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Case Report

Necrotic Erythema Nodosum Leprosum : A Case Report

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Erythema nodosum leprosum (ENL), the type-2 reaction in leprosy, is caused by a humoral immune response to *Mycobacterium leprae*. Severe ENL can lead to vesicular or bullous manifestations known as necrotic erythema nodosum leprosum (ENL). Despite the classic presentation of erythema nodosum, severe ENL very rarely presents as an ulcerative skin lesion. Necrotizing ENL is a rare manifestation, characterized by vesicular or bullous lesions resulting in necrotic ulcers preceded by constitutional symptoms of fever. Our patient was diagnosed with leprosy 19 years ago, was irregular in taking treatment and presented swelling in the lower leg with skin color changed to brownish purple. Based on clinical and histopathology the patient was diagnosed as necrotic ENL and treated with MDT and methylprednisolone. Patient consumed the medication for 3 weeks, the lesions turned into necrotic tissue, however, the patient died in the third week due to septic bacteremia. Atypical ENL manifestations require attention in diagnosis to start timely appropriate management.

Key words : Erythema Nodosum Leprosum (ENL), Necrotic ENL

Introduction

Necrotic erythema nodosum leprosum is an uncommon manifestation of type 2 reaction, which is found in cases of lepromatous and borderline lepromatous leprosy. Severe ENL can present with vesicular or bullous manifestations, which then burst and are called necrotizing erythema. Extracutaneous manifestations include neuritis, iridocyclitis, orchitis, and lymphadenopathy. Fever and other constitutional symptoms are usually associated with it (Dhillon et al 2015).

Cases of necrotic ENL lesions have been reported previously from Mexico and South America. Several reports of necrotic and bullous ENL have also been published from India (Barman et al 2015).

Careful attention is essential in establishing a clinical diagnosis of necrotic ENL because it is a variant that is relatively rare and can lead to complications and a high level of morbidity.

Case Report

A 27 years old male came with complaints of swelling on the face, hands, and feet since the previous day. Initially, the swelling was felt in the lower leg area, followed by the hands and face.

These complaints of swelling were accompanied

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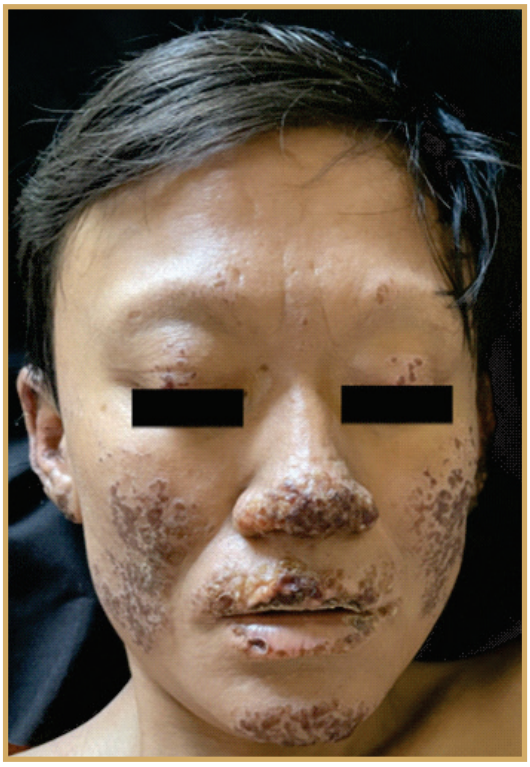

Fig. 1 : The dermatological examination of the facial region.

by pain with and knee joint pain. In the area of swelling, the skin color changed to brownish purple. Patient also complained of appearance lump on the lower right leg since the previous day, which measured about 3 cm in size and was not affected by pain. He was subsequently diagnosed with leprosy 19 years ago with symptoms of wounds and the shortening of fingers on his both hands. He then received Multi-drug therapy (MDT) but did not consume it regularly.

On general examination patient was febrile with 38.9°C. Dermatological examination of the facial region revealed multiple hyperpigmentation plaques with crusted, saddle nose and madarosis (Fig. 1).

There were multiple plaques, purplish brown vesicles, and bullae throughout the observation for 3 weeks, the lesion became a necrotic ulcer on the manus (Fig. 2) and pedis region (Fig. 3).

The Erythema Nodosum Leprosum International Study (ENLIST) assessment score was 19. Palpation of the peripheral nerves revealed enlargement of the right and left common peroneal nerves.

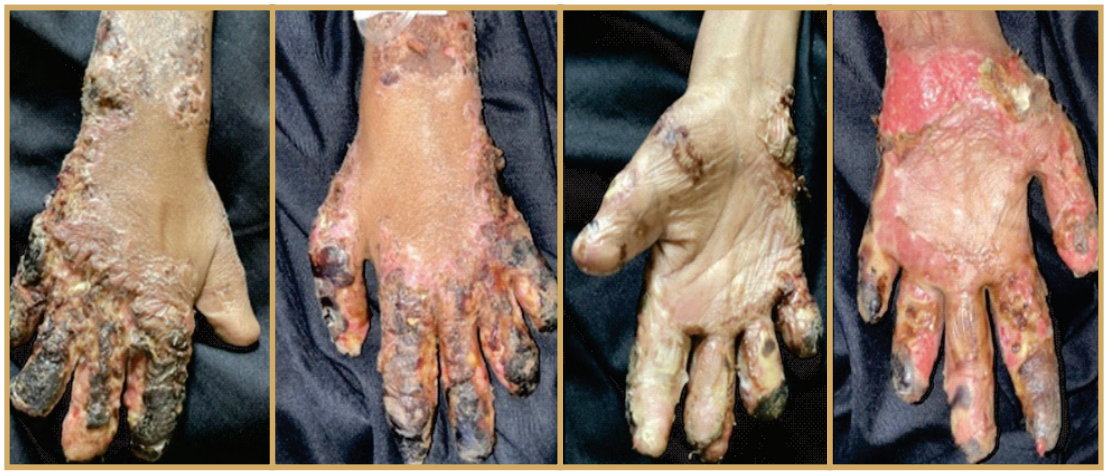

Fig. 2 : The dermatological examination of the manus region.

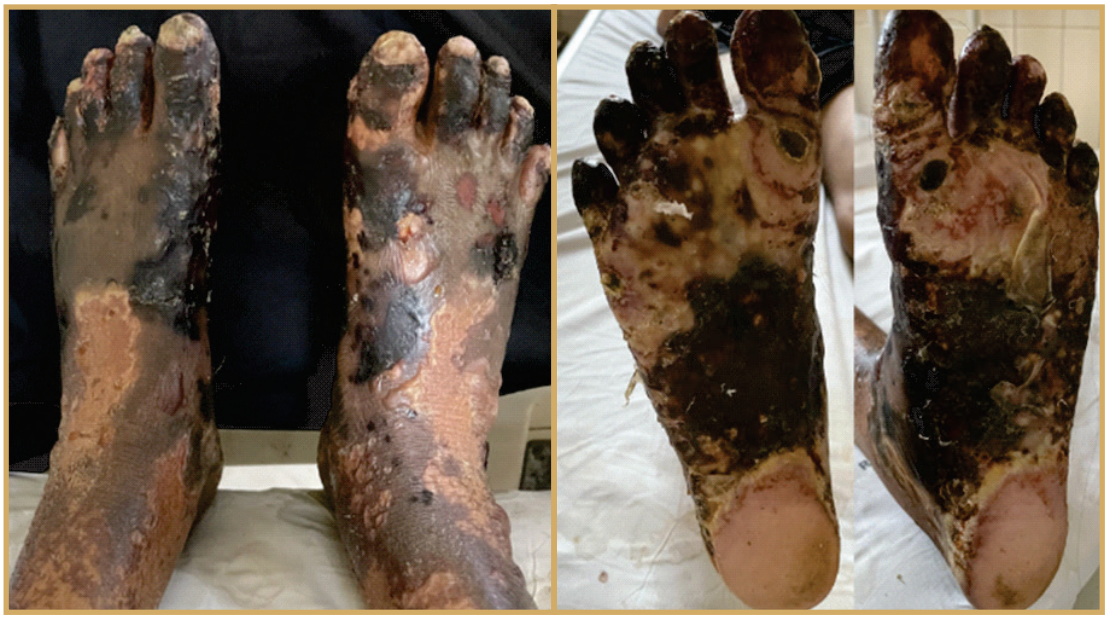

Fig. 3 : The dermatological examination of the pedis region.

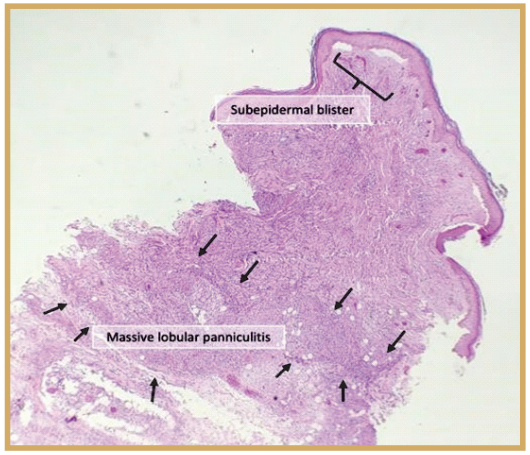

Fig. 4 : The skin tissue is layered with the epidermis and shows a granulomatous reaction (HE, 40x).

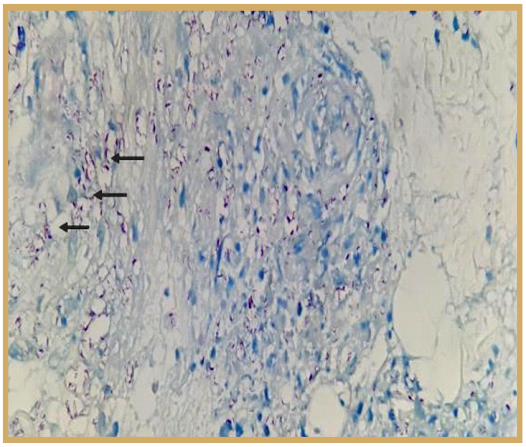

Fig. 5 : Acid-fast bacilli, stained red, which are located between the tissues (ZN x100).

Based on necrotic erythema lesion and histopathological findings the patient was diagnosed with necrotic erythema nodosum leprosum, treated with multibacillary multidrug therapy (MDT), which includes oral methylprednisolone 16 mg in the morning, 16 mg in the afternoon. Patient could consume medicines for 3 weeks, after that the lesions turned into necrotic tissue, the patient died in the third week due to septic bacteremia.

Patient underwent an abdominal ultrasound examination because laboratory showed renal azotemia and revealed bilateral parenchymatous renal disease and mild splenomegaly. Establishing the diagnosis and ruling out the etiology of vascular disorders, the patient underwent a radiological examination of duplex ultrasonography (DUS) on the superior and inferior extremities. There were no signs and symptoms of acute limb ischemia, acute heart failure, or cardiovascular emergencies.

On the punch biopsy examination taken from a sample of the edge of the bullae in a pedis ulcer. The results showed that the skin tissue was covered by epidermis which showed focal subepidermal blister. Dermis show intense inflammatory infiltrates consists of foamy histiocytes and neutrophils which extend to adjacent sub-cutaneous adipocytic tissue forming massive lobular panniculitis (Fig. 4).

Discussion

Vasculonecrotic erythema nodosum occurs in cases of multibacillary LL and BL after starting multibacillary multidrug therapy; the ulcer heals with a fibrotic scar. The clinical features of ENL and LP can be very similar, and cases of overlap of the two have also been published (Benjith et al 2023, Ranugha et al 2013).

Severe ENL can become vesicular or bullous and then rupture, referred to as necrotic erythema. Ulcers are a rare symptom of leprosy resulting from ENL (rare), loss of sensation (peripheral), or Lucio phenomenon. Despite the classic presentation of erythema nodosum, severe ENL rarely presents as an ulcerative skin lesion (necrotic erythema). Lucio's phenomenon is similar to necrotic erythema but without fever or constitutional symptoms. Histologically, LP shows epidermal necrosis, the presence of dense or granular bacilli in the endothelial cells, and the proliferation of endothelial cells; the superficial dermis may show leukocytoclastic vasculitis, dermal necrosis, and mild mononuclear infiltration. Necrotic ENL is characterized by the presence of granular/ fragmented bacilli in macrophages (a sign of a treatment-induced reaction), foamy histiocytes, and dermal neutrophil infiltration, along with pan-vasculitis, more so in the hypodermis and panniculitis (Dhillon et al 2015, Bhattacharjee et al 2020).

To conclude atypical ENL manifestations of leprosy and its complications require attention in diagnosis to determine appropriate management.

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