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Original Article

A Rare Combination of Pure Neuritic Leprosy with Morphea Leading to Diagnostic Confusion

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A circumscribed sclerotic plaque of morphea can sometimes be mistaken for tuberculoid leprosy and vice versa can also happen. However, the co-existence of a patch of morphea mimicking as Leprosy patch in an underlying case of neuriticleprosy, can be very misleading. We present a case with glove and stocking anaesthesia and peripheral nerve enlargement with a single large hypopigmented, non-anaesthetic macule on trunk, clinically diagnosed as Hansen's disease (Borderline Tuberculoid - BT). Slit skin smears proved to be negative for AFB and histopathology of the skin lesion was consistent with morphea, which lead us to do a nerve biopsy. Sural nerve biopsy proved it to be Hansen's neuritis with occasional bacilli. The patient was started on MDT-MB and followed up. This is a rare case of co-existing morphea with Hansen's disease. It would have been easily misclassified if we had presumed the cutaneous lesion to be a case of Hansen's (BT) patch and not done a cutaneous nerve biopsy which led to diagnosis of multibacillary leprosy.

Key words : Pure neuritic leprosy, Morphea

Case Report

A 56 year old agriculturist hailing from north east presented with complaints of diminished sensations including inability to perceive heat sensation over his hands and feet since last 4 years, a painless, non-healing ulcer on his left sole and left hand since 1 year. Patient denied history of taking any treatment for these problems.

On examination, sensations to hot and cold temperatures, superficial touch and pain were markedly reduced over both forearms including hands, and over the lower limbs, knees and below. Bilateral greater auricular (Fig 1), ulnar, radial, popliteal nerves were uniformly thickened and tender on palpation. A single large well-defined non-anaesthetic, hypo pigmented mildly atrophic macular patch (Fig 2) measuring about 6 x 8 cms with a mildly erythematous border was found over the left side of abdomen extending

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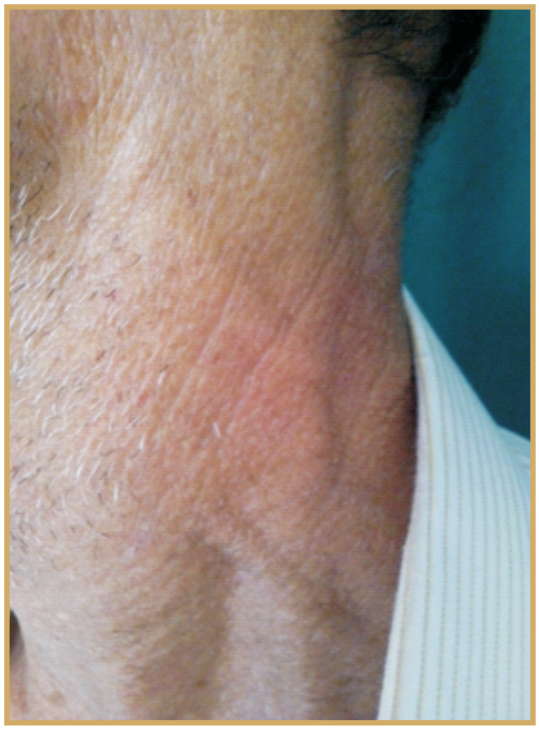

Fig 1 : Enlarged left greater auricular nerve

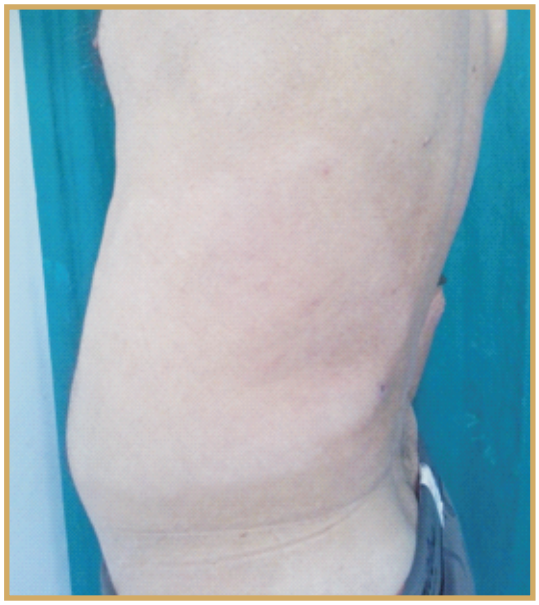

Fig 2 : A single large well-defined non-anaesthetic, hypo pigmented mildly atrophic macular patch measuring about 6 x 8 cms with a mildly erythematous border was found over the left side of abdomen extending onto the back.

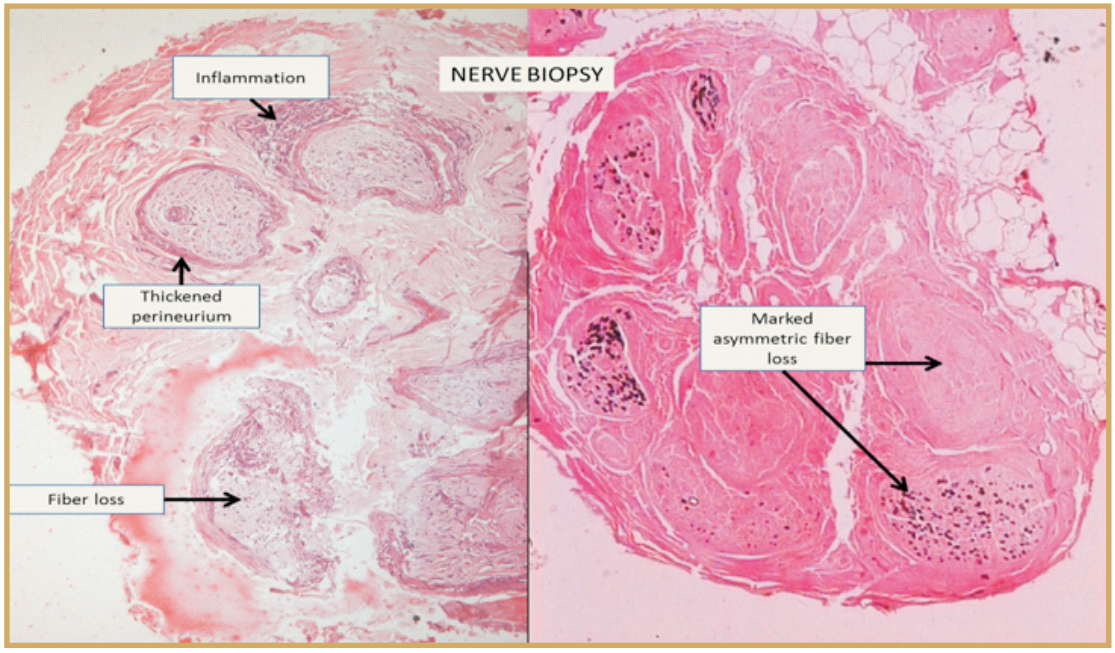

Fig 3 : Nerve biopsy showing features of chronic neuritis

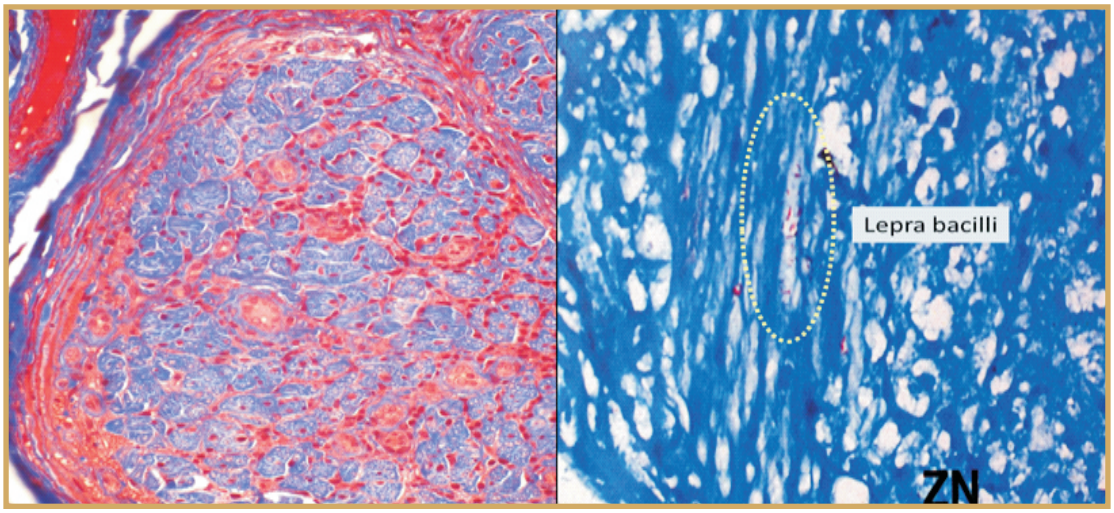

Fig 4 : Nerve biopsy showing Acid fast bacilli on ZN staining

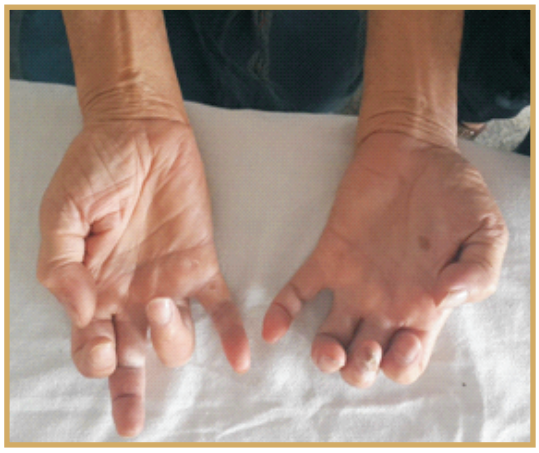

Fig 5 : Complete clawing (radial and ulnar) of the right hand and partial (radial) clawing of the left hand.

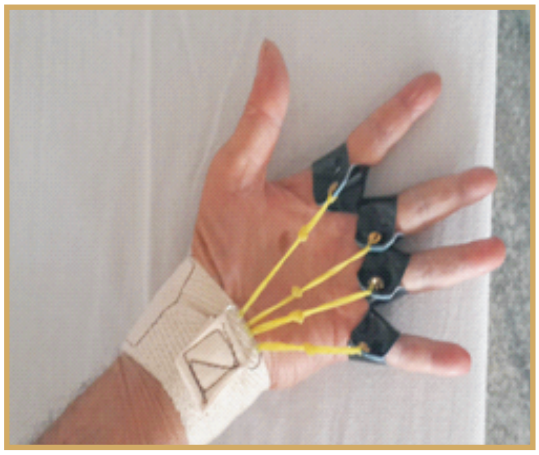

Fig 6 : Splinting and physiotherapy for recovery from palsy.

onto the back. With these clinical findings a clinical diagnosis of Hansen's disease was made.

Baseline hemato-biochemical parameters were normal. Slit skin smears revealed no acidfast bacilli. Skin biopsy from the hypopignmented patch was done. The histopathology revealed a thinned out epidermis, flattened rete ridges, plenty of hyalinised dermal collagen, decreased adnexae, mild perivascular lymphocytic infiltrate with no evidence of granulomas. The features were consistent with morphea. In view of the different skin biopsy finding a sural nerve biopsy was done which proved it to be Hansen's neuritis (Fig 3, 4) with occasional bacilli. The patient was

started on MDT-MB. Testing for anti-nuclear antibodies (ANA) was done, result was negative.

One month after the initiation of therapy, the patient developed painful swelling of hands and feet along with difficulty to perform fine movements in fingers. On examination, he had gross oedema of hands and feet, tender enlargement of all the peripheral nerves, complete clawing (radial and ulnar) of the right hand and partial (radial) clawing of the left hand (Fig 5). There was no evidence of new skin lesions or any changes over the existing skin patch (morphea), proving the patch to be unrelated to the Hansen's disease. A diagnosis of type I lepra reaction was made. The patient was started on 60 mg of oral Prednisolone per day along with physiotherapy (Fig 6, 7). A decrease in the symptoms was noted within 5 days of initiation of prednisolone therapy. It has been planned to taper the steroid gradually over next few months.

Discussion

Clinically hypo pigmented patches can be a manifestation of both morphea and leprosy in addition to several other conditions.

Morphea is characterised by localised sclerosis of the skin, is due to an unknown cause, a symmetric distribution, confined to one body area, with single or few lesions. It is caused due to the defect in fibroblastic cells in which alterations in the growth factors (platelet-derived growth factor) (Zheng et al 1998) and receptor expression (TGF-β) (Kubo et al 2001, Mahajan et al 1996). These alterations appear to lead to increased connective tissue growth factor (CTGF) gene expression and finally fibrosis (Igarashi et al 1996, Stratton et al 2001). The plaque lesions are indurated purplish in colour. After some months they become thickened and a characteristic lilac-coloured edge develops. At times they may present with diminished sensibility (Goodfield et al 2010, Sehgal et al 2002).

Leprosy is a chronic granulomatous disease caused by *Mycobacterium leprae* primarily involving the peripheral nerves and skin. The diagnosis can pose a clinical challenge with its diverse manifestations. For treatment purposes clinically patients with leprosy are classified as the paucibacillary (1-5 lesions) or the multibacillary (more than 5 lesions) subtype, reflective of the host immune response; the paucibacillary subtype is characterized by a predominantly Th1 cell-mediated immune response and the multi bacillary subtype is characterized by a predominantly Th2 humoral response (Fleishmajer et al 1977).

Leprosy is primarily a disease of the nerves and Schwann cells. The nerves are the first targets of *M. leprae* and hence neural involvement and its manifestations may be the earliest features of *M. leprae* infection. These may manifest as sensory or motor symptoms such as anhidrosis, alopecia's, paraesthesias, dryness of skin especially acral areas, trophic ulcers and deformities.

This early neuritic stage may continue as such in nerves or progress or disseminate based on host immunity. According to Dharmendra and Jopling's classification of leprosy (Jopling and McDougall 1988) the polyneuritic leprosy is mostly an unrecognised form the disease which may be incompletely treated. However, Indian classification, (Dharmendra 1985) recognises a clear neuritic or polyneuritic type. It is estimated that up to 10% of cases can involve only nerves without skin involvement.

In this case the patient had reported with diminished sensations, nerve thickening and trophic ulcer which suggested an initial diagnosis of leprosy. But skin biopsy from the hypo-pigmented patch revealed features of morphea, leading to a diagnostic confusion. The nerve biopsy revealed the underlying Hansen's neuritis. This stresses the importance of a complete and

thorough examination and investigation in cases with such diagnostic difficulty (Jose et al 2013).

We are presenting this case as a rare combination of pure neuritic leprosy with morphea which should be kept in mind as it can create confusion in the management of such cases.

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