

Tender Erythematous Plaques on Legs

A 54-year-old woman complained of tender, non-pruritic plaques on both legs which had erupted 2 weeks earlier. These lesions were accompanied with arthralgia, weakness, and fever. No other symptoms suggesting infection were referred such as diarrhoea, abdominal pain or sore throat. Her past medical history revealed chronic hypertension, which was well controlled with enalapril 5 mg daily, and right mastectomy due to carcinoma 15 years earlier. Clinical examination revealed 7 cm to 10 cm diameter circumscribed, infiltrated, erythematous coalescent plaques on both legs. Some isolated papules were observed (Fig. 1). No lymphadenopathies were found. The maximum level of temperature reached was 39.4 °C. Pharyngeal swab was collected for bacteriological study, and no positive result was obtained.



Fig. 1. Non-tender, well delimited erythematous plaques on both legs.

Laboratory results showed leukocytosis with a high C-reactive protein (CRP). She was started empirically on vancomycin, but the rash showed no improvement after 3 days of treatment. Chest X-ray study was unremarkable. Skin specimens were obtained and histological findings are shown in Figures 2 and 3.

What is the most likely diagnosis?

- A. Erythema multiforme
- B. Mycosis fungoides
- C. Erythema nodosum
- D. Sweet's syndrome
- E. Blastomycosis

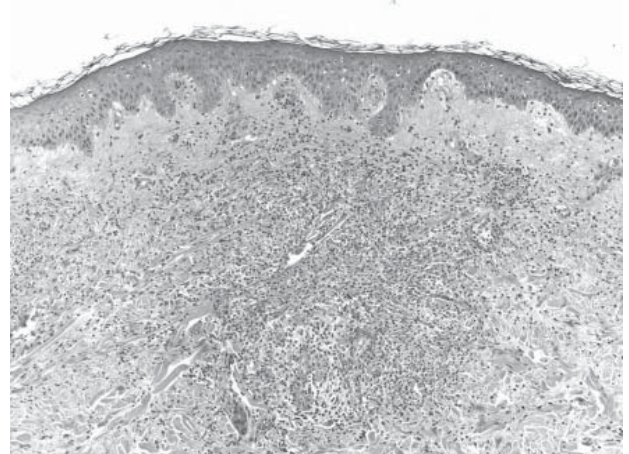


Fig. 2. Panoramic view of findings in histological exam. Findings in subcutis were unremarkable (H and E, original magnification x 5).

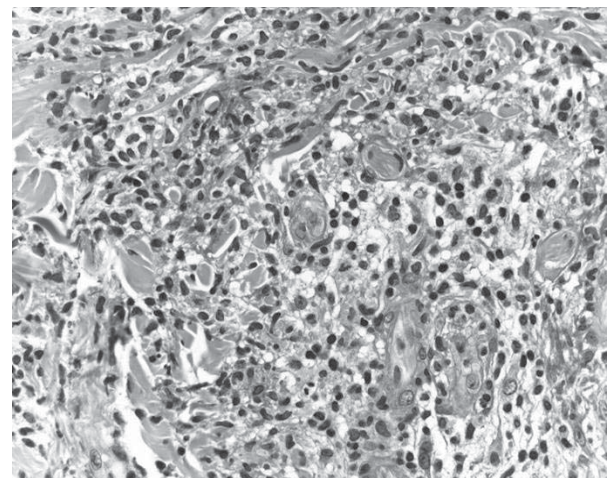


Fig. 3. Neutrophils are most common cells in the inflammatory infiltrate. These cells were not present in the subcutis tissue (H and E, original magnification x 20).

Sweet's syndrome is a condition marked by fever and painful skin lesions that appear mainly on the face, back and legs. Symptoms improve dramatically with systemic corticosteroid therapy.

Erythema multiforme is a type of skin condition of unknown aetiology. Most erythema multiforme are associated with herpes simplex or mycoplasma infections, and cutaneous adverse drug reactions. It often takes on the classical "target lesion" appearance.

Answer: D

Mycosis fungoides is the most common form of cutaneous T cell lymphoma. Plaques, which usually have a fine scale, tend to have a slow development and are preceded by macules and patches.

Erythema nodosum is an inflammatory disorder that involves legs. The lesions begin as firm, hot, red, tender nodules under the skin. Within a few days they may become purplish, then over several weeks, fade to brownish, flat patches. In about half of cases, the exact cause of erythema nodosum is unknown. Some cases may occur with infections (*Streptococcus*, most common), pregnancy and sensitivity to drugs.

Blastomycosis is a rare infection developed when a fungus called *Blastomyces dermatitis* is inhaled from wood or soil. It usually affects people with weakened immune system. Symptoms include chest pain, cough, unintentional weight loss and rash. From the lung, the fungus reaches the blood flow and then the cutaneous tissue. Ulcerated nodules or patches are the most common presentation.

Discussion

The differential diagnosis of erythematous plaques with systemic symptoms includes both infections and non-infections diseases such as erythema nodosum, pyoderma gangrenosum, sarcoidosis, subacute cutaneous lupus, vasculitis, lymphoma, erythema multiforme, leishmaniasis, cellulitis, and atypical mycobacterial infection.

Sweet's syndrome, also known as acute febrile neutrophilic dermatosis, is characterised by fever, neutrophilia, cutaneous eruptions consisting of erythematous papules and plaques, and a dermal non-vasculitic neutrophilic infiltration on skin biopsy. These plaques are painful but non-pruritic.¹ There are 2 main types of Sweet's syndrome—the idiopathic form is characterised by classic lesions of edematous “juicy” red plaques on the upper trunk. Middle-aged women are most likely to develop it. The other common form of Sweet's syndrome is seen as paraneoplastic phenomenon. Therefore, physicians should keep this in mind. Paraneoplastic Sweet's phenomenon is one of the most important features occurring mainly in conjunction with acute myelogenous leukaemia or myelodysplastic syndrome.

Blood count show leukocytosis, and polymorphonuclear cells with marked edema are present in the dermis, without involvement of the vascular endothelium nor evidence of vasculitis.² Workup for patients with Sweet's syndrome should include laboratory studies (with neutrophilia typically presented) including erythrocyte sedimentation rate (ESR) and CRP, chest X-ray (if pulmonary symptoms are presented) and skin biopsy to confirm the diagnosis.

For a definitive diagnosis of Sweet's syndrome, both major and 2 minor criteria should be met. The 2 major criteria are:

(i) abrupt onset of painful erythematous plaques or nodules; and (ii) neutrophilic infiltration in the dermis without leukocytoclastic vasculitis. The minor criteria are: (i) Skin lesions; (ii) Accompanied by periods of general malaise and fever; (iii) Laboratory values during onset: ESR > 20mm, CRP positive, neutrophils >70% leukocytosis >8000 (3 of these 4 values are necessary); and (iv) Excellent response to treatment with systemic corticosteroids.^{3,4}

Conclusion

The patient's symptoms and skin lesions rapidly improved with prednisone 30 mg per day. No recurrence has been observed during the follow-up.

REFERENCES

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