Indian J Lepr 2013, 85 : 83-86

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Case Report

Non pitting edema, arthritis and ichthyosis; presenting manifestation of leprosy

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Received : 17.08.2012 Revised : 15.04.2013 Accepted : 25.05.2013

Leprosy is a chronic granulomatous infection caused by *Mycobacterium leprae.* Predominantly involving skin and nerves and having classic clinical description, the diagnosis may be clear-cut in majority of patients but may be challenging in others. Non-pitting edema, ichthyosis and arthritis are uncommon in leprosy and very rare in tuberculoid type where the diagnosis may be challenging unlike relatively clear-cut clinical picture in lepromatous type. Musculoskeletal manifestation is seen in 1-5% of cases and usually associated with reactional states where again the diagnosis sometimes becomes evident. High index of suspicion is therefore recommended in patients with unexplained systemic illness especially in endemic areas. Herein we report a young male with non pitting edema and symmetric peripheral arthritis involving all four limbs, and ichthyosis as presenting manifestation of borderline leprosy where the neuro-cutaneous manifestation developed two months after the said presenting features and in absence of a reactional state.

Key words: Non-pitting edema, ichthyosis, symmetric arthritis, borderline leprosy.

Introduction

Leprosy is a slowly progressive disease caused by *M. Leprae* and its clinical manifestation is very characteristic. The spectrum of disease varies from polar tuberculoid characterised by hypoaesthetic skin lesions and nerve thickening to polar lepromatous characterised by nodular skin lesions, peripheral sensory neuropathy and/or auto-amputation of digits in long standing cases (Modlin and Rea 2008). Microscopic edema is classically described as a feature of type 1 lepra reaction but clinical edema can rarely occur during the course of leprosy (Gelber 2012). Arthritis and ichthyosis are unusual features of leprosy and very rarely they are the initial manifestation causing diagnostic challenges.

Case Report

A 23 years old male presented to medical OPD with non pitting edema involving all four limbs for two months along with arthritis involving bilateral wrist, elbow, ankle and knee joints. Typical morning stiffness was absent. There was

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a diffuse scaly skin lesion (ichthyosis) coinciding with the initial illness (Figure 1). He never had fever, diarrhoea, dysuria or oliguria, dyspnoea or cough. There was no history of jaundice and contact with tuberculosis. He was nondiabetic, nonhypertensive and there was no addiction.

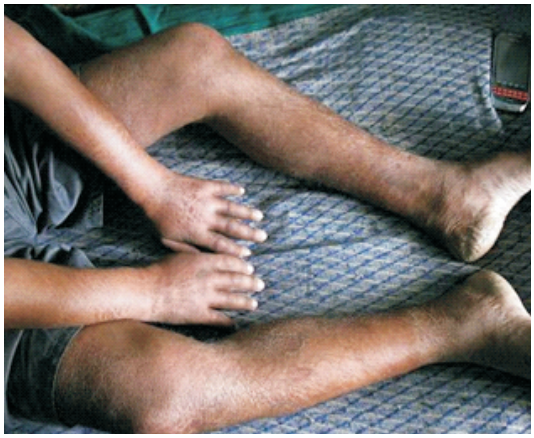

Fig 1 : Non pitting edema and ichthyosis involving all four limbs

The patient was conscious and oriented with average built and nutrition, and vitals were normal. The joints were warm and tender. The skin was dry, thickened and scaly. The edema mainly involved the extremities. Anaemia, cyanosis, jaundice, clubbing or lymphadenopathy was absent. The peripheral nerves were normal. Rest of the general and systemic examination was unremarkable. The patient was admitted for further evaluation when a feasible diagnosis was not made on OPD basis.

During second week of hospital stay there appeared a well defined erythematous macule over right cheek and then over trunk and extremities, which were hypoaesthetic to fine touch and pain [Figure 2, 3]. Over the next week examination of the peripheral nerve revealed nontender thickened great auricular, supra-orbital, ulnar, radial and common peroneal nerves [Figure 4].

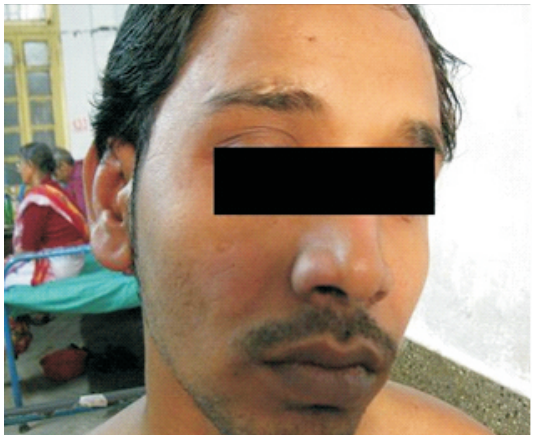

Fig 2 : Hypopigmented lesion involving right cheek

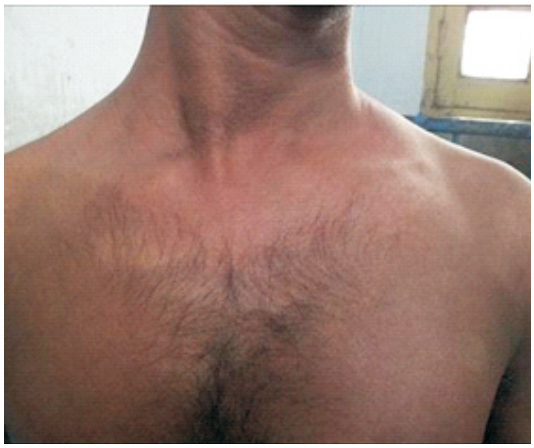

Fig 3 : Hypopigmented patch involving upper chest

Investigation before admission revealed haemoglobin 14.8g/dl, total leucocyte count 8800/cmm, N70 L21 E3 M6; RBC 5.13 × 10^6/cmm, platelet 2.3 × 10^5/cmm. ESR 15mm in 1st hour and CRP 0.56 mg/dl. Bilirubin(T) 0.55mg/dl, AST 48 U/l, ALT 45 U/l, ALP 60 U/l, protein(T) 7.1 gm/dl, albumin 3.8, globulin 3.3 g/dl. Urea 23 mg/dl, creatinine 0.78 mg/dl, uric acid 6 mg/dl. fT_4 1.15 ng/dl, fT_3 3.24 pg/ml, TSH 1.14 µIU/ml. Rheumatoid factor was negative. Further work up revealed anti CCP Ab (chemiluminiscence) 1.7 U/ml (<5). CPK 148,

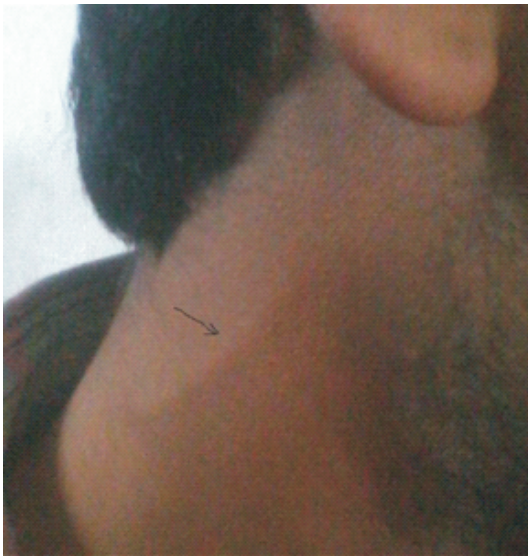

Fig 4 : Thickened right great auricular nerve

CPK-MB 18U/l. Serum aldolase 1.7U/l (1.5-8). ASO titre was 115 IU/ml (N<200). Urine analysis was normal. Slit skin smear from hypo aesthetic patch and edematous skin were negative for acid fast bacilli (AFB). Chest X-ray didn't reveal any abnormality. A clinical diagnosis of borderline lepromatous/borderline borderline (BL/BB) leprosy was made. We started multi drug anti leprosy therapy, patient's clinical condition improved remarkably, arthritis and edema subsided within a month.

Discussion

Leprosy is a chronic granulomatous infection caused by *Mycobacterium leprae*. It primarily involves skin and nerves, but other organs may be affected including eyes, respiratory tract, lymph nodes, testicles and joints (Modlin and Rea 2008). The disease is categorised according to the principal presenting feature that depends on specific immunity to *M. Leprae* (Ridley and Jopling 1966). Polar tuberculoid (TT) usually have hypoaesthetic well defined macules with or without peripheral nerve thickening. Polar lepromatous (LL) on the other hand manifests with multiple poorly marginated skin nodules on the face, ear lobes and other body parts along with distal symmetric polyneuropathy. Nerve thickening are variable features but hypoaesthetic lesions are late feature of LL (Gelber 2012). Clinical edema of limbs is found uncommonly in LL but it is an extremely rare finding in tuberculoid leprosy (Archibald and McDougall 1977). Edema may also be seen in "diffuse lepromatosis" exclusively seen among Caribbean and Mexican population (Gelber 2012). Classic immunologically mediated reactional states are characteristic features of leprosy. Type 1 lepra reaction is seen in borderline leprosy and is characterised by appearance of new macular lesions and increased erythema of the existing lesions as well as neuritis and histologically manifested as dermal edema (Gelber 2012). It is a clinical diagnosis and the nerve thickening characteristic of type 1 reaction is classically 'painful and tender' which was not the scenario in this patient (Modlin and Rea 2008). Moreover there was a chronic phase of edema, arthritis and ichthyosis lasting for over two months prior to appearance of the neurocutaneous lesions that exclude the possibility of a lepra reaction. Icthyosis is rarely seen in leprosy and very rarely it is the initial presentation (Chauhan et al 2010). Musculoskeletal manifestation may be seen in leprosy (1-5%) in the form of arthritis, enthesitis, sacroiliitis, cryo-globulinaemic vasculitis and dermatomyositis but they are hardly seen in absence of a reactional state (Chauhan et al 2010). Rarely patient may present with distal symmetric polyarthris involving hand joint resembling rheumatoid arthritis as seen in our case (Chauhan et al 2010). Kidney disorders and myxedema were excluded in our patient with appropriate tests.

The striking part of our patient is that limb edema, peripheral arthritis and ichthyosis were the initial presenting feature of the disease causing diagnostic dilemma and typical skin and nerve lesions developed only after two months of disease onset which ultimately helped in diagnosis. Therefore clinician should have high index of suspicion of a diagnosis for leprosy in patients presenting with unexplained systemic illness even in absence of classical lesions of leprosy.

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How to cite this article : Moulick A, Jana A, Sarkar N et al (2013). Non pitting edema, arthritis and ichthyosis; presenting manifestation of leprosy. *Indian J Lepr.* **85** : 83-86.