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ERYTHEMA NODOSUM LEPROSUM : A LONG BATTLE TO FIGHT

Dermatology

Dr. Apoorva Gupta* Junior Resident 3, Department of Dermatology, School of Medical Sciences and Research, Sharda University, Greater Noida, Uttar Pradesh, India. *Corresponding Author

Dr. Goel Shitij Professor and Head, Department of Dermatology, School of Medical Sciences and Research, Sharda University, Greater Noida, Uttar Pradesh, India.

ABSTRACT

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KEYWORDS

INTRODUCTION

Leprosy is a chronic, slowly progressive, granulomatous infection caused by the bacillus Mycobacterium leprae. Usually 15-50 percent of lepromatous leprosy (LL) patients develop Erythema Nodosum Leprosum (ENL) reactions within the first year of therapy. Classically, ENL presents as crops of tender, erythematous papules, plaques, or nodules that are evanescent and last only 7-10 days.Extracutaneous manifestations include fever with constitutional symptoms, neuritis, arthritis, epididymo-orchitis, dactylitis, synovitis, iridocyclitis, rhinitis, epistaxis.[1]

Case Report

A 42 year old male presented with history ofrecurrent attacks of painful nodular skin lesions, since one year. The lesions first appeared over the right lower limb and slowly developed over the other sites. In the current episode, the patient had multiple painful nodular skin lesions over both the ears, lower extremities and trunk since 4 days. The current episode was more severe and associated with high grade and continuous fever, one episode of epistaxis, swelling of both the lower limbs, increased lacrimation & hoarseness of voice. There was a history of rapid tapering of oral corticosteroids over the last one month before the onset of symptoms. On asking, the patient revealed history of intake of Universal Multi Drug Therapy for Leprosy (UMDT)for one year, 10 years ago.

On examination, the patient had multiple well defined painful and tender subcutaneous, erythematous nodules ranging from size 0.5 to 1cm & pustules on both the ears, lower extremities, and trunk. A few nodules were crusted with serosanguinous discharge. Right ulnar and radial nerves were thickened but non-tender, associated with wasting of thenar muscle. Histopathological examination done from a nodule over right thigh revealed separation of epidermis from dermis by a narrow zone of collagen (Grenz zone).

Dense infiltrate of neutrophils was seen which was extending from the dermis up to the subcutis. Macrophages with vacuoles and globi were in large number. Fite-Faraco Stain confirmed the presence of acid-fast bacilli. Slit Skin Smear revealed taken from the earlobe and skin nodule showed bacteriological Index of +3. Finally a diagnosis of LL Hansen's with Erythema nodosum leprosum was made. Patient was started on UMDT with prednisolone 40 mg/day. On subsequent follow-up, patient was better with healing of all the lesions.

DISCUSSION

Type 2 Lepra reaction, popularly known as ENL is an immune complex syndrome. It is an example of type III hypersensitivity reaction (Coombs and Gell classification). Type 2 reaction (T2R) occurs mostly in LL and sometimes in BL leprosy. Patients with high bacillary index are more prone to get ENL.[2]Type 2 reaction occurs mostly during antileprosy treatment. A few cases present for the first time with features of reaction before leprosy is diagnosed and treatment started. ENL lesions may appear anywhere, deep in the dermis and subcutaneous tissue and these may not be clinically apparent on the surface of the skin. There is sudden appearance of crops of evanescent (lasting for few days) pink (rose) coloured tender papules, nodules, or plaques variable in size. The nodules are dome-shaped and ill-defined, warmer, and blanch with light finger pressure. They may appear anywhere on the skin except the hairy scalp, axillae, groin and perineum, the warmer areas of the body. The fresh crops of ENL lesions usually appear in the evening when endogenous cortisol production is at its lowest.[3] New ENL lesions appear, as old lesions subside. An individual ENL lesion after a period of 24–48 hours show a change of colour from pink/red to bluish and brownish and finally dark, in a week or 10 days. ENL lesions may commonly become vesicular, pustular, bullous, and necrotic and break down to produce ulceration called as erythema nodosum necroticans.[4] In severe T2R there may be swollen, painful, and tender nerve trunks with loss of function. Careful nerve examination is essential to detect recent nerve function impairment (sensory, motor, and autonomic functions) of the respective affected nerves during and after each episode of T2R. However, skin involvement may not be always associated with manifest nerve lesions. Other associated features are myositis, arthritis, synovitis, rhinitis, epistaxis, laryngitis, iridocyclitis, glaucoma and painful dactylitis. There may be periosteal pain (particularly in tibia), tender and swollen lymph nodes (especially inguinal and femoral groups), acute epididymo-orchitis, nephritis and proteinuria, renal failure, hepatosplenomegaly, anaemia, and amyloidosis. The ENL lesion shows features of either LL or BL histopathology depending on the clinical spectrum the patient belongs to and there is dense infiltration of the superficial and/or deep dermis and/or subcutaneous tissue by neutrophils. Vasculitis is a predominant feature in some cases. Severity of Type 2 Reaction may be of mild or severe type. Mild type is defined when the patient has only a few ENL skin lesions and no signs of involvement of other organs. Mild T2R can be managed with analgesics and anti-inflammatory drugs such as

aspirin and other NSAIDs. Aspirin is given in the dose of 600 mg 6 hourly with meals. Oral corticosteroids constitute the first line armamentarium in the management of severe T2R. They act by inhibiting both the early and late phases of inflammation. Corticosteroids decrease chemotaxis of neutrophils and inhibit the enzyme prostaglandin synthetase. Steroid administration is also associated with suppression of cell-mediated immunity by depletion of T-cells, particularly helper T-cells, with consequent restoration of altered helper/suppressor T-cells ratio and resultant decrease in the liberation of proinflammatory lymphokines. Prednisolone should be started in a dose of 1 mg/kg/day till clinical improvement, then tapered every week by 5–10 mg over 6–8 weeks. A maintenance dose of 20–30 mg may be needed for several weeks to prevent recurrence of ENL. No doubt, quick response to prednisolone is observed in most of the cases in the first attack of T2R. However,steroid dependence is a very important problem to handle. Tapering the dose of steroid is often associated with recurrence of reaction.[1]

CONCLUSION

Generally, ENL is seen at the beginning of UMDT institution for the treatment of Leprosy. Though there are case reports of late presentation of Lepra reactions but presentation of ENL occurring 10 years after treatment with MDT-MB is unusual. In our case, typical clinical and histopathological features helped us to diagnose and treat the case. Thus suspicion, early diagnosis & definitive long term treatment with glucocorticoid is essential to manage the reactions.

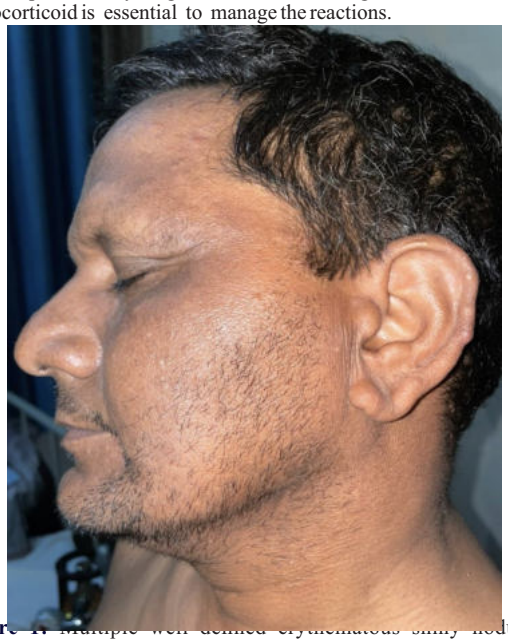

Figure 1: Multiple well defined erythematous shiny nodules approximately 0.5 cm in diameter along with diffuse infiltration present over the left ear. Lateral madarosis is present over the left eyebrow.

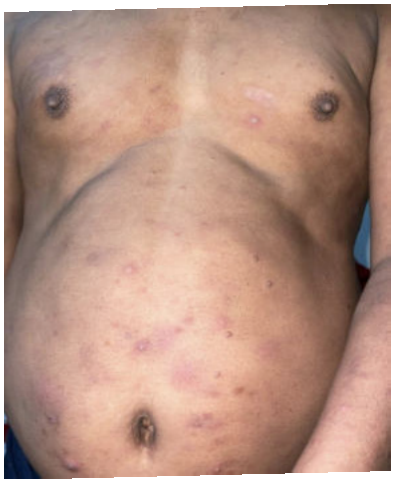

Figure 2: Multiple well defined dome-shaped erythematous shiny nodules along with surrounding erythema present over the abdomen and chest.

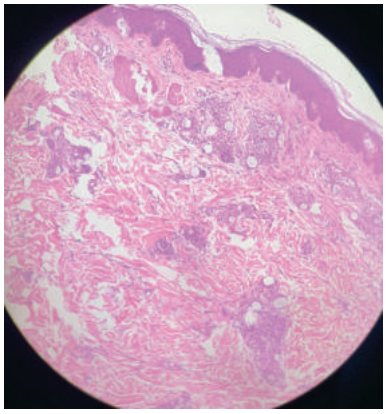

Figure3: Epidermis is separated by a narrow zone of collagen (Grenz zone). Multiple macrophages with vacuoles and globi present in the upper dermis. Dense neutrophilic infiltrate extending from the dermis up to the subcutis.

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