***no patient handout*

Erythema gyratum repens - Skin

Synopsis

Erythema gyratum repens (Gammel disease) is a paraneoplastic syndrome that is thought to arise as a result of the cross-reactivity of tumor antigens with antigens in the skin. It often presents prior to the detection of the internal malignancy as a pruritic, annular, erythematous eruption whose edges advance at a rapid rate.

Erythema gyratum repens is associated with internal malignancy in 85% of patients, most commonly lung carcinoma. Other cases have been reported with cancers of the esophagus, breast, cervix, stomach, pharynx, bladder, prostate, uterus, tongue, colon, and pancreas. Erythema gyratum repens has also been noted in association with several nonmalignant conditions such as tuberculosis, linear IgA dermatosis, psoriasis, pityriasis rubra pilaris, pemphigus vulgaris, bullous pemphigoid, ichthyosis, and hypereosinophilic syndrome. The disorder has only been reported in whites. The male-to-female ratio is 2:1, and the average age of onset is 63 years.

Codes

ICD10CM:
L53.8 – Other specified erythematous conditions

SNOMEDCT:
77300003 – Erythema gyratum repens

Look For

Serpiginous, erythematous plaques with fine scale trailing the leading edge. The lesions have what has been described as a "wood-grain" pattern due to the development of "rings within rings." The edges of the lesions advance at a rapid rate, up to 1 cm/day.

The hands, feet, and face are typically spared.

Diagnostic Pearls

Additional skin manifestations that may be seen include palmoplantar hyperkeratosis, dystrophic nails, ichthyotic changes, and nonspecific vesicles and bullae.

Peripheral eosinophilia is seen in over one half of patients.

Differential Diagnosis & Pitfalls

The differential includes other figurate erythemas:

- **Erythema annulare centrifugum (EAC)** – Erythema gyratum repens advances at a much faster rate than EAC.

- Erythema migrans (marker of **Lyme disease**)
- Annular **urticaria**
- Urticarial phase of **bullous pemphigoid**
- **Erythema multiforme**
- **Tinea corporis**
- **Erythema marginatum** (rheumatic fever)
- Necrolytic migratory erythema (associated with **glucagonomas**)
- If the rash is psoriasiform, check for other stigmata of **psoriasis**.
- **Subacute cutaneous lupus erythematosus**
- **Granuloma annulare**
- **Secondary syphilis**
- **Leprosy**
- **Sarcoidosis**
- **Tinea imbricata**

Best Tests

Erythema gyratum repens has a distinct and often striking appearance; therefore, the diagnosis is often made clinically.

Skin biopsy findings are nonspecific and include hyperkeratosis, parakeratosis, patchy spongiosis, and perivascular lymphohistiocytic infiltrate.

Management Pearls

Patients will develop erythema gyratum repens an average of 4–9 months prior to diagnosis of their internal malignancy in 80% of cases. Therefore, recognition of this paraneoplastic disorder should prompt an exhaustive search for the underlying malignancy. Patients will often require referral to oncology – medical, surgical, and/or radiation.

Therapy

In paraneoplastic cases, therapy is aimed at treating the underlying malignancy via surgery, chemotherapy, radiation, or a combination of these modalities. The erythema and pruritus usually resolve rapidly when the appropriate treatment is administered. In patients with a large metastatic tumor burden, the eruption usually does not resolve until shortly prior to death.

Erythema gyratum repens has been reported to resolve spontaneously in one case not associated

with malignancy. Systemic corticosteroids have been tried with modest results. Topical steroids, vitamin A, and azathioprine have been tried without success.