Lepromatous leprosy presenting as Erythema multiforme – A rare presentation

Janardhan Bommakanti1*, G.K. Prasad2, Vani.G3, N.Vidyavathi4

1Associate Professor, 2Prof & Head, 3&4Residents, Department of Dermatology, Venereology & Leprosy (DVL), MediCiti Medical College, Medchal mandal, Ranga Reddy district, Telangana, India.

ABSTRACT
A 30 year old female presented as Erythema multiforme (EMF) with classic target lesions. Histology was also consistent with EMF. Differential diagnoses of Sweet’s syndrome & Urticarial vasculitis were entertained. Two weeks later she developed Erythema Nodosum Leprosum (ENL) like lesions. Skin Slit smears at second visit were positive for Acid fast bacilli (lepra) [AFB(L)] and histopathological / Fite Faraco stain for lepra bacilli was consistent with leprosy. We are reporting this case because of rare presentation of lepromatous leprosy as EMF.

KEYWORDS: Erythema multiforme, Lepromatous leprosy, Erythema nodosum leprosum.

INTRODUCTION
Hansen’s disease manifests in different forms, classified according to Ridley-Jopling classification based on clinical, microbiological, pathological & immunological criteria. Hansen’s disease is associated with type 1 (Reversal) and type 2 (Erythema nodosum leprosum) immunological reactions, during which the disease process appears to worsen dramatically. These reactions may occur at any time before, during or after treatment.

Erythema Nodosum Leprosum (ENL) or Type 2 Lepra reaction occurs in lepromatous leprosy (LL) and also in borderline lepromatous leprosy (BL). ENL presents as erythematous, often tender, papules, nodules, plaques, vesicles, pustules & blisters associated with constitutional symptoms. Rarely, it may present as Erythema multiforme, which also is an immunological reaction pattern. [1] We report this case presenting as EMF initially, later manifesting classical features of ENL. So far only 2 cases have been reported as per our knowledge.

CASE REPORT
A 30 yr old married female, housewife, resident of Toopran, Telangana state presented with multiple red coloured, raised, ring shaped skin lesions over face & extremities, appearing in crops [figure 1], associated with intermittent fever & chills of 1 week duration. Lesions were painful, no history of cough / sore throat / blisters over lip margins. No history of itching, nasal stuffiness or epistaxis. No history of sensory loss or muscle weakness. No history of drug intake. On examination, multiple target lesions of various sizes were distributed over face, extensor aspects of both upper & lower limbs. Conjunctivae, oral & genital mucosae are not affected. No loss of sensations over lesions. No lymphadenopathy.

Differential diagnosis of Erythema multiforme, Urticarial vasculitis & Sweet’s syndrome were considered. On investigation, blood count showed leukocytosis. X-ray chest PA view– Normal, Herpes virus (HSV) 1& 2 IgM/IgG serology was negative. Vitals were in normal range. Biopsy from the Target lesion was suggestive of Erythema multiforme. Final diagnosis was Erythema multiforme. Managed symptomatically.

She came back two weeks later with diffuse swelling over nose, painful red coloured raised lesions over face, both upper & lower limbs [figure 2&3]. On detailed enquiry, she gave history of erythematous, painful lesions over body three months earlier, which subsided spontaneously. Cutaneous examination revealed multiple tender warm erythematous nodules over bridge of the nose, trunk, and extensor aspects of both upper limbs and lower limbs. Peripheral nerve examination revealed thickened and tender left ulnar nerve, bilateral lateral popliteal & posterior tibial nerves. No motor deficit. Skin Slit Smear for Acid Fast
Bacilli (L) was Positive with Bacillary index (BI) 4+. Biopsy done at 2nd visit from nodule over right arm was consistent with Erythema nodosum lepromatous (H&E stain). Fite Faraco stain [figure 4] demonstrated AFB (Lepra).

Figure:1 Typical Target /iris skin lesions (EMF) over forehead

Figure:3 Diffuse nodular swelling over the nose and target lesion over forehead

Figure:2 EMF lesions on right arm & ENL lesions on right forearm (2nd visit/8th day)

Figure:4 Fite Faraco stain x1000 magnification shows clumps of lepra bacilli

DISCUSSION

EMF is a reaction pattern to many different triggering factors with immunological basis. In up to 50% of the cases there is no demonstrable provoking factor. The most common association is with preceding herpes infection type 1 / 2 or mycoplasma infection. Other bacterial / viral infections and drugs are also incriminated. In our case Mycobacterium leprae was the most probable triggering factor. Clinically EMF manifests with target / iris skin lesions, macular, papular or urticarial lesions. It can occur at any age in both sexes. In general the course may last over a few days to 2-3 weeks. Repeated attacks of EMF are commonly associated with recurrent herpes simplex infection. In this case there was no clinical or serological evidence of herpes virus infection. [2]

EMF is of 4 types. Minor/ simplex form, major, localised vesiculo bullous & atypical. Minor form accounts approximately 80% of cases of EMF showing typical target lesions which are less than 3 cm diameter and restricted to few areas of the body, which is seen in this case. They are rounded & have 3 zones: central area of dusky erythema / purpura, middle pale zone of edema & outer ring of erythema with well defined edge. Atypical lesions have only 2 zones. Lesions appear in successive crops for few days and fade in 1-2 weeks sometimes leaving dusky discolouration. EMF lesions may be few to very profuse.
Photo aggravation is well known. Mild EMF cases managed symptomatically but in severe cases, systemic corticosteroids may be given, which is debatable. [2]

ENL lesions are usually painful, erythematous nodules occurring in crops in lepromatous / borderline lepromatous leprosy patients. Coombs & Gell’s type III hypersensitivity reaction is implicated in pathogenesis of this condition. Deposition of circulating or extravascular immune complexes in various tissues is responsible for manifestations. Usually associated with fever, chills, malaise & arthralgias. ENL may present with papules, plaques, vesicles, pustules, blisters & frankly necrotic lesions. The triggering factors are concurrent illness, vaccinations, stress, medications, pregnancy & lactation.[3]

The bacteriological index is usually high in ENL patients.[4] Switching of anti-leprosy drugs to 2nd line drugs like Ofloxacin resulted in pustular variety of ENL.[5] A case of ENL with severe skin ulceration had to be treated with thalidomide in addition to systemic corticosteroids.[6] Sweet syndrome/Gomm button disease like presentation was has been reported by Kou & Chan et al.[7] Bentekan et al reported case presenting initially with laryngeal dyspnoea.[8] Rarely, ENL may present as Erythema multiforme like lesions. This patient, presented initially as Erythema multiforme, with no obvious etiologic factor. Later, she developed full features of ENL. Detailed clinical history, examination and histology both Haematoxylin & Eosin (H&E) and Fite Faraco stain were consistent with Lepromatous leprosy with ENL.

The patient was started on multi bacillary- multidrug treatment (MB MDT) along with systemic corticosteroids. [9] EMF like skin lesions in leprosy patient was mentioned by Ramu G & Dharmendra et al [10] & also by Das, Roy, Giri et al who reported one case which presented initially as granuloma annulare and other as EMF.[11] We report this case of Erythema nodosum leprosum presenting initially as Erythema multiforme which is very uncommon. Therefore high index of suspicion of leprosy is necessary with uncommon presentation particularly in endemic areas.

REFERENCES


*Corresponding author: Dr. Janardhan Bommakanti E-Mail: jannub6@rediffmail.com