Reactional State in Lepromatous Leprosy Simulating Sweet’s Syndrome

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Abstract

Erythema nodosum leprosum (ENL) or Type-2 lepra reaction is a manifestation of type-III hypersensitivity response, and usually occurs in certain cases of lepromatous and borderline lepromatous leprosy. ENL may present as widespread crops of erythematous, inflamed nodules and papules. Rare variants of ENL mimicking pemphigus or Sweet’s syndrome (SS) have been documented. Here, we report an unusual case of persistent ENL in a 52-year-old lady, which we could diagnose with the help of skin biopsy and histopathological examination.

Introduction

Leprosy reactions are immunological phenomena that occur before, during or after the completion of multi-drug therapy (MDT). Two major complications of leprosy are type-1 reactions and ENL. Type-1 lepra reaction is a delayed type of hypersensitivity reaction occurring in borderline leprosy cases. Type-2 lepra reaction or ENL is an antigen antibody-mediated immune complex reaction, and is usually seen in lepromatous or sometimes borderline lepromatous leprosy.

Case Report

A 52-year-old female presented with painful generalised rash all over her body without any pruritus for 1½ year. She was diagnosed by the local dermatologist to be suffering from erythema nodosum leprosum (ENL), and was put on to rifampicin, dapsone, clofazimine, methyl prednisolone and pantoprazole after performing slit-skin smear test. The patient symptomatically improved but the rash persisted.

Examination revealed a conscious, ill-looking female who had mild pallor. Neither she had icterus, cyanosis, clubbing, oedema, nor was there any lymphadenopathy. Her vitals were stable and the temperature was 37.2°C. Her skin was purplish with scattered tender pustules and nodules over her face and limbs. Other systemic examinations were within normal limits except for the thickened peripheral nerves.

A provisional diagnosis of Hansen’s disease with lepra reaction was made. Routine blood count and biochemistry showed a leucocytosis with total leucocyte count of 18,200/mm³ and a differential count of 82% neutrophils, 15% lymphocytes and 3% monocytes, and an ESR of 64 mm in 1st hour (Westergren). Blood sugar, urea, creatinine, electrolytes and liver function tests were within normal limits. Intravenous antibiotics were started. The skin lesions worsened with pustular appearance and some of them ruptured giving rise to punched-out lesions (Figure 1). A biopsy from the skin lesion was sent for histopathological analysis, staining for bacteria / acid-fast bacilli (AFB) / fungi, and culture. She was prescribed rifampicin, clofazimine, dapsone, colchicine and a rapidly tapering course of dexamethasone. The lesions did not improve even after 7 days of therapy compelling us to add thalidomide; thalidomide was prescribed
in three divided doses of 100 mg each, and the patient was regularly monitored for potential side effects of venous thromboembolism, drowsiness, constipation, concurrent renal damage and neuropathy. The patient improved remarkably with the above treatment. Histopathology report was consistent with Sweet’s syndrome showing evidence of dense perivascular infiltrate composed of predominantly neutrophils, leucocytoclasis with a notable number of histiocytes and a few lymphocytes in the infiltrate (Figure 2, Figure 3). Evidence of oedema was seen in the upper dermis with numerous thin walled blood vessels and infiltrates. The papillary dermis was unaffected. Staining for bacteria, AFB, fungi were negative and the culture revealed no growth.

Discussion
Sweet’s syndrome is an acute neutrophilic dermatosis characterised by fever, neutrophilia, tender erythematous skin lesions (papules, nodules and plaques) and a diffuse infiltrate consisting predominantly of mature neutrophils that are typically located in the upper dermis in the absence of leucocytoclastic vasculitis. The oedema in the upper dermis of the lesions, results in their transparent, vesicle like appearance and has been described as an illusion of vesiculation. Infections associated with SS are Streptococci, Yersiniosis, Blastomycosis, Mycobacterium chelonae and others.

The differential diagnoses entertained in a case of ENL are erythema nodosum, panniculitis and sepsicaemia. Rarely ENL clinically present as urticarial vasculitis, severe bullous variety mimicking pemphigus or Sweet’s like lesions. Lesions of erythema nodosum are usually evanescent unlike ENL. Abscess or septicaemia was ruled out by staining for the micro-organisms and subsequent culture. Leucocytoclastic vasculitis would have shown vessel wall destruction, extravasated erythrocytes, fibrinoid necrosis of the vessel wall, karyorrhexis, and neutrophils in the vessel wall. Simultaneous occurrence of Sweet’s syndrome and ENL is very rare.

Our patient was previously suffering from leprosy as was proved by positive slit-skin smear from multiple sites, and thickening of peripheral nerves. Occult malignancies, rheumatological disorders and inflammatory bowel disease, commonly associated with SS, had been ruled out from history and appropriate investigations. She was not on any offending drugs prior to her illness. She improved
symptomatically with MDT for leprosy though her skin lesions persisted.

SS type leprosy reaction, though rare, has been reported previously. Wade-Fite stain or Fite-Faraco stain used for staining both living and dead *Mycobacterium leprae* or monoclonal antibodies against *Mycobacterium leprae* antigens can absolutely differentiate SS from Sweet’s like ENL lesions.\(^5\) It could not be performed in our case, hence we report it as a case of Hansen’s disease presenting with severe Sweet’s like reactional state.

The first line medications for this disease include corticosteroids, potassium iodide and colchicine while the second line medications include indomethacin, cyclosporine, clofazimine and dapsone. In addition, effective treatment of Sweet’s syndrome has also been described with cyclophosphamide, chlorambucil, immunoglobulin, interferon-α, etretinate, etanercept, infliximab and thalidomide.\(^1\) Our patient did not improve with the first and second line medications and was therefore put on to thalidomide with which she improved dramatically.

**Conclusion**

ENL is a serious immunologic complication of borderline lepromatous and lepromatous leprosy. The presentation is varied and sometimes mimics SS, thereby confounding the physician. Our patient had type – 2 lepra reaction with constitutional symptoms. It responds remarkably well to Thalidomide treatment. Concurrent renal damage and other side effects, if any, should be assessed. An early diagnosis with the help of skin biopsy and intensive management are essential to minimise deformity and disability.

**References**


