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Case Report

Primary Neuritic Hansen's Disease presenting as Ulnar Nerve Abscess in a Human Immunodeficiency Virus Positive Patient

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Leprosy has been increasingly known to have an enigmatic relationship with human immunodeficiency virus infection. Co-infection may result in atypical manifestations of leprosy. A 45-year old human immunodeficiency virus- positive male, agricultural laborer presented with a swelling over right elbow, right hand deformity, generalized itching and recurrent vesicles over the perinasal area. Clinical and investigational findings were consistent with mononeuritic type of Hansen's disease with right sided silent ulnar nerve abscess, partial claw hand. CD4+ count of the patient was 430 cells/ cmm. This patient also hadherpes simplex labialis, with HIV-associated pruritus. To the best of our knowledge such an atypical presentation has not been reported earlier.

Keywords : Leprosy, HIV, Coinfection, Primary neuritic Hansen's, Nerve abscess

Introduction

Leprosy is a chronic infectious disease caused by *Mycobacterium leprae* which damages the skin and peripheralnerves (Britton and Lockwood 2004). Human Immunodeficiency Virus (HIV) infection has altered the epidemiology of mycobacterial diseases. This has led to an increase in several illness associated with anumber of mycobacterial diseases, particularly those of the *Mycobacterium avium* intracellulare complex in the industrialized world and of tuberculosis (TB) in sub-Saharan Africa, the former Soviet Union, Asia including India (Massone et al 2011)). The interaction between TB and HIV led the WHO to declare TB a global emergency (WHO 2002). Although an outbreak of leprosy was expected, most reports suggest HIV to have little influence on the course of leprosy (Goodless et al 1994, Schettini et al 1996, Naafs 2000; Silva et al 1997; Ustionasski et al

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2006). However, after the introduction of highly active antiretroviral therapy (HAART) leprosy may present as immune reconstitution inflammatory syndrome (IRIS), typically as paucibacillary leprosy with type 1 reaction (Landay et al 2002, Lawn et al 2003, Pignataro et al 2004, Visco-Comandini et al 2004, Trindade et al 2005, Kharkar et al 2007, Batista et al 2008, Menezes et al 2009, Couppié et al 2004, Martiniuk et al 2007). (Nery et al 2000) concluded that HIV co-infection did not seem to change the natural course of the disease or cause difficulties in diagnosis and treatment. Lucas (1993) considered that leprosy was a missed disease in AIDS. Since most clinical signs of leprosy are dependent on cell mediated immunity, it has been postulated that when HIV patients with latent leprosy infection receive anti retroviral treatment, their immune system recovers, recognising *M. leprae* antigens unfolding the clinical disease (Naafs 2000). Sharing of several antigenic epitopes between *Mycobacterium leprae* and HIV proteins may be modulating the response (Husain et al 2007). In the present study we are reporting the clinical profile of a patient with HIV-leprosy coinfection presenting as primary neuritic Hansen's disease with ulnar abscess and ulnar claw hand.

Case Report

A 45 year old male agricultural laborer, presented with itching all over the body, swelling over right elbow, right hand deformity, recurrent fluid-filled lesions over the perinasal area for the last six months. He also complained loss of sensation over right ulnar area. Initially, the patient noticed a small swelling over right elbow with gradual increase in size, with subsequent development of right claw hand. There was no history of trauma, non healing ulcer, hypopigmented patches over the body or systemic complaints. Family history was non-contributory. His general physical findings and vital parameters were normal expect for mild pallor. Cutaneous examination revealed soft, oval swelling measuring about 2 cm x 5 cm, with mild tenderness just above the right elbow region (Fig 1). Ulnar nerve behind the medial epicondyle was thickened, tender and connected to the swelling. Sensation for all modalities were absent over the medial half of dorsum of hand and medial one-third of palm with ulnar clawing (Fig 2). No other nerves were thickened. Tingling along ulnar nerve distribution on tapping the swelling was noted. No hypo pigmented patches found over any part of the body. There was no infiltration over face, ear lobes and any other part

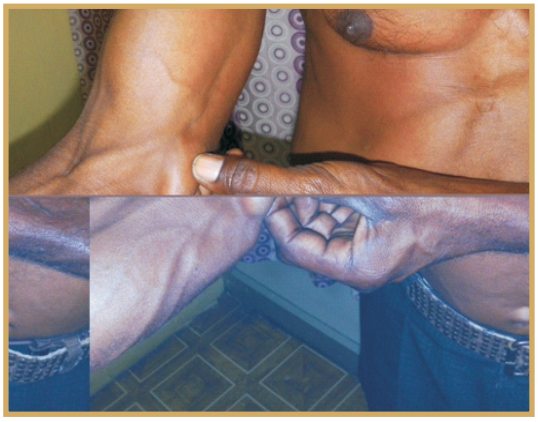

Fig 1 : Right Ulnar nerve abscess

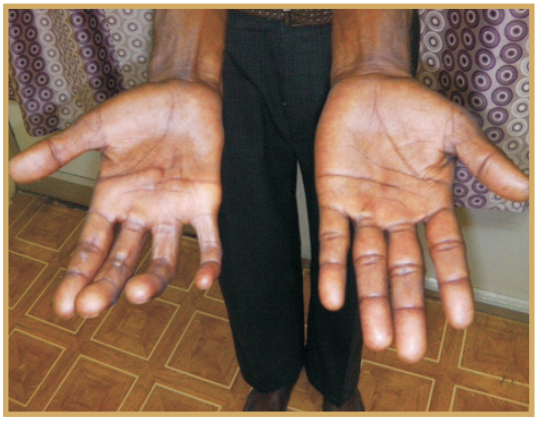

Fig 2 : Right sided Ulnar Claw hand

of the body. Wasting of the hypothenar eminence of right hand and flattening of the medial border of right forearm. Complete hemogram and biochemical profile was unremarkable expect for mild anemia. Enzyme linked immunosorbent assay for HIV-1 was reactive with CD4 count, 430 cells/mm^3. Serology for syphilis, hepatitis B and C were negative. Skin slitsmear examination from both ear lobes and eyebrows were negative for AFB. Voluntary muscle testing score for right abductor digiti minimi was 3 on the Medical Research Council (MRC) scale. Ultrasonography was suggestive of ulnar nerve abscess. Histo-pathology from the abscess showed ill formed granulomas in the dermis involving the nerve fibres. Granulomas comprised of multinucleated giant cells, epitheloid cells and lymphocytes, AFB were not detectable - with these findings borderline tuberculoid leprosy was made. A final diagnosis of mononeuritic type of Hansens with right sided silent ulnar nerve abscess, partial claw hand and also having herpes simplex labialis (clinical diagnosis), with HIV associated pruritus was made and the patient was started on multibacillary therapy includes Rifampicin 600mg, Clofazimine 300mg and Dapsone 100mg monthly once supervised dose, Dapsone 100mg and Clofazimine 50mg daily self- administered for one year. Systemic steroids started at 60mg of prednisolone per day gradually tapered over a period of 6 months. Patient was not started on ART as CD4 count was more than 350 cells/mm^3. Physiotherapy was started with gradual improvement in motor functions were noted. Patient did not come back after completion of one year MB-MDT treatment and was lost for follow-up.

Discussion

It seems likely that the immunological changes caused by the HIV infection and the HAART triggers new manifestations that brings the patients for diagnosis and treatment. Health authorities and health workers in countries in which leprosy and HIV infections are endemic should be aware that a latent leprosy infection can be present in HIV infected patient and that this infection can be revealed when HAART is introduced. Moreover, that an immunological reaction may occur leading to loss of nerve function.

In the era of HIV pandemic, lepromatous cases were expected to increase, due to the reduction in cell mediated immunity in HIV. But contrary to this belief, HIV does not affect the migration of *M. leprae* specific CD4+ T cells to the lesion site or their response to the antigen (Sharma and Malhotra 2008). However, sharing of several antigenic epitopes between *M.leprae* and HIV virus might be responsible for different relationship with leprosy as compared with tuberculosis and *M.avium* as well as some other mycobacterial infections (Husain et al 2007).

There are a few reports of leprosy associated with HIV (Schettine et al 1996, Goodless et al 1994). Spectrum of the skin lesions ranging from hypopigmented tuberculoid lesions to nodular lepromatous lesions have been described in HIV infected patients (Singhal 2010). Few atypical and rare skin manifestations of leprosy have been reported in association with HIV.

A case of borderline tuberculoid leprosy in a HIV-positive patient who developed a marked reversal reaction was reported and it was noted that HIV co-infection seemed to be associated with an increased rate of reactional states and more severe cases of neuritis. Atypical and rare skin manifestations, such as verrucous lesions and ulcers, may appear after HAART resulting in increased CD4+ T-lymphocyte count and drop in viral load.

Contrary to the above, although antiretroviral drugs were not initiated in our case, yet patient presented with neuritis and a silent nerve abscess, without other cutaneous signs. Worsening of nerve damage might be expected in patients with dual infection as HIV infection may alter the immune response in nerves towards *M.leprae*. HIV is itself neuropathic; so, it could also act synergistically. However, conclusive evidence to support this hypothesis is lacking (Singhal 2010).

Immunologically driven inflammation is also responsible for much of the clinically apparent nerve injury. Nerve function impairement is more rapid and severe in patients with aggressive cellular response i.e. in tuberculoid disease and during reactional states, especially type 1 reaction. As HIV principally affects cell mediated responses, these pathogens may also have potentially interesting immunologic interactions in the human host. Unlike tuberculosis, where HIV infection affects granuloma formation depending on the degree of immune suppression as reflected by blood CD4+ counts, host granulomatous response is preserved among individuals co-infected with HIV and leprosy (Singhal 2010).

Bacterial parasitization of peripheral nerves is a unique feature that is characteristic of leprosy. In majority of the immunocompetent patients, the resulting neural lesion remains as a granuloma but in a few cases the granuloma may soften and develop into an abscess. Progression to abscess formation is most commonly seen in patients with tuberculoid leprosy. Rarely, however, nerve abscess may also develop in other types of leprosy (Kumar et al 1997).

The usual habitat for *M.leprae* in the nerve is the schwann cell but occasionally the ensheathed axon becomes involved. The Schwann cells assume a phagocytic function and evolve into macrophages or epitheloid cells resulting in the formation of a granuloma. Invasion of the endoneurium may follow and the whole endoneurial zone may appear to be occupied by epitheloid cells with or without the presence of bacilli. Caseation may occur in microscopic foci within the granulomas or areas of necrosis may coalesce, forming a cold abscess particularly when the immunity is high. Cold abscesses occur more frequently in the tuberculoid form in India (Singh and Ojha 1969, Char and Cross 1986). Pure neural leprosy without skin involvement is rare and comprises 4-6% of leprosy cases. Majority in this group present with sensory impairment, 5-10% with deformity and rarely with nerve abscesses (Sharma and Malhotra 2008).

Our patient, although immunocomprised, presented with mononeuritic involvement without cutaneous sign, silent nerve abscess and associated ulnar claw hand. To the best of our knowledge, this is the first case report of such an association. In our case, high-resolution ultrasound helped in unearthing a silent nerve abscess, highlighting the long term utility of this non-invasive modality in early detection of silent neuritis or nerve abscess, thereby preventing deformities and disabilities. Ultrasound has been used early for this purpose (Taneja et al 1992) and our study re-emphasizes the use of this tool to study the nerve affliction in leprosy.

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Key messages

Coinfection with leprosy and HIV may be commoner in endemic areas. Awareness, recognition and early management of atypical manifestations of leprosy in the presence of HIV helps in preventing the complications.

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