Erythema Nodosum Leprosum Associated with Lepromatous Leprosy: A Case Report and Review of Literature

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ABSTRACT

Hansen disease or leprosy is an ancient disease that is still present with us and could masquerade and pose diagnostic challenges. Clinical presentation of Hansen disease is variable from the chronic form of the diseases which are easily recognised to the acute reactions that occur following perturbation of the chronic phase. Erythema nodosum leprosum is commonly seen in patients with borderline lepromatous leprosy or lepromatous leprosyas in the present case.We report a case of erythema nodosum leprosumthat presented in association with lepromatous leprosy at the LAUTECH Teaching Hospital,Ogbomoso.This patient was initially misdiagnosed in a comprehensive health center as a case of acne vulgaris and was treated with oral antibiotics and cleansers without improvement. The patient responded to a combination of Dapsone, Clofazimine, Rifampicin, and Prednisolone. The patient has remained well in the last four years without any form of relapse.

INTRODUCTION

There is no other communicable disease in human with several clinical pictures as leprosy[1]. Leprosy is a chronic granulomatous communicable disease caused by *Mycobacterium lepra* or *Mycobacterium lepromatosis*[2]. The chronicity of leprosy could be perturbed with different forms of acute exacerbations referred to as lepra reactions[3]. The Type 2 lepra reactions also known as erythema nodosum leprosum (ENL) is an acute inflammatory reaction that manifests as erythematous, painful papules and nodules in the extremities of patients with lepromatous leprosy (LL) or occasionally in borderline lepromatous leprosy (BLL) [3]. Frequently, ENL occurs while patients are on multidrug therapy (MDT) and reactions could occur at any point in the course of treatment, even up to one year after initiating MDT[3].

The ENL could be acute, subacute or chronic[4]. Acute forms of ENL occurring in a new patient, not exposed to MDT could be easily missed if there is no high suspicion for leprosy. In the literature, ENL varies from mild cases that spontaneously resolve to the severe forms of the reaction, that ulcerate and then heal with scarring. The various presentations of ENL in the form of vesiculobullous, pustular, ulcerated, hemorrhagic and erythema multiform-like reactions are a major diagnostic concern in clinical practice[5-8]. Rarely, ENL may be the first presenting manifestation of lepromatous leprosy as in the case under discussion[9]. We, therefore, present a classical form of ENL that presented to the clinic for the first time in association with lepromatous leprosy.

CASE REPORT

A 17-year-old female undergraduate complained of painful recurrent raised rash in the face, and extremities and some yellowish discolorations of her skin over the past 15 months. She was not on any medication before the appearance of the eruption. She has had about 2-3 previous episodes. The most recent episode started about two weeks before

presentation. There was no history of fever, pruritus, gum bleeding, body weakness, multiple sexual partners or contact with individuals with a similar rash. There was a positive history of the feeling of numbness on some of the discolored skin area but no history of paralysis of any limb, no limb ulcers or previous hospital admission or surgery or blood transfusion. There were new crops of painful and shining papules and nodules all over her body including the face, the ears, nose, and extremities. Some of the nodules later contained pus before they disappeared. The patient had no hoarseness or stridor but had a history of stuffy nose and occasional epistaxis. Her vision was clear and no eye pain or photophobia. She was not a known patient with asthma, hypertension or diabetes and no family history of similar rash. She presented to a comprehensive health center where she received a diagnosis of acne vulgaris and was treated with oral antibiotics and facial cleanser without improvement.

Examination of the skin showed symmetrically distributed erythematous and painful papules, nodules on the face with infiltration of the lip margins, nose, ears, and the chin. There were painful erythematous plaques infiltration and nodules on the arms, forearm, thighs, and shins. No visible limb deformity, or nerve thickening, ulceration or madarosis. There was background post inflammatory hyperpigmented macules and patches that suggested previously resolved lesions, Figure 1-3.

Her cardiovascular system showed regular pulse rate of 88 beats per minute; with normal volume and no thickened arterial wall or locomotor brachialis. Her blood pressure was 96/64mmHg with no displaced apex beat, no thrill or murmur. Heart sounds one, and two only were heard. Her abdomen was full and moved with respiration. No area of tenderness, no organ enlargement or ascites. The patient had normal active bowel sound. Other systems were essentially normal. The complete blood count, electrolytes, urea and creatinine, abdominal ultrasound, urinalysis were essentially normal. Microscopic histology of the skin showed dermis diffusely infiltrated by macrophages with large vesicular nuclei and granular cytoplasm. Some of the macrophages have vacuolated cytoplasm and within the macrophages are seen numerous *Mycobacterium leprae* microorganisms, Figure 4-6. The macrophages and *M. leprae* were seen surrounding some adnexal skin structures. A diagnosis of lepromatous leprosy with ENL (type 2 reaction) was made. The patient was commenced on Dapsone, Clofazimine, Rifampicin, and Prednisolone. Patient's condition improved on treatment and presently on follow-up.

DISCUSSION

The ENL is a type 3 Coomb and Gell hypersensitivity reaction usually seen in lepromatous and borderline lepromatous patients with high bacterial load[1,4,6,7]. Erythema nodosum leprosum could occur anytime, but it commonly occurs later during treatment and in the long-standing untreatedpatient[1,5,6,7]. Our patient remained untreated for a long time because she was misdiagnosed and treated in community health center as a case of acne vulgaris with facial cleanser and antibiotics.

It is known that antigen-antibody complexes are formed during the treatment in multibacillary leprosy and those long-standing untreated cases that have high bacillary load due to the death of bacilli; deposition of these complexes in varioustissues causes an inflammatory response with different symptoms[1, 6, 8]. More than half of lepromatous leprosy and about 25% of borderline lepromatous patients can develop Type 2 reactions[8]. Characteristic changes in the skin of affected people include the appearance of crops of multiple red to dark brown, painful papules and nodules over the face commonly and predominantly on the extensor surface of the limbs[1,2,7,8]. The trunk is usually sparingly affected[11] as also noted in the present patient. The affectation of peripheral nerves in the immunological reaction could lead to substantial loss of limb functions[6]. The old leprosy lesions at the onset of ENL usually undergo no changes[1],and after the disappearance of a crop of the lesion, the skin may undergo some discoloration changes leaving behind dyspigmented areas as observed in our patient[1]. The ENL lesions could recur at the same sites in about 10-36% of cases[11], and if they do not resolve, a chronic painful panniculitis could develop and may persist for months to years[11]. The lesion of ENL is almost always symmetrical, and the face is involved as in the present case in 50% of situations

and episodes could last as shorts as 2-3 days to weeks[11]. Intermittent fever is a usual accompaniment of ENL, but subacute and chronic cases may not present with pyrexia[11]. Other features of ENL though not seen in this patient, including hepatosplenomegaly, neuritis, arthritis, iritis, iridocyclitis, glomerulonephritis, dactylitis, orchitis, epididymitis and adenitis[1,2,6].

Concerning precipitating factors in ENL, while opinions are diverse in the literature on the role of gender, season and skin color on predisposition to ENL, recognized precipitating factors include infections and infestation like malaria, sepsis, and other intercurrent infections. Others include stress, pregnancy, lactation, menstruation,and medication[1,6,10,11].

Various rare and atypical variants of ENL have been described in the literature. The atypical types may present or herald as necrotic ulceration[7], pustulation[10,11], persistent and localized ENL[13], primary nerves involvement[14], urticarial vasculitis[15], sweet syndrome[16], pyrexia of unknown origin[17] and bullous eruptions[18]. Pustulation reaction is a rare form of ENL reaction[11] but occurred at some point in our patient.

Initiation of treatment is germane in the management of leprosy. The MDT is necessary for treatment as well as for the elimination of leprosy. The medication may, however, precipitate reactions. If the patient is already on MDT, therapy should not be discontinued. Corticosteroids are the mainstay of treatment in ENL. Corticosteroid limits inflammation, decreases pain and prevent nerve damage. Prednisolone in a dose of 40 to 60 mg/day, tapered over 6 to 12weeksis effective[19]. Other useful medication in the treatment of Type 2 reactions may include colchicine, thalidomide, clofazimine, chloroquine, and levamisole[20].

CONCLUSION

Despite the success recorded with the rollout of effective multidrug therapy in the treatment of leprosy, the disease remains in the communities. The pleomorphic nature of its presentations requires thecreation of more awareness among medical workers and non-specialists to limit disabilities and other far-reaching consequences on the sufferers andthe community.

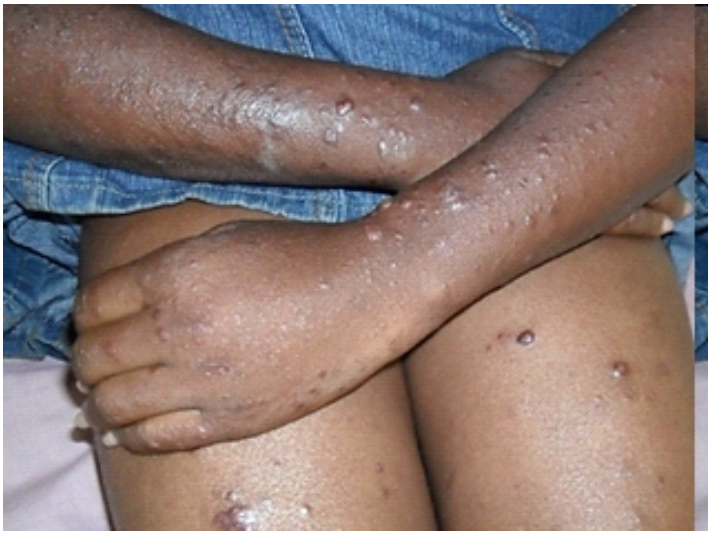

Figure 1: Clinical photograph showing erythematous papulonodular lesion on the extensor surface of the forearms and thighs

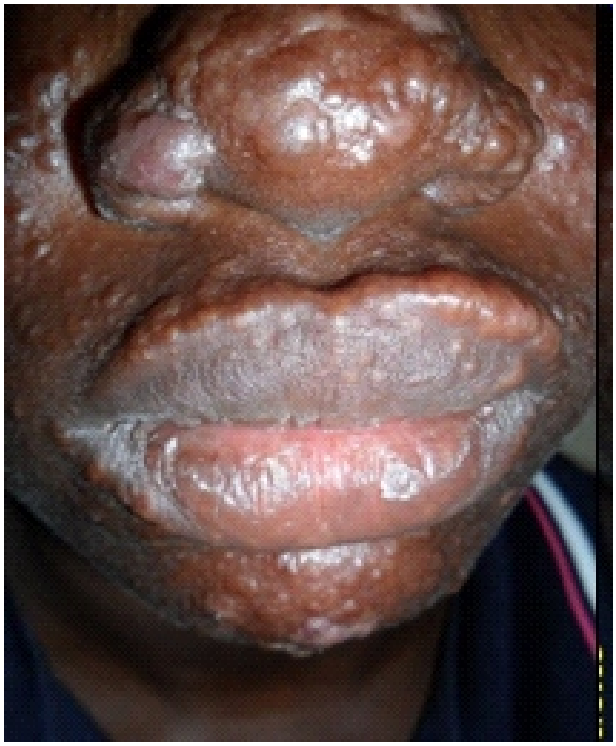

Figure 2: Clinical photograph showing erythematous papulonodular infiltration of the nose, lip margins and the chin.

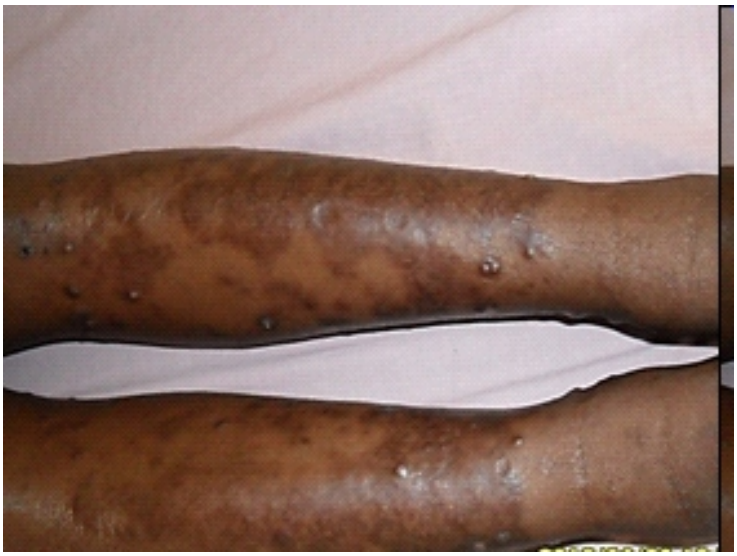

Figure 3: Clinical photograph showing erythematous nodules and plaques on the extensor surface of the legs.

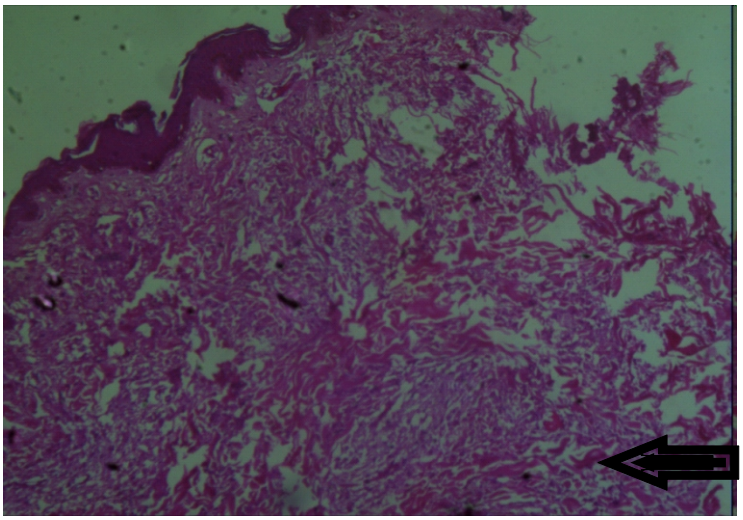

Figure 4. Photomicrograph of a case of lepromatous leprosy showing circumscribed dense dermal masses which are composed predominantly of clusters of macrophages (foam cells),few lymphocytes and plasma cells. A dense mass is shown by the black arrow. (Haematoxylin and eosin, X100).

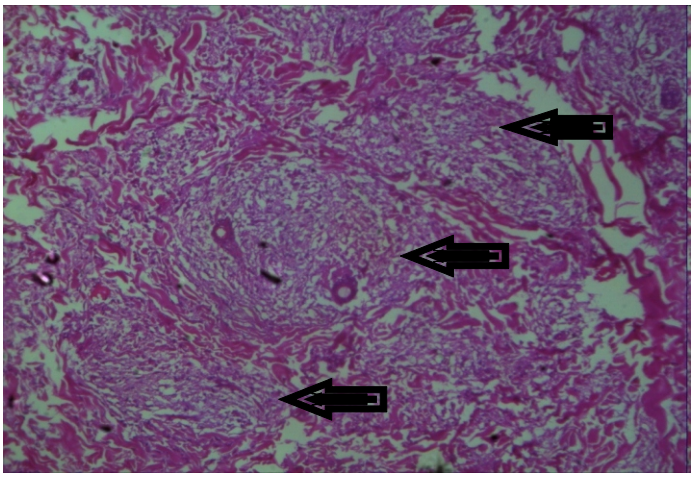

Figure 5. Photomicrograph of a case of lepromatous leprosy showing dermal masses, which are composed predominantly of clusters of bacilli containing macrophages(foam cells),few lymphocytes and plasma cells. The masses are shown by the black arrows. (Haematoxylin and eosin, X100).

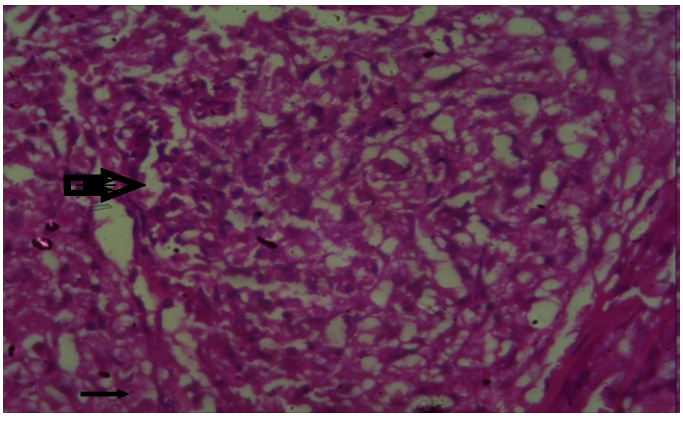

Figure 6. Photomicrograph of a case of lepromatous leprosy showing a dermal nodule composed of clusters of macrophages withfoamy cytoplasm, a few lymphocytes and plasma cells. (Haematoxylin and eosin, X400).

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