevalent than Lyme disease
• TX: Doxycycline
ERYTHEMA (CHRONICUM) MIGRANS

• Histology: Routine histology is nonspecific. Many specimens contain eosinophils and plasma cells; Warthin-Starry stain (silver stain) will occasionally reveal the organism
  – Decreased Langerhans cells in the dermis, multiple apoptotic cells in the epidermis
  – Inflammatory infiltrate contains macrophages, CD4+ helper T cells, CD45+ RO memory T cells
ERYTHEMA (CHRONICUM) MIGRANS

• Tx: only 1% of those bitten get Lyme disease, routine treatment not recommended unless:
  – If in an endemic area (>20% of ticks are infected) and bitten by a tick identified to be Ixodes and attached >36hrs: prophylaxis within 72hrs with single dose of Doxy 200mg
ERYTHEMA GYRATUM REPENS

• Gammel’s disease

• **Paraneoplastic** figurate erythema

• Pathogenesis: immune cross-reaction between tumor antigens and cutaneous antigens
ERYTHEMA GYRATUM REPENS

Figure 1  Conditions associated with erythema gyratum repens (83 cases).

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ERYTHEMA GYRATUM REPENS

Figure 4 Diseases associated with non-paraneoplastic erythema gyratum repens (25 cases).

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ERYTHEMA GYRATUM REPENS

• Figurate erythema that is migratory and composed of concentric rings with a **wood-grain** appearance.

• Lesions develop scale at edges and **advance at a rapid rate** = up to 1cm per day (much faster than EAC)
  – 85% of patients have an **underlying neoplasm**; most commonly **lung**, breast, or esophagus/stomach, may coincide with pulmonary TB
  – Rash develops from 1yr prior to 1yr after the diagnosis of the neoplasm
ERYTHEMA GYRATUM REPENS

“WOOD GRAIN” APPEARANCE

Andrews disease of the skin and Rongioletti, F., Fausti, V., & Parodi, A
ERYTHEMA GYRATUM REPENS

• Histology: Non-specific; hyperkeratosis, focal parakeratosis, moderate patchy spongiosis, mild perivascular lymphohistiocytic infiltrate; eosinophils and melanophages may be seen
  – Accumulation of active Langerhan’s cells in upper layers of epidermis
  – DIF: IgG and C3 in the floor of the blister cavity (only seen in some patients, not required for confirmation)
ERYTHEMA GYRATUM REPENS

- Tx: Resolves when underlying condition is treated.
ERYTHEMA MULTIFORME
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• Classified by etiology
  – Herpes Simplex
  – Mycoplasma
  – Contact Dermatitis
  – Drug induced
  – Radiation induced
  – Idiopathic
ERYTHEMA MULTIFORME

• EM minor
  – Self limited, recurrent
  – “target” lesions= peripheral erythema, edematous pale ring and central dusky purpura

• EM major
  – On a spectrum with SJS and TEN
  – More severe, most likely drug related
  – More mucosal involvement
ERYTHEMA MULTIFORME

Andrews Disease of the skin
ERYTHEMA MULTIFORME

Keratinocyte necrosis
ERYTHEMA MULTIFORME

– Histo: “basket weave” stratum corneum, cellular necrosis out of proportion to lymphocytes

– Tx: prevent HSV outbreaks, acyclovir
Resources

- Batycka-Baran A, Zychowska M, Baran W, Szepietowski JC, Maj J.
- *Erythema Annulare Centrifugum Associated with Ovarian Cancer*. Acta Derm Venereol. 2015 May 27