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Case Report

Pure Neuritic Leprosy with Type 2 reaction

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Pure neuritic leprosy (PNL) is considered a distinct form of leprosy by Indian Association of Leprologists. Reactions usually reported in PNL are Type 1 reactions manifesting as neuritis with a rare involvement of the skin. We report a case of Type 2 reaction in PNL. This case, a 30 year old male, presented with history of shock like sensation over both upper limbs and lower limbs and tingling numbness for duration of one year. Clinical examination did not reveal any observable patches, plaques or nodules on the entire body. Touch, pain and temperature sensations were impaired on the ulnar aspect of both upper limb and even over both lower limbs. Motor examination revealed weakness of small muscles of both hand. Enlargement of ulnar, ulnar cutaneous nerves, lateral popliteal, anterior tibial and sural nerve was found on both sides with minimal tenderness over the ulnar nerves. Slit skin smears were negative for acid fast bacilli. Nerve conduction study showed asymmetric mixed sensorimotor polyneuropathy. Biopsy from hypoaesthetic area on right forearm revealed mild lymphocytic and macrophage accumulation around the dermal vessels, sweat glands and neurovascular bundles, suggestive of Indeterminate Hansens. Nerve biopsy from right sural nerve gave findings of Lepromatous Hansens neuritis with the epineurium thickened, medial hypertrophy of nutrient artery and neovascularisation. Wade Fite Faraco stain showed multiple acid fast bacilli. Diagnosis of Pure Neuritic Leprosy with Type 1 reaction was made and patient was started on MB-MDT. During the treatment he developed ENL which was successfully managed with Steroids and Thalidomide. It will be important to keep such mixed atypical presentations in mind when dealing with suspected cases of PNL.

Keywords : Pure Neuritic Leprosy, Type 2 reaction

Introduction

Neural leprosy is considered a distinct form of leprosy by the Indian Association of Leprologists (IAL 1982, Rao & Suneetha 2016). It constitutes about 418% of leprosy patients as per Indian studies (Sharma & Malhotra 2008) Type 1 reactions have been reported in pure neuritic leprosy (PNL), which are mainly manifested by

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increase in intensity of neuritis or associated with increased nerve function impairment (Kumar et al 2004). Skin lesions have been reported in reversal reactions in patients with PNL either very early (Indeterminate) or with advanced multibacillary neural pathology (Mishra et al 1995, Guilloton et al 2002). Here we report a case of Pure Neuritic Leprosy with type 2 reaction.

Case Report

A 30 year old male presented in our OPD with history of shock like sensation over both upper limbs and lower limbs and tingling numbness for duration of 1 year. He did not give history of noticing any skin lesions. No history of any motor weakness or sensory disturbances in hands and feet.

On examination he was afebrile with no pallor, icterus, lymphadenopathy and pedal edema. There were no observable patches, plaques or nodules on the entire body. Touch, pain and temperature sensations were impaired on the ulnar aspect of both upper limb and even over both lower limbs. Motor examination revealed weakness of small muscles of both hand. Enlargement of ulnar, ulnar cutaneous nerves, lateral popliteal, anterior tibial and sural nerve was found on both sides with minimal tenderness over the ulnar nerves.

All his blood investigations, ECG and CXR were within normal limits. Bacteriological index was negative. Nerve conduction study showed asymmetric mixed sensorimotor polyneuropathy. Biopsy from hypoaesthetic area on right forearm revealed mild lymphocytic and macrophage accumulation around the dermal vessels, sweat glands and neurovascular bundles, suggestive of Indeterminate Hansens (Fig. 1).

Nerve biopsy from right sural nerve gave findings of Lepromatous Hansens neuritis with the epineurium thickened, medial hypertrophy of nutrient artery and neovascularisation. The perineurium was also thickened, with subperi-

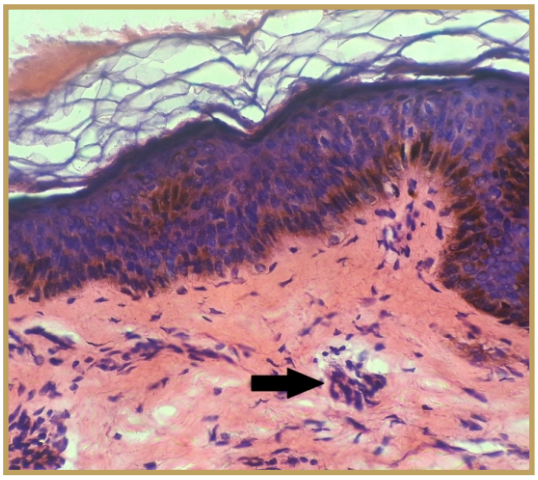

Fig 1 : Biopsy from hypoaesthetic area - Mild lymphocytic and macrophage accumulation around the dermal vessels, sweat glands and neurovascular bundles, H&E 40X.

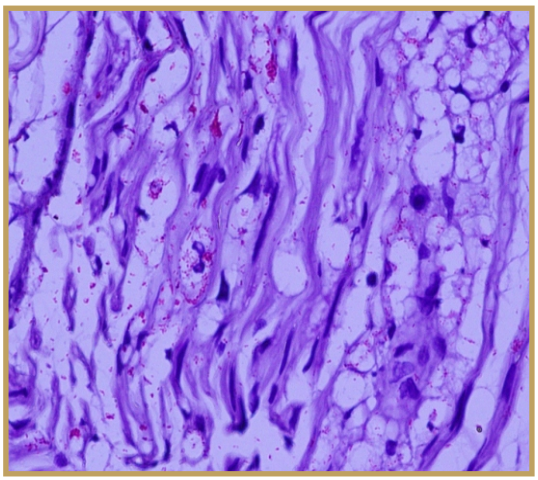

Fig 2 : Biopsy from sural nerve - Both endoneurium and perineurium showing foam cell collections with many acid fast Lepra bacilli. Wade Fite - Faraco stain x 400

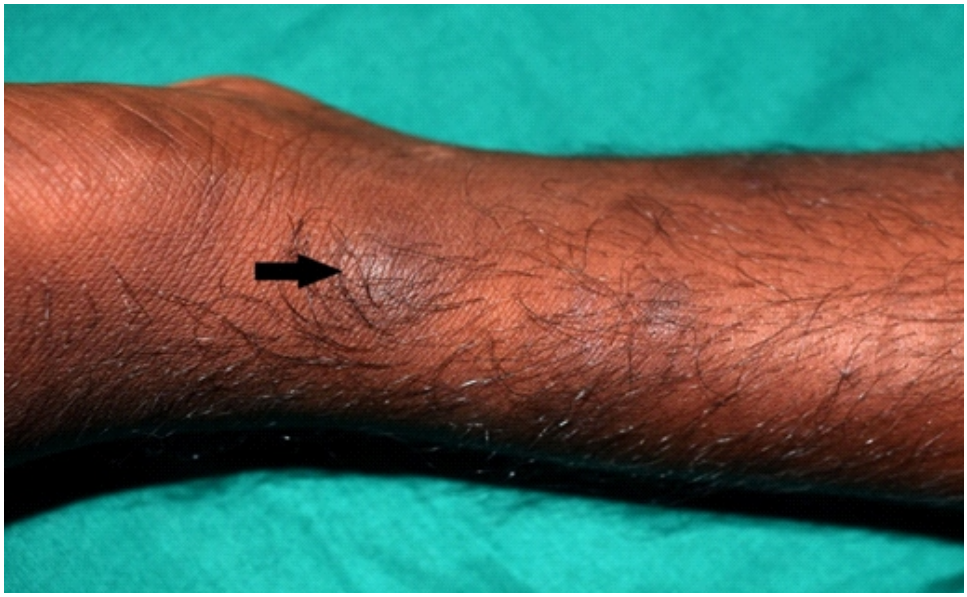

Fig 3 : ENL lesions over the lower limb

neurial edema. Both endoneurium and perineurium showed foam cell collections. Wade Fite Faraco stain showed multiple acid fast bacilli (Fig. 2). Diagnosis of Pure Neuritic Leprosy with Type 1 reaction was made and patient was started on MB-MDT with adequate rest of the affected nerve and tablet Prednisolone was given in tapering dose for one month.

After around two month of treatment the patient reported with complaints of painful crops of evanescent lesions on face, upper limbs and lower limbs. Also history of fever, malaise, body aches was present. Cutaneous examination revealed red, tender, ill defined, nodules and papules were present on bilateral upper limbs, face and lower limbs (Fig. 3). Biopsy from these lesions showed focal collection of foam cells and inflammatory infiltrate composed of lymphocytes and neutrophils in the dermis and subcutaneous fat suggestive of lobular panniculitis. The diagnosis of ENL was made and the patient was started on Thalidomide and steroids. On follow up there were no new lesions and previous lesions healed with post inflammatory hyperpigmentation.

Discussion

"Neural leprosy" with only nerve involvement without obvious skin lesions was first proposed as a separate type of leprosy by Wade in 1952. The Indian Association of Leprologists (IAL) recognized "neural leprosy" as a distinct type of leprosy and included it in their official six group classification in 1955 and named it "polyneuritic leprosy" (Prasad 2005). The features of pure neuritic leprosy (PNL) as approved by IAL in 1982 included absence of skin lesions, nerve enlargement, sensory loss along distribution of nerves, smear negative and histology of either of tuberculoid, borderline or non-specific type (IAL 1982).

The prevalence of PNL varies from 4-18% with single or multiple nerve involvement. PNL may be an early stage in the pathogenesis of the disease

before the appearance of skin lesions (Kumar et al 2004, Rao & Suneetha 2016).

The widely accepted classification for leprosy proposed by Ridley Jopling or the classification of WHO does not include neuritic leprosy in its group system (Prasad 2005). However, IAL classification recognizes pure neuritic leprosy as distinct entity (IAL 1982). Considering these two classifications, and based on histology and nerve involvement, a diagnosis of lepromatous leprosy with type 2 reaction could be considered in this patient. But according to IAL classification, he was diagnosed as PNL as there was no skin lesion and the slit skin smear was negative at the time of initial presentation, though there was extensive nerve involvement. Though following the initiation of therapy and withdrawal of steroids he developed ENL lesions, but again when he was started on Thalidomide and Steroids the skin lesions again disappeared.

Histopathological spectrum of nerves in PNL ranges from tuberculoid to lepromatous leprosy. Few studies have demonstrated that the biopsy from apparently normal skin or nasal mucosa had majorly leprosy features (Kaur et al 1991, Suneetha et al 2000-1, Jardim et al 2003, Rao & Suneetha 2016).

Reactions in the form of Neuritis in PNL may be present in 40 -50 % mainly in the form of reversal reactions. Features of type 2 reactions are not reported. Reactions in PNL can go unnoticed unless associated with skin lesions (Guilloton et al 2002, Jardim et al 2003, Kumar et al 2004, Rao & Suneetha 2016). Intensity of reactions depends on the immune response to *Mycobacterium leprae* or bacterial load in the nerves. Many studies have shown a moderate-to-heavy bacterial load within the nerves with features of lepromatous changes. Similar finding was noted in our patient (Kaur et al 1991).

The occurrence of Type 2 reaction in our patient was probably due to high bacillary load in the nerves and the breach in blood nerve barrier which occurs due to various reasons leading to bacteraemia; also following the introduction of MB-MDT there might be lysis of bacilli, with increased antigenic products and immune complex formation which has led to type 2 reaction during therapy.

By definition, patients with pure neuritic leprosy should not have clinical skin lesions. But studies have shown that though the skin appears clinically uninvolved, histopathological changes of leprosy are present. Hernandez et al (2011) reported histopathological changes of leprosy in the apparently normal looking skin of 32.1% patients with 12.7% of them having either epithelioid or macrophage granulomas. This study concluded that the absence of visible hypopigmented skin lesions in these patients is probably related to the deep location of the granuloma in the dermis, which are, therefore, unable to exercise any direct influence on the melanocytes in the epidermis (Hernandez et al 2011).

To conclude while Type 1 reactions with or without skin lesions are known to occur in PNL, however, the PNL cases having high bacillary load and lepromatous involvement of the nerves, patients can be predisposed to develop type 2 reaction.

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