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Erythema Necroticans- A Rare Presentation of Lepra Reaction A Report of two Cases

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Abstract
Leprosy is chronic inflammatory disease caused by Mycobacterium leprae, affecting skin, peripheral nerves and other tissues. During long course of disease, immunological alterations occur in the form of reactions which result in sudden increase in activity of disease.
Here we present two cases, in which one case directly presented with Erythema Necroticans without preceeding symptoms of Leprosy and the other is known case of Borderline lepromatous leprosy with Erythema Necroticans.
Keywords: *Erythema necroticans, Lepra bacilli, Fite faraco stain.*

Introduction

Leprosy reactions are immunologically mediated episodes of acute or subacute inflammation which interrupt the relatively uneventful usual chronic course of the disease affecting skin, mucous membrane and / or other sites.[1] Leprosy reactions include Type 1 reaction, Type 2 reaction or Erythema Nodosum Leprosum and Lucio phenomenon.

Erythema Nodosum Leprosum is a type 3 immune complex mediated hypersensitivity reactions characterized by sudden appearance of crops of evanescent, pink colored tender papules, nodules or plaques variable in size associated with constitutional symptoms. Erythema Necroticans is a rare severe form of ENL in which lesions become vesicular, pustular, bullous and necrotic and break down to produce ulceration called as Erythema Nodosum Necroticans.[1]

Case Report

Case 1

A 49 yr old male patient, a known epileptic since 15 years on phenytoin and carbamazepine had a trauma on left little toe, for which he consulted a quack and his left little toe was amputated and was given some injectables, following which he developed multiple ulcerations and raw areas associated with mild fever, there was no history of hypopigmented or erythematous lesions, epistaxis, joint pains, slippage of footwear. Initially diagnosis of drug reaction was made and was admitted. On examination general condition was normal, vitals were normal, pedal edema and

puffiness of face was present. There were multiple large ulcers of varying sizes, with sloping edge, floor coverd with slough, few ulcers with crusts, few healed with scarring are distributed relatively symmetrically over both ear lobes, extensor aspects of fore arm, buttocks, scrotum and over legs. There is decreased sensation over both lower limbs, and over lesions. Investigations like complete blood picture, liver and renal function tests were within normal limits.

Slit skin smear was negative from the ulcer and from both ear lobes. Biopsy was taken from ulcer edge, Histopathological examination showed epidermis with areas of crusting, orthokeratosis, hypergranulosis, moderate acanthosis and intact basal layer. The superficial and deep dermis showed sparse interstitial and perivascular lymphocytic infiltrate, the subcutis showed foamy macrophages and lymphocytes. Fite faraco stain revealed abundant solid stained and fragmented lepra bacilli.

Patient was diagnosed as a case of Lepromatous leprosy with Erythema nodosum necroticans and was started on Multi bacillary Multi drug therapy and Injection Dexamethasone, Non steroidal anti inflammatory drugs and supplements.

Figure 1.1: Ulcers covered with slough and crusts distributed over both ear lobes (A, B) over extensor aspect of arm and forearm (C) over lower limb (D)

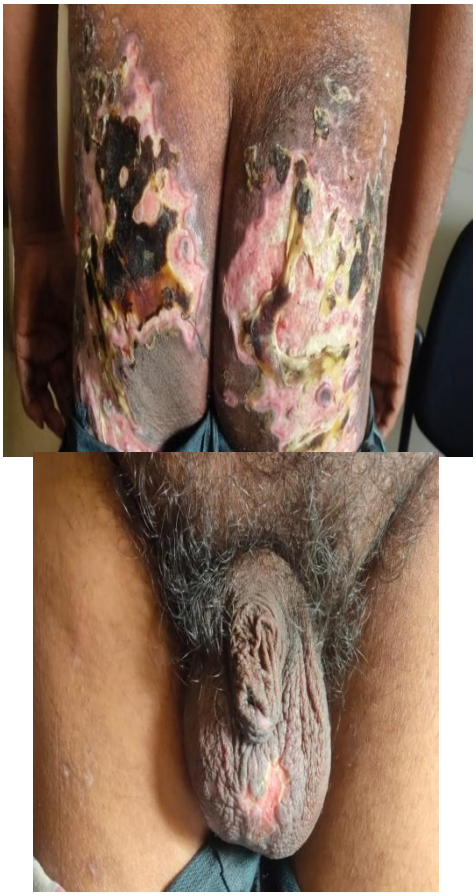

Figure 1.2: Ulcers involving both buttocks and scrotum.

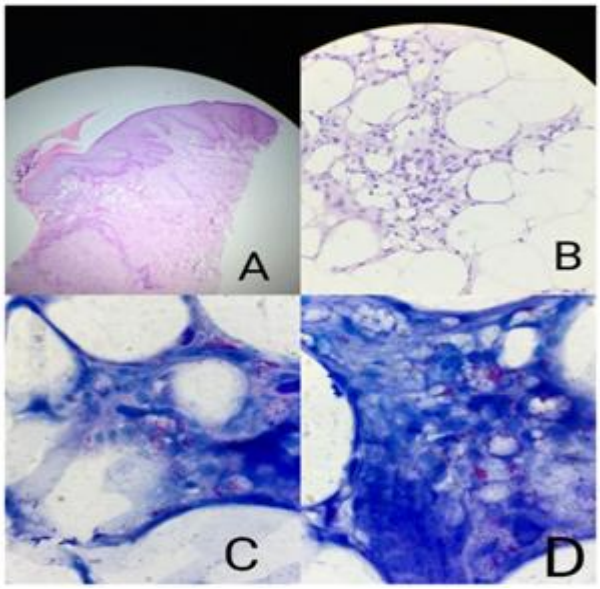

Figure 1.3: Histopathology

A)10X image showing normal epidermis and dermis.

B)40X image showing lymphocytes and foamy macrophages

C & D – Fite faraco stain showing solid and fragmented bacilli.

Case- 2

A 16 year old female, known case of Borderline Lepromatous Leprosy, Used multibacillary multidrug therapy for 8 months complaints of fever and ulcerated lesions over face since 1 week. On dermatological examinated multiple hemorrhagic fluid filled blisters were present over ear lobes, hyperpigmented crusts over cheeks and forearm and raw areas are present over forearms, diffuse hyper pigmentation over soles of feet. All the investigations like complete blood picture, liver and renal function tests were within normal limits. Slit Skin smear reveals multiple bacilli with bacillary index of 6+. Biopsy reveals ill defined macrophage granuloma. Case was diagnosed as Erythema Necroticans.

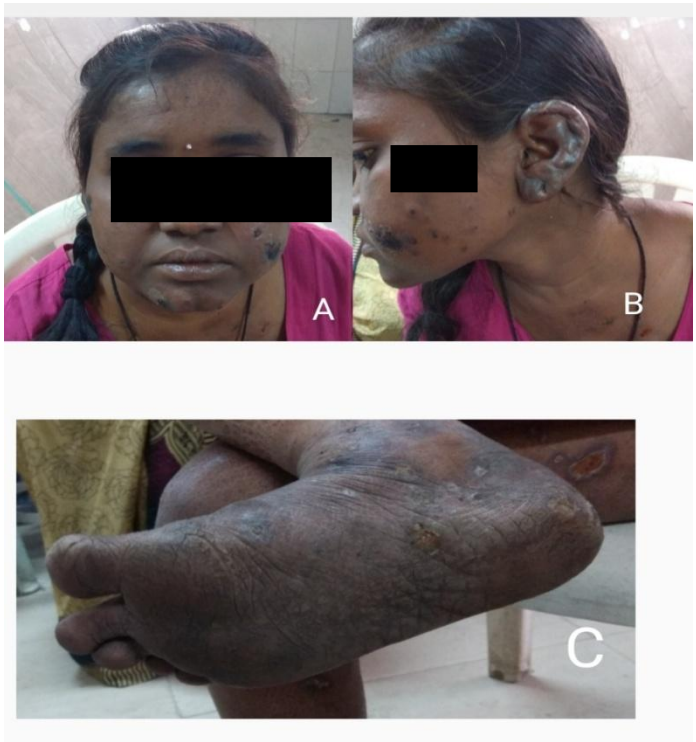

Figure 2.1

A- Blackish crusts over face.
B- Hemorrhagic bullae over ear lobe
C- Blackish pigmentation over sole of foot

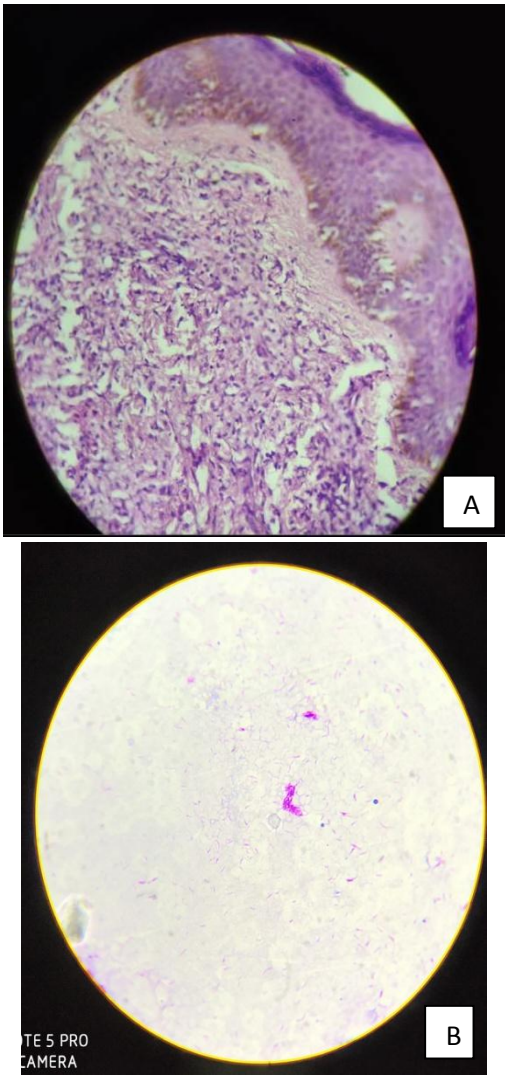

Figure 2.2

A-Ill defined macrophage granuloma.
B- Slit skin smear showing 6+

Discussion

Erythema Nodosum Leprosum is an immune complex syndrome causing inflammation of skin, nerves, other organs and generally malaise. It occurs mostly in Lepromatous leprosy and sometimes in Borderline lepromatous leprosy. Antigen-antibody complexes are formed during the treatment in multibacillary leprosy patients and in patients with longstanding untreated disease with high bacillary load due to the death of bacilli, deposition of these complexes in various tissues evoked inflammatory response with constitutional symptoms.

Risk factors for ENL include lepromatous leprosy with skin infiltration , bacillary index>4+, patients less than 40 yrs age, intercurrent infections,

trauma, Surgical intervention, Physical and mental stress, strongly positive mantoux test, pregnancy, parturition and ingestion of potassium iodide.

Erythema Necroticans is a severe form of ENL first reported by Verma and Pandhi[2] in 1993 where the lesions are vesicular, bullous and pustular which become necrotic and later ulcerate.

Literature search revealed a case report by N. Al Hayki and B. Al-Mahmoud where Erythema necroticans is presenting manifestation of subtle borderline lepromatous Leprosy[3] and inanother report by Verma and Pandhi[2] it is the presenting manifestation of Lepromatous leprosy, which is similar to our case no 1, where patient had no previous history of leprosy and directly presented with erythema necroticans . Surgical stress might be a precipitating factor in this case, also there is involvement of scrotum which is considered as relatively immune zone in Leprosy.

T S Rajashekar et al[4] reported certain zones such as scalp, palms and soles, genitalia, groins, axillae, eyelids, transverse band of skin over lumboscaral area, and midline of back and perineum have been described to be relatively immune to the development of lesions in leprosy which is attributed to their relatively high local temperature. Scrotal skin has been reported to be relatively cooler than the core temperature for effective spermatogenesis. However, due to the use of heavy undergarments, it is likely that the temperature of the scrotal skin may remain elevated.[5] Thappa DM et al [6] reported scrotal skin lesions in Lepromatous leprosy.

Biopsy taken from ulcer edge had no epidermal and dermal changes, that is no granulomas or foamy macrophages or peri appendageal infiltrates, but subcutaneous tissue revealed few foamy macrophages and lymphocytes. The case was diagnosed based on Fite faraco stain which showed multiple solid stained and fragmented bacilli in subcutaneous tissue. Such histopathological findings of leprosy without characterstic epidermal or dermal changes are unusual and to best of our knowledge they were not earlier reported in the literature. Skin lesions improved rapidly after MB MDT.

Our second patient is a known case of Borderline lepromatous leprosy presenting with ulcers, bullae and black colored crusts, there is pigmentation over soles of feet which may be due to clofazamine. The involvement of palmo plantar skin was reported by **Indira et al**[7] in which they screened 280 patients of leprosy and found the palmoplantar involvement in 10% of the cases. Lesions occurred more frequently in borderline leprosy than in lepromatous leprosy. Palms and soles are included in relatively immune zones because the thick epidermis of palms and soles along with the good amount of fibro fatty tissue provides an insulating property and hence a high nerve bed temperature, which renders the palmoplantar localization of Mycobacterium leprae less likely[8].

Similar involvement of palms in the case of BT Hansens was reported by **Yasmeen Jabeen Bhat**[9]**.**

Conclusion

These cases where reported because of rarity of this condition and it occurred as a presenting manifestation which is uncommon and involvement of relatively immune zones like scrotum and soles.

Source of Support: Nil

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