Erythema nodosum leprosum (ENL) mimicking Pityriasis lichenoides et varioliforme acuta (PLEVA): An atypical presentation

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Introduction

Erythema Nodosum Leprosum (ENL) or Type 2 lepra reaction is found in the lepromatous pole of leprosy.1 It usually presents as tender, erythematous, evanescent nodules associated with systemic features. Besides nodular lesions, various reported atypical presentations of ENL include pustular, bullous, ulcerated and erythema multiforme-like lesions. Atypical presentations often lead to delay in diagnosis and treatment which may lead to severe systemic complications in ENL. We are reporting a 50 year old male who presented with recurrent papular and pustular crusted skin lesions associated with mild fever and was misdiagnosed as PLEVA initially, but later on was proved to have lepromatous leprosy with erythema nodosum leprosum on histopathology.

Case report

A 50 year old farmer presented with recurrent episodes of fluid filled skin lesions for 3 months which was healing leaving hypopigmented macules and scars on his body. These episodes were associated with mild fever and joint pain. Examination revealed few asymptomatic skin coloured to erythematous papules of <0.5 cm, multiple vesicles, few
vesicles with central crust (variolliform) and multiple healed hypopigmented and hyperpigmented macules with scars over his trunk and extremities (Figures 1 and 2).

The patient had received various courses of antibiotics with no improvement. There was no past history suggestive of any infections or any other drug intake. Based on these findings a clinical diagnosis of PLEVA was considered. All routine investigations were within the normal range. Serology for HIV was negative. A biopsy from a crusted vesicle revealed subepidermal separation, foamy macrophages and neutrophilic infiltration in the dermis, perivascular neutrophilic and lymphocytic infiltration suggestive of lepromatous leprosy with ENL (Figures 3, 4 and 5).

AFB staining of the same tissue showed foamy macrophages containing bacilli in single and globi suggesting the lepromatous pole (Figure 6).

Following histopathology findings, a slit skin smear from the back and right ear lobe was done, which showed a bacillary index of 5+ and MI (morphological index) of 90%. The diagnosis was revisited and repeat clinical examination did not reveal any other features suggestive of lepromatous leprosy and other features of ENL except glove and stocking anaesthesia. A final diagnosis of lepromatous leprosy with ENL was made. The patient was
started on treatment with WHO MDT-MB (A) and oral prednisolone 40 mg per day. His skin lesions improved over a week. The patient is currently under treatment with MDT-MB(A) and oral steroid on tapering doses without any recurrence of lesions.

Discussion

Erythema nodosum leprosum (ENL) or Type 2 reaction is an immune-mediated complication of leprosy commonly occurring in lepromatous or borderline lepromatous leprosy.¹ It is a Type III hypersensitivity reaction, due to the formation of circulating immune complexes. Immune complex deposition triggers a vasculitic process resulting in release of TNF-α and subsequently an inflammatory episode resulting in skin lesions and systemic symptoms.² Vascular changes are common in ENL and it has been hypothesised that vasculitis is the major pathological event in ENL.² It causes an acute inflammatory reaction which can occur in any organ or tissue invaded by the leprosy bacillus.³ However, recent evidence suggests some role of cell mediated immunity in the pathogenesis of ENL.³
ENL usually occurs within the first 6 months of initiation of MDT, however, it can develop any time before, during or after the completion of treatment. The skin lesions are more common over the extremities and may present as superficial or deep seated, erythematous, tender papules or nodules which are evanescent and heal within 7–10 days.

**Figure 3.** Subepidermal separation with perivascular neutrophilic and lymphocytic infiltration and vasculitis suggestive of ENL (H & E x10)

**Figure 4.** Subepidermal separation with neutrophilic infiltrate (H & E x40)
with post-inflammatory hyper pigmentation. Besides typical ENL lesions, atypical presentations of ENL include pustular, bullous, ulcerated and erythema multiforme-like lesions.\textsuperscript{4–9}

Similarly PLEVA presents as an acute-to-subacute eruption of multiple, small, erythematous papules that rapidly develop into polymorphic lesions at various stages of evolution, such as vesicles, pustules, hemorrhagic crust covered papules, and shallow ulcers.

\textbf{Figure 5.} Foamy macrophages and epitheloid cells (H & E x40)

\textbf{Figure 6.} Foamy macrophages with positively stained bacilli in single and globii suggesting lepromatous pole of Hansen’s disease (ZN stain x40)
These lesions heal in weeks to months with hyper/hypopigmented and pox-like scars. Histopathologically, PLEVA is characterised by diffuse dermal inflammatory infiltrate concentrated along a basal layer with prominent lymphocytic and erythrocytic exocytosis into the epidermis.

The diagnosis of ENL is easily made by clinical examination in a patient of Hansen’s disease and rarely needs confirmation by histopathology. However, in the absence of classical features of Hansen’s disease diagnosing atypical presentations of erythema nodosum leprosum is often difficult and can result in misdiagnosis. The classical changes described in the histopathology of ENL include neutrophil rich inflammatory infiltrate in the deeper dermis, infiltrating within pre-existing lepromatous lesions and often associated with vasculitis.

Our case presented with repeated episodes of fever with skin lesions in the form of papules, vesicles with central crusting (varicella like) and lesions healing with hypopigmented scars. The patient was clinically diagnosed as PLEVA, but was later on proved to have lepromatous leprosy with ENL on histopathology. Except for glove and stocking anaesthesia there was no other clinical evidence of lepromatous leprosy in the patient.

In a review of literature we couldn’t find any PLEVA like presentation of ENL. We suggest in an endemic area of leprosy this type of clinical presentation of erythema nodosum leprosum should be considered in differential diagnosis, so that the patient can be diagnosed and treated early to avoid systemic complications of ENL.

References