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Case Report

Erythema Nodosum Leprosum Presenting as Annular Papuloplaque Lesions in a Christmas Tree Distribution

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Leprosy is a chronic granulomatous disease whose course is complicated by reactions. Type 2 reaction or erythema nodosum leprosum (ENL) is an immune complex mediated reaction which can present with varied manifestations. Here we describe a case of ENL in Lepromatous Leprosy patient who was on standard MDT and presented with annular papuloplaque lesions with collarette scale having Christmas tree distribution.

Keywords : Erythema nodosum leprosum, ENL, atypical, leprosy, Christmas tree presentation

Introduction

Leprosy is a chronic disease caused by *Mycobacterium leprae*. Its course is complicated by acute onset of inflammatory reactions which can occur before, during and after treatment of the disease. The reactions are classified as Type 1 reaction or reversal reaction and Type 2 reaction or erythema nodosum leprosum (ENL). Reversal reaction is a Type IV delayed type hypersensitivity reaction while erythema nodosum leprosum is immune complex mediated Type 3 hypersensitivity reaction (Ramesh & Pahwa 2010). These reactions can present with varied manifestations as the disease itself. The treatment of reaction is sometimes more challenging than the disease per se. Here we report a case of ENL in lepromatous leprosy who presented with annular papuloplaque lesions in a Christmas tree distribution.

Case Report

38 year old male presented with spontaneous blistering and numbness of hands and feet for the last 1 year. On physical examination there were multiple hypopigmented anaesthetic patches over upper limbs, back and lower limbs along with few skin coloured nodular lesions. Multiple thickened peripheral nerves with glove and stocking anaesthesia was also present. A single nodular lesion was also present over upper lip, and madarosis of both eyebrows was observed. Slit skin smear from ear lobe revealed several AFB on Ziehl Neelson staining with a Bacteriological Index (BI) of 6+. The patient was diagnosed as a case of Lepromatous leprosy (LL) and started on multibacillary multidrug therapy (MDT-MB). After two months the patient presented with multiple red, raised, painful evanescent lesions

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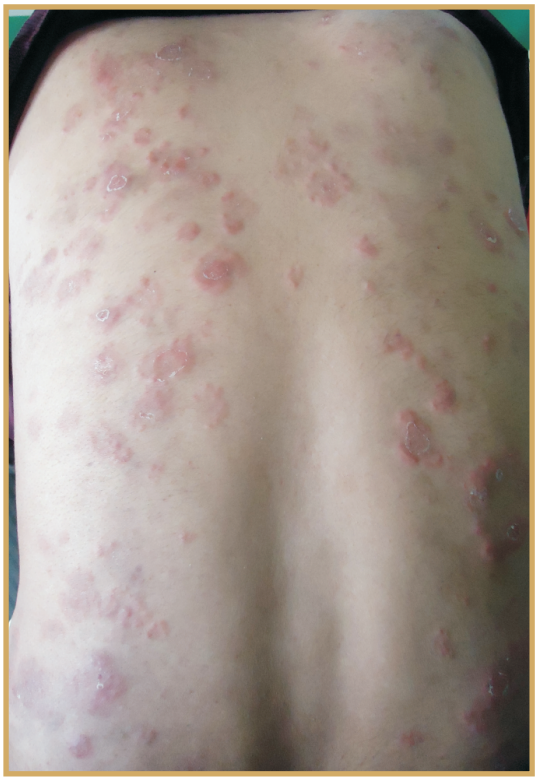

Fig 1 : Annular papuloplaque lesions over back with a Christmas tree distribution.

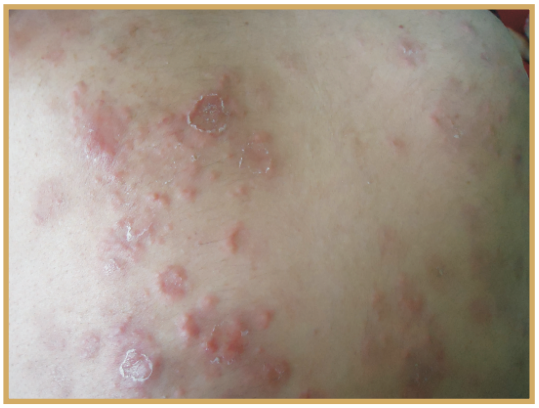

Fig 2 : Close up view of lesions showing collarette scale.

over face, back, outer aspect of arms and thighs. These cutaneous lesions were present for last

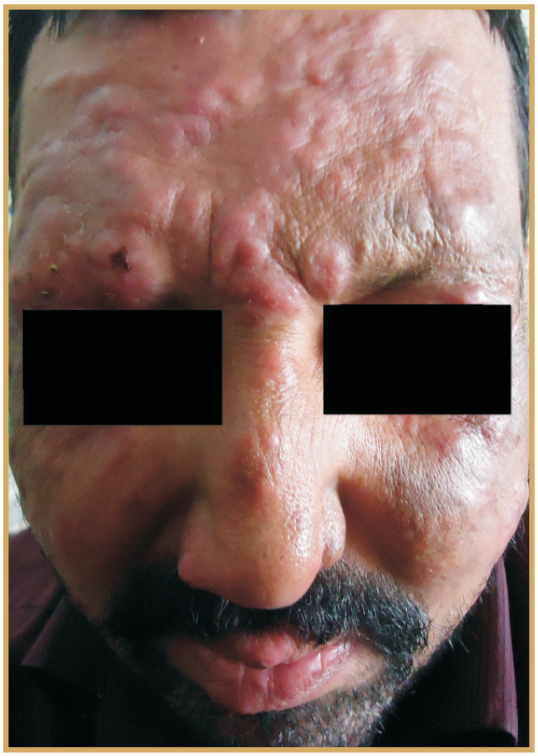

Fig 3 : Multiple erythematous nodules over face with madarosis.

15 days. These were also associated with history of fever and joint pains. On examination the patient was febrile, and there were multiple erythematous papuloplaque lesions arranged in annular configuration with collarette scale over the back. These lesions were present in a Christmas tree distribution (Figs 1, 2). Also there were multiple erythematous tender nodules over face, extensor surfaces of arms and thighs (Fig 3). These were associated with neuritis in right ulnar nerve. Complete hemogram was normal except for raised ESR which was 30 mm in first hour. Urine examination, renal function test and liver function test were within normal limits. Patient was diagnosed as having Type 2 reaction clinically, and started on oral prednisolone 40 mg per day, alongwith continuing MDT. Steroids were slowly

tapered off subsequently according to the NLEP guidelines. There was early resolution of lesions, and fever and the neuritis subsided completely within 2 weeks.

Discussion

Type 2 lepra reaction or Erythema nodosum leprosum (ENL) is usually seen in Lepromatous leprosy and borderline lepromatous leprosy (Yogeesh et al 2012). It is characterised by acute onset of crops of painful erythematous evanescent nodules on face, back and extensor surface of limbs. It is a multisystem disorder which manifests with fever, myalgias, bony pains, neuritis, arthritis, orchitis, lymphadenitis, uveitis and glomerulonephritis. Pathogenesis of Type 2 reaction involves increased expression of Th2 cytokineprofile i.e. IL-6, IL-8, and IL-10, IL-4 and IL-5. This results in increased neutrophil infiltration, activation of complement and formation of immune complexes which cause tissue damage (Pandhi and Chhabra 2013). In a study by Kar and Gupta L (2016), 13% of leprosy patients presented with ENL at the time of diagnosis of leprosy, 89.39% patients developed T2R while on MDT whereas 10.61% developed T2R after RFT (Released from treatment).

Atypical presentations of ENL include vesiculobullous, pustular, ulcerated, hemorrhagic and erythema multiforme-like lesions. (Ramesh and Pahwa 2010, Yogeesh et al 2012, Verma et al 2015, Chauhan et al 2006). Prabhu et al (2008) have reported a persistent and localized variant of ENL. Lesions simulating sweet syndrome have been reported by Kou and Chan (1987). Severe Type 2 reaction also manifests as necrotic ulcers and is known as Erythema nodosum necroticans. Kar et al (2009) also described a case of ENL manifesting with annular vesicobullous lesions in a patient of Borderline lepromatous leprosy. Dermal oedema and leucocytoclastic vasculitis have been suggested as the possible cause for bulla formation by them. Similar lesions were reported by Goyal et al (2016), in a young male who was on ofloxacin along with MDT. Shah et al (2009), reported Erythema Multiforme like and annular hemorrhagic bullous lesions in a female patient who was on standard MB MDT and daily OfloxacinIt was hypothesized that, Ofloxacin being a bactericidal drug, increases the antigenic load when combined with MDT and increases the severity of reaction. Mahajan et al (2012), reported a case of Type 2 lepra reaction which mimicked Hodgkin's lymphoma. Their patient presented with generalised lymphadenopathy, acquired ichthyosis and constitutional symptoms but there were no cutaneous lesions of ENL. Vijendran et al (2014) described arare presentation of ENL in a female, which were distributed in a photo distribution manner.

Our patient presented with annular papuloplaque lesions with collarette scale in a Christmas tree distribution. Earlier reports showed presence of vesicobullous lesions in annular configuration especially in patients who were on ofloxacin along with MDT. In our patient ENL lesions had annular configuration but they were not vesicobullous indicating less severity of reaction as he was only on MDT alone. The lesions showed collarettte scale and were in Christmas tree distribution which was unique in this case. Collarette scale are usually seen in Pityriasisrosea but is also present in subsiding lesions of Furuncle, Miliaria and Erythema Nodosum (Kangle et al 2006).

ENL usually develops during first six months of MDT. Patients with high BI (> 4+) have high prevalence of ENL. Other precipitating factors are physiological stress, mental stress, antileprosy drugs, pregnancy, infection and using antibiotics with antileprotic action for other ailments (Ramesh and Pahawa 2010). Severity of ENL is more in Caucasians and Mongolians than in Negroes (Yogeesh et al 2012).

There is wide experience of treatment of leprosy reactions including ENL (Kar and Gupta 2016). The drugs of choice in treatment of ENL with neuritis

are steroids. Thalidomide is also a good drug but does not help in relieving the neuritis, for which steroids have to be added. In ENL with ocular involvement and iridicyclitis steroids are mainstay of treatment alongwith atropine to dilate the pupils. Other anti-inflammatory agents like Asprin, NSAIDs, Colchicine and Sodium antimony gluconate have been used for treatment of milder ENL reactions. Increasing the dose of Clofazimine to 300 mgs also helps in the treatment and helps in tapering off of steroids in chronic and recurrent ENL reactions. Our patient responded well to systemic steroids, and there was no recurrence in the limited follow up period. In chronic and recurring ENL reactions Thalidomide, Methotrexate and immunomodulators including MIP have been used for treatment. With their usage the steroids could be tapered off effectively and side effects of long standing steroids can be prevented.

Several atypical presentation of Type 2 lepra-reactions have been described in literature. Our patient presented with annular papuloplaque lesions in a Christmas tree distribution with collarette scale. ENL lesions in a Christmas tree distribution have not been described in literature to the best of our knowledge. Unusual presentation of Type 2 leprareaction should be kept in mind for prompt diagnosis and treatment. Early diagnosis and treatment of Type 2 leprareaction can be used for treatment of neuritis and thus reduce the disability rate.

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