A case of ulcerative leprosy? Lucio's phenomenon

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Introduction

Reactions in leprosy are due to heightened immunologic response of the host to *Mycobacterium leprae* or its breakdown products. Type 2 lepra reaction is characterized by Erythema Nodosum Leprosum (ENL) and multisystem involvement. ENL manifests commonly as painful erythematous, indurated nodules. Sometimes ENL lesions can be papular, plaque, vesicular, bullous or rarely ulcerative[1].

Lucio's phenomenon is a rare syndrome reported mainly in Mexico and Central America and occasionally reported from a few other countries including the USA, India and Singapore[2]. It is also a type 2 reaction restricted to patients with diffuse non nodular lepromatous leprosy.

Case report

Our patient is a 48-year old female beggar with congenital motor weakness of both lower limbs. She was admitted to NCTH Ragama with a two month history of leg ulcers. These were painless and progressive. Over the last two weeks, the ulcers had progressed rapidly and new ulcers had appeared over the buttocks upper arms and face. Some of the ulcers started as blisters while the others began as purpuric macules.

On examination, she was moderately pale, afebrile and there was no lymphadenopathy. There were extensive deep and irregular ulcerations over the face with involvement of the tip of the nose and nasal mucosa and the ear lobes (Figure 1). There was nodularity in the ear lobes and infiltrated plaques over the forehead. Skin was thickened with a diffuse infiltration of skin in between the ulcers. There was loss of eye brows and lashes. There were similar ulcers over the legs, buttocks and upper arms. However nodular or plaque lesions were absent (Figure 2). Distal extremities were oedematous and there was a glove and stocking type sensory impairment. There was no significant nerve thickening or tenderness.

Slit skin smear from the ear lobe showed numerous acid-fast bacilli with a morphological index of +5. A skin biopsy from the upper arm showed an area of ulceration with underlying acute inflammation and necrosis. There was a diffuse infiltrate of histiocytes, foamy macrophages and scattered lymphocytes in the dermis. The subcutaneous fat shows septal inflammation. A few blood vessels showed leukocytoclastic vasculiis. On Wade-Fite stain, numerous acid-fast bacilli were seen in and around the endothelial cells of superficial and deep blood vessels. Full blood count showed anaemia with Hb-7.6 g/dl and MCV 67.4 fl. White cells and platelets were normal.

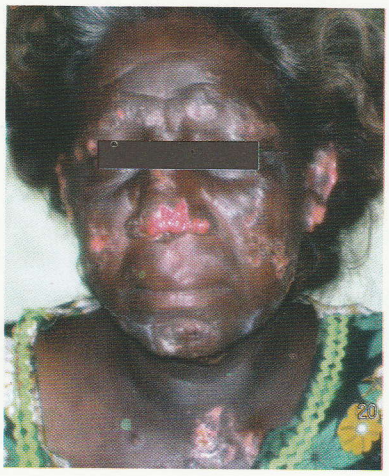

Figure 1

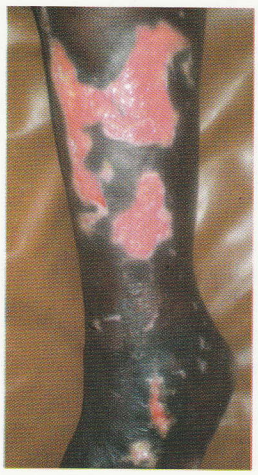

Figure 2

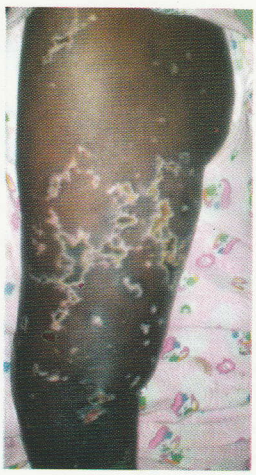

Figure 3

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The patient was started on multibacillary multidrug therapy (MDT-MB) together with oral prednisolone 40 mg daily, clofazamine 200 mg daily and intravenous penicillin and cloxacillin. She was given haematinics and gastric and bone protective measures were taken. The patient responded satisfactorily to treatment with skin lesions healing to form retiform atrophic scars (Figure 3).

Discussion

Ulcerative lesions are seen in leprosy, but they are rare. Ulcerations could result from severe ENL reaction or sometimes in severe type 1 lepra reaction. When there is extensive ulceration in leprosy, the possibility of Lucio's phenomenon should be considered.

The main difference between Lucio's phenomenon and ENL reaction is that the former is an ulcerative reaction occurring in the absence of cutaneous nodules, whereas the latter may occur in any type of lepromatous leprosy[3]. ENL usually presents as tender nodules that can sometimes ulcerate.

Lesions of Lucio's reaction usually begin as painful purpuric macules, plaques or vesicles which often ulcerate and heal with atrophic scars. The lesions are typically located over the extremities but may also affect the buttocks and trunk. Features of underlying lepromatous leprosy commonly described include diffuse thickening of facial skin, loss of eye brows and lashes, distal anaesthesia and destructive rhinitis. Systemic upsets are variable and may occasionally be fatal. However, unlike ENL it is not associated with fever or other manifestations like arthritis, neuritis, iridocyclitis or leukocytosis.

Histological findings in Lucio's reaction include leukocytoclastic vasculitis, endothelial proliferation and thrombus formation in dermal or subcutaneous vessels, with a sparse lymphocytic infiltration. Therefore the ulceration in Lucio's reaction is due to cutaneous infarction secondary to a necrotizing vasculitis. This is due to the deposition of immunocomplexes in dermal blood vessels and direct invasion of endothelial cells by the bacteria[4].

It is assumed that severely depressed cell mediated immune response permit the wide dissemination of the bacilli, which even reaches endothelial cells. The rapid and massive bacillary death resulting in the release of bacillary antigen causes overproduction of specific immunoglobulins which lead to immuncomplex formation.

Although the response to treatment has been reported as poor, especially with severe systemic upset, corticosteroids have a good effect. Thalidomide has no value. Pentoxyfilline has the ability to improve the regional microcirculation especially in ischemic areas and is therefore useful in the treatment of Lucio's phenomenon.

Many of the clinical and histological features in our patient, as mentioned earlier, are compatible with the diagnosis of Lucio's phenomenon. However the presence of nodularity in the earlobe is not suggestive of this condition.

References

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