Painful bullae in a febrile woman: a clue to first presentation of multibacillary Hansen's disease

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DESCRIPTION

A woman in her 40s presented to the emergency department with 8 days of fever and malaise. Along with the fever, crops of multiple painful reddish raised lesions appeared for the first time on the face, upper limbs and trunk. There was no history of loss of sensation or weakness of the extremities. No significant history of systemic complaint was present. She received only symptomatic management for these complaints from a general physician with no improvement of the lesions. On examination, she was febrile with a temperature of 38.5°C, heart rate of 104 beats per minute and blood pressure of 104/60 mm Hg. Multiple, tender, oedematous and erythematous plaques, few of which with central flaccid to tense bullae containing clear to turbid were distributed on forehead, trunk and extremities (figure 1A,B). There were no other significant findings including other skin lesions like hypoaesthetic or anaesthetic macules or plaques were not evident clinically. Palpation of peripheral nerves, sensory and motor examinations were within normal limits with no hand and feet deformity. Laboratory tests revealed mildly raised leucocytosis, neutrophilia (92%) and elevated inflammatory markers (procalcitonin of 3 ng/mL and C reactive protein of 14 mg/L). Due to the characteristic cutaneous lesions, skin punch biopsy for histopathological examination was done from a fresh erythematous nodule on the left forearm with a suspicion of Sweet's syndrome (SS) with its bullous variant, which revealed diffuse and intense dermal oedema with separated collagen, diffuse foamy histiocyte aggregates with periadnexal accentuation of lymphohistiocytic collections (figure 2A, H&E and 4×). Furthermore, Fite Faraco stain was done on skin biopsy section which showed several globi of lepra bacilli distributed mainly around the hair follicles and dermal blood vessels suggestive of erythema nodosum leprosum with Fite stain showing multiple globi of lepra bacilli (figure 2B). MB-MDT (multibacillary-multidrug therapy), prednisolone 40 mg, non-steroidal anti-inflammatory drugs were started with improvement of the lesions after 2 weeks of follow-up.

Reactions in leprosy occur as episodes of acute or subacute immunologically mediated inflammation which disrupts the relatively uneventful disease course affecting skin, nerves, mucous membrane or other sites.[1] Type 2 lepra reaction occurs mostly in multibacillary cases especially in lepromatous leprosy and borderline lepromatous leprosy cases.[1] Erythema nodosum leprosum (ENL) is a characteristic lesion of Type 2 reaction which manifests as crops of multiple erythematosus, evanescent, tender nodules and plaques with predilection of extremities, face and trunk. It is usually accompanied by constitutional symptoms like fever, malaise, arthralgia along with systemic complications.[2] Bullous lesions are one of the rare atypical presentation of Type 2 reactions and only a few cases have been reported in the literature.[1–3] Clinical signs of ENL may mimic SS in the setting of normal sensory and motor peripheral nerve examination. SS or acute febrile neutrophilic dermatosis presents as abrupt onset erythematous, tender nodules and plaques with constitutional symptoms.[3–5] The characteristic laboratory findings of SS include neutrophilia, raised erythrocyte sedimentation rate (ESR), C reactive protein with dense neutrophilic infiltrate in the dermis without evidence of leucocytoclastic vasculitis.[3] The diagnosis of ENL may be missed especially when it presents as the first manifestation of Hansen's disease in a treatment naive patient leading to delay in initiation of definitive treatment, prevention of disability and deformity.

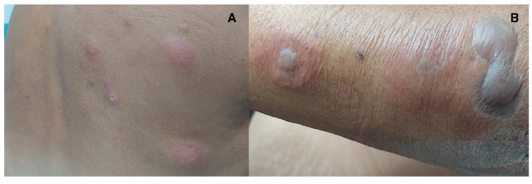

Figure 1 (A) Multiple well-defined, erythematous, tender plaques on the the trunk. (B) Well-defined, erythematous plaques with centrla bullae seen on extensor aspect of left forearm.

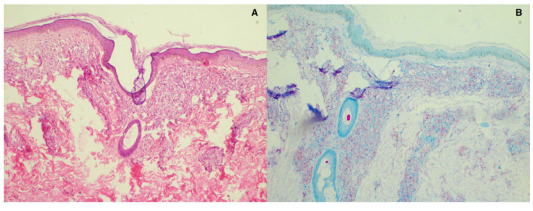

Figure 2 (A) Extensive dermal oedema with disintegrated collagen, diffuse foamy macrophages in the papillary dermis and around adnexa; neutrophils and lymphohistiocytic aggregates are seen in perivascular and periadnexal areas (H&E,40×). (B) Fite stain shows numerous globi of lepra bacilli around superficial vessels and hair follicles (Fite 40×).

Learning points

- Erythema nodosum leprosum may masquerade as Sweet's syndrome (SS).
- Bullous lesions are rare atypical presentation seen in both erythema nodosum leprosum (ENL) and SS.
- Bullous ENL may often be misdiagnosed when it is the first manifestation in a setting of normal sensory and motor peripheral nerve examination.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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