Livedo reticularis in type 2 lepra reaction: A rare presentation

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ABSTRACT

Type 2 lepra reaction or erythema nodosum leprosum (ENL) is an immune complex syndrome that occurs mostly in lepromatous leprosy and sometimes in borderline lepromatous leprosy patients. We present an untreated case of lepromatous leprosy, who presented with type 2 lepra reaction and livedo reticularis. Livedo reticularis, though seen in lucio phenomenon, is not a part of type 2 lepra reaction. The case is being reported for its rarity.

Key words: Lepromatous leprosy, livedo reticularis, type 2 lepra reaction

INTRODUCTION

Type 2 lepra reaction or erythema nodosum leprosum (ENL) is an immune complex syndrome that occurs mostly in lepromatous leprosy and sometimes in borderline lepromatous leprosy patients. It generally occurs in patients on treatment but may be encountered in untreated cases. It is an example of type III hypersensitivity reaction or Arthus phenomenon. The deposition of circulating or extravascular immune complexes in various tissues is responsible for the manifestations of type 2 reactions.[1,2]

We present an untreated case of lepromatous leprosy, who presented with type 2 lepra reaction and livedo reticularis. Livedo reticularis, though seen in lucio phenomenon, is not a part of type 2 lepra reaction. The case is being reported for its rarity.

CASE REPORT

A 40-year-old female presented with hyperpigmented macules on the hands since 2 months. It started on palms and gradually progressed to involve hands, forearms and neck. The lesions were asymptomatic except for mild burning sensation on exposure to sunlight. She had noticed loss of eyebrows since 1 month. She did not complain of fever or joint pains. There was no other significant history.

General physical and systemic examination revealed no abnormality. Reticulate hyperpigmented macules with areas of atrophy were seen over the neck, back, medial aspect of forearms, lower limbs, palms, and soles (Figures 1 and 2). Supraciliary madarosis was present. Skin biopsy results were awaited.

The patient presented with new painful, reddish lesions all over the body after 20 days. Examination revealed multiple, tender, erythematous papules and nodules over face, neck, chest, back, and both upper and lower extremities. Livedo reticularis was present on the back, abdomen and lower limbs (Figures 3 and 4). There was no nerve thickening or sensory loss. A diagnosis of lepromatous leprosy with type 2 reaction was made and a repeat biopsy was taken from the back. Routine blood investigations such as complete hemogram, thyroid profile, liver function tests, renal function tests, venereal disease research laboratory (VDRL) test, and HIV were within normal limits. She was positive for HbsAg. The initial biopsy report revealed thinned out epidermis lined by keratinized stratified squamous epithelium. Subepidermal clear zone was seen. Dermis showed sheets of foam cells, histiocytes along with lymphoplasmacytic infiltrate extending up to the reticular dermis. These sheets of foam cells were seen in perivasculary as well as periadenexal region (Figure 5). Fite-Farraco stain for lepra bacilli was positive. The biopsy findings...
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suggested the diagnosis of lepromatous leprosy. The second biopsy report showed epidermal atrophy with plenty of foamy macrophages and mononuclear cells along with occasional fragmented neutrophils in the upper dermis [Figure 6]. The Fite-Farraco stain revealed plenty of acid fast bacilli [Figure 7]. The second biopsy report confirmed type 2 lepra reaction.

**DISCUSSION**

Livedo reticularis is a reticular erythematous violaceous discoloration of the skin that typically affects the limbs, although it can be generalized. It is secondary to organic or functional disorders of the dermal arteries or arterioles. Lucio phenomenon is one of the causes for livedo reticularis which causes vessel inflammation, thereby leading to reticular discoloration of the skin.[3]

Lucio leprosy is characterized by diffuse skin infiltration, madarosis, loss of facial skin creases, and absence of papules and nodular lesions.[2] Lucio’s phenomenon is an acute event in the evolution of Lucio’s leprosy, characterized by the onset of erythematous violaceous maculae and infiltrated plaques that progress with central necrosis followed by ulceration. They frequently occur in patients who were never treated or in those who have followed treatment irregularly. According to the international literature, three criteria are adopted for Lucio phenomenon: cutaneous ulceration, vascular thrombosis, and invasion of blood vessel walls with lepra bacilli.[4] In the present case, the patient presented for the macular pigmentation of hands and neck with madarosis and after 15 days she presented with livedo reticularis, fever, malaise, and arthralgia. Though the presentation is typical for Lucio phenomenon; absence of

![Figure 1: Reticular pigmentation of the neck before reaction](image1)

![Figure 2: Hyperpigmented macules over palms before reaction](image2)

![Figure 3: Livedo reticularis over back during reaction](image3)

![Figure 4: Livedo reticularis over lower limbs during reaction](image4)
In livedo reticularis, the biopsy specimen taken from the erythematous area may be normal. In our case, as the biopsy was taken from the erythematous area, there were no features of livedo reticularis, like thickened vessel wall and occlusion by the thrombus.

There were no features suggestive of connective tissue disorder in the present case. The patient did not agree for antinuclear antibody assay. Though there were no features of connective tissue disorders, coexistence of such conditions cannot be ruled out.

This case has been presented for its unique manifestation as livedo reticularis in type 2 lepra reaction in an untreated case of lepromatous leprosy.

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