

Lupus Erythematosus Tumidus

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epidermal component was noted. Pain and touch sensation were normal.

Differential diagnoses

These include cutaneous lupus erythematosus, erythema annulare centrifugum, leprosy, tinea corporis and erythema chronicum migrans.

CASE SUMMARY

History

A 33-year-old housewife presented with mildly itchy rash over her face and upper trunk for two months. There was photosensitivity. There were no systemic symptoms such as joint pain, weight loss or fever. She was a non-smoker and non-drinker. Her past health was good.

Physical examination

There was a large erythematous annular lesion with oedematous edge over her right upper back (Figure 1). Similar skin lesions were present on the left side of her face, right arm and anterior chest. No significant

Investigations

Her complete blood count, ESR, liver and renal function tests were normal. The ANF titre was 1:120. The anti-DNA, anti-ENA, rheumatoid factor, C3, C4 and immunoglobulin levels were all normal. *Borrelia burgdorferi* antibody was negative. Skin scrapping for fungus was negative.

Diagnostic skin biopsy revealed superficial and deep perivascular as well as periadnexal inflammatory infiltration in the dermis and vacuolar alternation of the basal epidermal cells. There was conspicuous dermal deposition of mucin. No fungus was detected. Direct immunofluorescent was negative.



Figure 1: Erythematous annular lesions with oedematous edge over the upper back and arm

Diagnosis

The diagnosis was lupus erythematosus tumidus.

Management

Sun protection was advised. Topical corticosteroid was prescribed with initial response. Hydroxychloroquine 200 mg QD was given later because of worsening of her skin condition. Good response was seen within two months.

REVIEW ON LUPUS ERYTHEMATOSUS TUMIDUS

Lupus erythematosus tumidus (LET) is a rare subset of chronic cutaneous lupus erythematosus (LE). Gougerot and Bournier first described its hallmark clinical features in 1930¹ and it has been very rarely reported since. In English, "tumid" means swelling (of parts of the body).

Clinical features

LET is characterized by non-scarring, erythematous, succulent, urticaria-like, single or multiple plaques with no surface changes over sun-exposed areas.² It can be mildly pruritic and occasionally presents with fine scaling. The plaques may clear spontaneously without scarring and then recur in the original distribution. Photosensitivity is common in patients with LET. Onset tends to cluster in summer. While patients may have concurrent lesions of discoid LE, LET lesions may be present in patients with systemic LE.³

A slight male predominance (22 males and 18 females) was reported in one study² but other papers showed a female predominance. The mean age at onset was 36.4 years (range 9 months to 54 years).

Investigations

An elevated ANA titre ($\geq 1:160$) was detected in 10% of patients. Anti-Ro and anti-La were positive in 5%. There was a low C3/C4 level in 5%. Raised IgA, IgG and/or IgM levels were found in 8-10%.

Phototesting is an important diagnostic test in LET. Reproduction of skin lesions after UVA and/or UVB irradiation was detected in 70% of patients. Uninvolved

skin on upper back was irradiated with single doses of 100 J/cm² UVA and/or 1.5 MED (Minimal erythema dose) UVB daily for 3 consecutive days. Test areas were evaluated until specific lesions appeared for up to 4 weeks after last irradiation. Criteria for positive result required that induced lesions clinically resembled LE, histology finding compatible with LE, and skin lesions developed slowly and persisted for several days or weeks in contrast to other photodermatoses.

Skin biopsy typically shows perivascular and periadnexal superficial and deep lymphocytic infiltration, interstitial mucin deposition, and in some cases, scattered neutrophils. There is no epidermal involvement or alternation of the dermoepidermal junction. Direct immunofluorescence is negative.

Ackerman considered LET to be an authentic manifestation of discoid LE, but devoid of changes at the dermoepidermal junction and in the epidermis and papillary dermis.⁴

Differential diagnoses

LET shows different clinical and histological features as compared with subacute cutaneous LE and erythema annulare centrifugum. In contrast to polymorphic light eruption, LET shows a much more delayed reaction after sun exposure and takes longer to heal. It may be indistinguishable from Jessner's lymphocytic infiltration. In fact, some wonder whether LET and Jessner's lymphocytic infiltration may be the same disease, with the latter representing an earlier form of the disease. LET can be distinguished from pseudo-lymphoma histologically as the latter typically shows top-heavy, wedge-shaped infiltrate composed mostly of small lymphocytes and plasma cells or eosinophils with no mucin deposition. Reticular erythematous mucinosis (REM) usually presents as reticulated, erythematous patches with well-defined margins, typically on the chest and upper back. Some authors consider REM to be a variant of DLE or LET.

Treatments

As with all photodermatoses, sun-protection is important. Topical corticosteroids together with sun-protection may induce resolution of skin lesions in 45% of patients. LET responds rapidly to anti-malarial drugs (both hydroxychloroquine and chloroquine phosphate).

It has been reported to induced complete resolution in 20 of 22 treated patients (91%).²

Systemic corticosteroid or immunosuppressants may be required in a minority of patients. Spontaneous resolution may also occur.

Learning points:

Diagnosis of LET is based on clinico-pathological correlation, results of phototesting and its rapid response to anti-malarials.

References

1. Gougerot MH, Bournier R. Lupus erythematoses tumidus. Bull Soc Fr Derm Syph 1930;1291-2.
2. Kuhn A, Richter-Hintz D, Oslislo C, et al. Lupus erythematosus tumidus – a neglected subset of cutaneous lupus erythematosus: report of 40 cases. Arch Dermatol 2000;136:1033-41.
3. Ruiz H, Sanchez JL. Tumid lupus erythematosus. Am J Dermatopathol 1999;21:356-60.
4. Ackerman AB. Histologic diagnosis of inflammatory skin diseases. Baltimore: Williams & Wilkins, 1997:529.

Answers to Dermato-venereological Quiz on page 139

Answer (Question 1)

1. Pseudoxanthoma elasticum.
This is an inherited generalized degenerative disease of elastic tissue, of which autosomal dominant and recessive variants exist. The disorder consists of characteristic skin lesions involving flexural sites, ocular involvement and cardiovascular manifestations.
2. Histologically, the elastic fibres are irregular, thickened and fragmented, appearing as widely dispersed granular material amidst normal collagen fibres. The elastic fibres can be highlighted with the von Kossa stain.
3. The gastrointestinal tract (gastrointestinal haemorrhage), cardiovascular system (hypertension and occlusive vascular disease) and eyes (angioid streaks and retinal haemorrhage) are typically involved.

Answer (Question 2)

1. Angiolymphoid hyperplasia with eosinophilia. It presents as pink, purple, or deep-red domed shaped nodules on the scalp, around the ears, and on the neck. Nodules may be solitary, or they may be multiple and discrete or confluent. Some lesions may present clinically as subcutaneous nodules. Nodules of angiolymphoid hyperplasia are more frequent in middle-age women.
2. Histologically, the lesions are predominately intradermal and appear as ill-defined, lobulated masses composed of numerous variably sized vascular channels. The vessels are lined by large endothelial cells with markedly histiocytoid appearances. There is an eosinophilic infiltrate in the surrounding stroma.
3. Blood eosinophilia is present in 10-15% of the patients.