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Letter to Editor

Uncommon Presentation of a Common Disease - Response to letter by Jain et al

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Dear Sir

We thank Dr Jain and colleagues for their interest in our paper and raising some important issues on the diagnosis and management of this patient. The diagnosis of ganglionopathy may have differing clinical, imaging, electrodiagnostic and pathological correlates. The patient with ganglionopathy may present either with sensory ataxia or with neuropathic pain or both (Smith et al 2005). Our patient had sensory ataxia, pseudo-athetosis and incoordination suggesting large fibre involvement. Presence of dysaestheia, loss of pain and temperature in distal lower limb were suggestive of small fibre involvement. In such a situation leprosy must be considered even if there is no nerve enlargement or maculoanaesthetic patch. We have reported severe pseudoathetosis due to leprosy (Misra et al 2003). Ganglionopathy like symptoms are generally attributed to autoimmune disorders, intoxication, diabetes and malignancy. We excluded these conditions and treated with multidrug therapy. Following which the patient showed progressive improvement and is now normal and off drugs. Leprosy is known to cause "quiet nerve paralysis", or "silent neuropathy" because its clinical manifestations appear only when 25%–30% of the nerve fibres in a nerve trunk become non-functional. Below this threshold, clinical neurologic examination does not show any cutaneous sensory deficits (Shelley & Shenoy 2018). In our patient, neuropathic symptoms were minimal but slit smear was positive for *M. leprae* confirming the diagnosis of leprosy. In leprosy, isolated ganglion involvement is unlikely because the infection of peripheral nerves has been attributed to an ascending neuropathy originating in sensory cutaneous nerves which travel proximally to involve larger nerve trunks carrying mixed sensory and motor fibres (Sabin et al 1993). Though the patient had only neurological symptoms and signs, cannot be classified as pure neuritic leprosy because *M. leprae* was positive in slit smear. Based on symptoms, signs and investigations, our case can be categorised as possible ganglionopathy (Camdessanché et al 2009). In such a situation adjacent root and nerves may also be involved. MRI abnormalities would have diagnosed this patient as probable ganglionopathy and a positive biopsy would have led to the diagnosis of definite ganglionopathy. Biopsy was not considered because patient was improving on MDT. The vigour of investigations should be rational. In this patient when the diagnosis was based on

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simple slit smear examination, other causes were excluded by laboratory tests, and malignancy by PET. Cerebrospinal fluid examination was also not considered important in our patient. No doubt leprosy involves small fibres but the large fibres may also be involved in severe nerve damage, under low immunity and during lepra reaction which may result in pseudoathetosis, and even Charcot's joint (Misra et al 2023). When the large nerve fibres are affected, impairment of vibration sense may lead to areflexia. In our patient, presence of sensory loss in hands while it was present below knee and areflexia is not consistent with length dependent axonopathy.

We have prescribed prednisone 20mg which was tapered in due course. If there is any worsening or aggravation of symptoms higher dose may be prescribed; thereby side effects of high dose of corticosteroid can be avoided. We feel that the patient had full recovery by MDT for 1 year and low dose of prednisone.

Enhancement of dorsal root ganglia in MRI would have supported the diagnosis of ganglionopathy but its absence does not exclude it. In this patient, biopsy of ganglion was not considered prudent as the patient was improving and we strongly felt that the clinical syndrome was due to leprosy. The aim of this case report was to highlight that in the patients simulating sensory neuropathy of ganglionopathy before considering malignancy, immunological and toxic disorders, leprosy should be considered because it is rewarding to diagnose and treat.

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