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Case Report

Rare Presentation of Type 2 Lepra Reaction with Suppurative Lymphadenitis and Pustular Sweets Syndrome-like Lesions

S Chauhan[1], GK Verma[2], L Negi[3], A Singh[4]

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The unusual presentations of lepra reactions (LRs) or leprosy often warrant the exclusion of close mimickers especially if cardinal signs of leprosy are lacking. In such cases, demonstration of acid-fast bacilli (AFB) either on slit skin smear microscopy (SSS) or histopathology or on cytology can aid in early diagnosis of leprosy. Here a 72-year-old untreated case of leprosy is reported for atypical presentation of type 2 lepra reaction (T2LR) in the form of massively enlarged, generalized lymphadenitis with secondary suppuration in groin and axillary groups. The patient also had fever, constitutional symptoms, sweets syndrome-like tender plaques, and pustules over the extensors of the extremities. Despite having visible deformities in her hands for the past 10 years, the patient denied any treatment for leprosy. The demonstration of AFB on lymph node cytology and subsequently on repeated SSS from ears confirmed the diagnosis of leprosy. After that other clinical possibilities like sweets syndrome with underlying lymphoma or concomitant tubercular lymphadenitis were excluded.

Keywords : Atypical, Type 2 Lepra Reaction, Pustular, Necrotic, Suppurative Lymphadenitis, Sweets Syndrome.

Introduction

Type 2 lepra reaction (T2LR) is an immune complex-mediated systemic inflammation with various unusual presentations. Erythema nodosum leprosum (ENL) is the most characteristic cutaneous manifestation of T2LR. Various atypical forms of ENL like vesicular, bullous, necrotic, and pustular have been described (Luo et al 2021). These atypical variants usually simulate other dermatoses and indicate severe cutaneous and systemic involvement.

Being a multisystem disease lymph node involvement is well-reported in leprosy and T2LR. Leprosy related lymphadenopathy is painless and involves the inguinal group in 76.2%, cervical and axillary in 69.5%, and axillary in 64.7% (Kiran et al 2009). Painful lymphadenitis is a feature of T2LR and occurs in about 15% of cases (Luo et al 2021). Generalized lymphadenopathy with secondary suppuration is a rare presentation of T2LR. We could find a single case report of suppurative lymphadenitis reported in March 2023 by Meena et al (2023). To enrich the literature on leprosy, we are reporting our experience.

Case report

A 72-year-old woman presented with multiple painful swellings with secondary suppuration in both groin and axilla for the past 5 months. She

[1] Dr Sandhya Chauhan, MD (Dermatology), Assistant Professor
[2] Dr Ghanshyam Kumar Verma, MD, Professor
[3] Dr Lalita Negi, MD (Pathology), Associate Professor
[4] Dr Aarti Singh, MBBS, Post graduate student (3rd year)
[1,2,4] Departments of Dermatology, Venereology and Leprosy & [3]Pathology, Indira Gandhi Medical College, Shimla- 171001 (HP), India
Corresponding Author: Dr Aarti Singh, **Email**: draartisinghderm1247@gmail.com

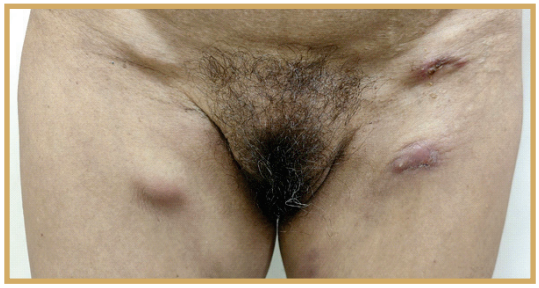

Fig. 1 : 72-year-old female of T2LR showing multiple enlarged, erythematous, fixed, and suppurative lymph nodes in the inguinal and femoral groups.

also had extremely painful multiple red raised and pustular lesions on her buttocks, upper and lower limbs. These complaints were associated with high-grade fever, severe constitutional symptoms, and pain in multiple joints. There were visible deformities in both hands for the past 9-10 years which the patient reported secondary to trauma. The patient denied completely any numbness or weakness in her hands.

On examination the patient was sick-looking, her temperature was 101°F, and her pulse rate was 120 beats per minute. She had multiple tender enlarged, non-matted but fixed cervical, axillary, and inguinal lymph nodes with secondary suppuration in axillary and inguinal groups (Figs 1 & 2). Cutaneous examination showed multiple well-defined, tender, erythematous papules, plaques, and pustules predominantly on her extremities with few lesions on the face, buttocks, and trunk (Figs 3 & 4). The classical lesions suggestive of leprosy like hypopigmented macules, plaques, nodules, or infiltration were absent (though a biopsy was sent from barely perceptible hypopigmented macules on the back). The peripheral nerves in both upper and lower limbs were symmetrically thickened, and temperature loss was demonstrated below the knees. But sensations were intact in deformed hands despite having motor weakness.

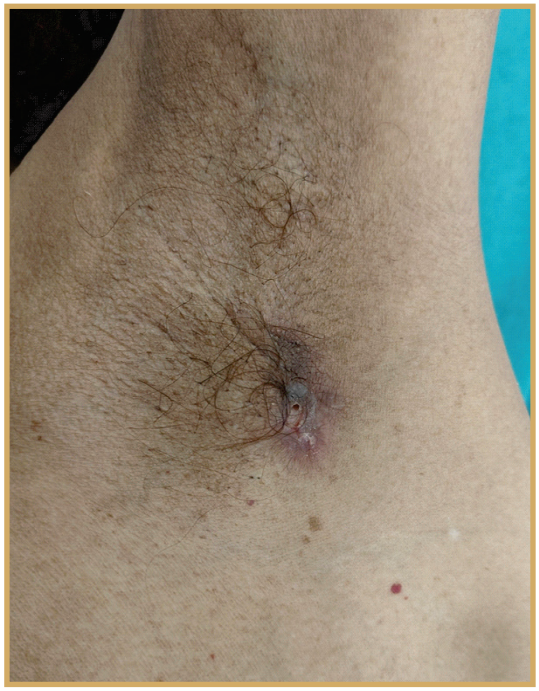

Fig. 2 : Left axilla showing suppurated lymph node with sinus formation and peri sinus erythema.

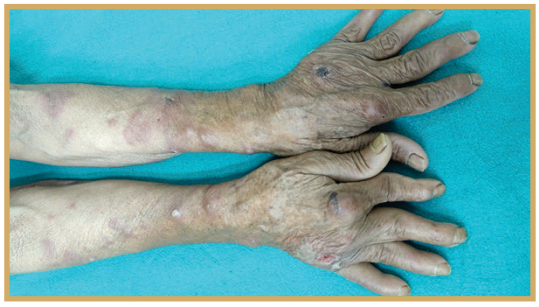

Fig. 3 : Extensors of forearms and dorsal hands revealing multiple erythematous plaques, with few pustular, ulcerative, and necrotic lesions. Flexion and claw hand deformities are also evident.

Baseline slit skin smear (SSS) from both ears was negative and histology from the hypopigmented (doubtful lesion) macule didn't show any evidence of leprosy. Nerve conduction studies revealed axonal sensory motor neuropathies in both upper and lower limbs. A clinical possibility

of pure neuritic leprosy (multibacillary type)

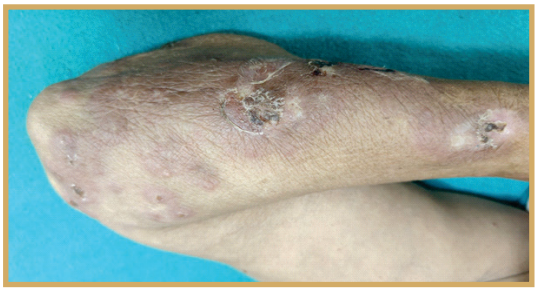

Fig. 4 : Flexors of right forearm revealing multiple erythematous and targetoid plaques of size 1*1 cm to 10*5 cm with central ulceration, necrosis, and healing by crusting.

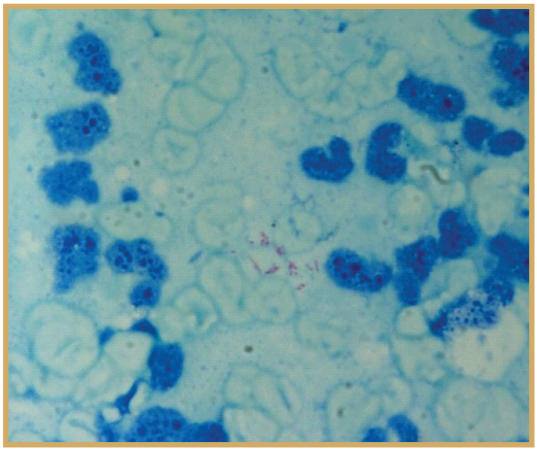

Fig. 5 : Acid-fast lepra bacilli on slit skin smears from ear lobule on 100x.

with pustular ENL was considered. Because of multiple enlarged lymph nodes, negative SSS, and non-specific histology; all imaging (X-rays, USG) and laboratory investigations were done to exclude underlying lymphoma, pustular sweets syndrome, and co-existing tubercular etiology.

The routine investigations revealed leukocytosis (18000/microlitre), a raised erythrocyte sedimentation rate (72 mm/h), and a C-reactive protein (140 mg/L). The patient had a negative Mantoux test, normal X-ray chest, negative ZN staining, and gene expert for tuberculosis from lymph node aspirate. The repeated SSS from both ears and modified ZN staining on multiple lymph nodes aspirate demonstrated foci of fragmented and granular acid-fast bacilli with BI 4+ and MI 0% (Fig. 5). After the demonstration of AFB in skin and lymph node aspiration other invasive investigations like nerve and lymph node biopsies were not done. The patient was diagnosed with lepromatous leprosy and severe T2LR with unusual features like suppurative lymphadenitis and pustular sweets like ENL. Indoor management was done with oral prednisolone in the dosage of 1 mg/kg/weight and injectable antibiotics (Amoxicillin with clavulanic acid 1.2 gm IV twice daily) for 10 days. After clinical improvement, patient was discharged on standard multi-drug therapy (MDT) and tapering doses of oral prednisolone.

Discussion

The cardinal signs of leprosy are sufficient to diagnose classical cases of disease, but unusual presentations often challenge the diagnosis. Leprosy has been a great mimicker to sarcoidosis, connective tissue diseases, Jessner's lymphocytic infiltrate, and even cutaneous lymphomas (Mahajan et al 2012). Bagla et al reported generalized lepromatous lymphadenitis masquerading as lymphoma in a 70-year-old male (Bagla et al 2005). Singh et al reported, two cases, who were initially diagnosed as tuberculosis and lymphoma, and later lymph node biopsy revealed the diagnosis of leprosy (Singh et al 1985).

Generalized lymphadenopathy with secondary suppuration as a presenting manifestation of leprosy is a rare observation and represents severe T2LR. Yang et al reported an unusual case of extensive lepromatous lymphadenitis with severe systemic manifestations. (Yang et al 2018). Recently Meena et al reported a case of lepromatous leprosy with suppurative lymphadenitis (Meena et al 2023). The present case had generalized lymphadenopathy, sweets syndrome-like lesions (painful plaques, nodules, and pustules), severe constitutional symptoms, and leucocytosis with raised ESR and CRP.

Despite strong suspicion of leprosy, lymphoma with paraneoplastic sweets syndrome and tuberculosis were close differentials. The visible deformities thickened peripheral nerves, painful lymphadenopathy, and co-existing pustular ENL were pointers towards leprosy with T2LR. In lymphoma and tuberculosis, lymphadenopathy is painless, and co-existing leprosy-specific skin lesions are not seen. In the present case, negative SSS (initially) and non-contributory histopathology warranted the exclusion of lymphoma and tuberculosis.

Approximately 15-50% of lepromatous leprosy (LL) patients experience ENL within one year of therapy (Vijendran et al 2014). In a case series by Vijendran et al, various atypical variants of ENL were described and one of the cases had a sweets syndrome-like presentation (Vijendran et al 2014). Kou and Chan (1987) also reported a patient of ENL simulating Sweet's syndrome.

The atypical variants of ENL indicate severe T2LR and systemic involvement. Sepsis and even fatality due to secondary infection have been reported with ulcerative/necrotic ENL. Davis et al (2002) reported a fatality. Vijendra et al (2014) reported sepsis secondary to ulcerative/ necrotic ENL. The present case had severe constitutional and systemic symptoms with fever, lymphadenitis, arthritis, leucocytosis, and raised acute phase reactants. Apart from MDT and high doses of oral steroids, broad-spectrum parenteral antibiotics are recommended for management. Early diagnosis and prompt management can prevent sepsis-related morbidities and mortality in patients with pustular, ulcerative, and necrotic ENL.

Conclusion

The clinical diagnosis of leprosy is not difficult but atypical presentations may indicate severe disease and warrant exclusion of close mimickers. Awareness of atypical variants such as presented in this report and their prompt management is essential to prevent mortality and morbidity in leprosy patients.

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