CASE REPORT

Extensive Ulnar Nerve Necrosis: A Complication of Tuberculoid Leprosy

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Summary Pure neural leprosy without cutaneous manifestations is a relatively rare manifestation of leprosy. It can present as a mono- or poly-neuritis with sensory and/or motor impairment. Neural leprosy may or may not be associated with thickening of the involved nerve. We report the case of a 14-year old boy with extensive ulnar nerve necrosis who was diagnosed to have tuberculoid leprosy. What makes this case unique is that we have here a case of pure neural leprosy with a single nerve turned ‘necrotic.’

Introduction

Mycobacterium leprae (M. leprae) has a predilection for affecting peripheral nerves, with dermal nerves being affected commonly. However, involvement of peripheral nerve trunks alone without skin manifestations of dermal nerve involvement, termed pure neuritic leprosy (PNL), is unusual.¹

These patients can present with mono- or poly-neuritis with sensory and/or motor impairment. Moreover, the symptoms of neuritis may or may not be associated with thickening of the involved nerve.² Our patient presented with extensive ulnar nerve necrosis due to leprosy.

CASE REPORT

A 14 year old boy presented to the surgical out-patient clinic with history of weakness of his right hand for the past 2 years. He also complained of a slowly but progressively increasing swelling over the inner aspect of his arm above the elbow over the past one and a half years. He had been on anti-leprosy treatment for an initial period of 6 months under a peripheral outreach center for suspected pauci-bacillary leprosy. Due to the persistence of ulnar nerve necrosis, he was referred to the plastic surgery department for further evaluation and management.

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thickening the patient was placed on a further 6 months of anti-leprosy therapy. He was, however, not placed on steroids.

Local examination at the time of presentation to our hospital (1 year after initial suspicion of leprosy), revealed a claw-hand deformity of the right hand with (gross) wasting of intrinsic muscles of the right hand. In addition there was sensory loss in the ulnar nerve territory. A small soft non-fluctuant subcutaneous swelling was noted on the medial aspect of the arm between the biceps and the triceps muscles (Figure 1).

An ultrasound examination of the right arm revealed the swelling to be arising from the ulnar nerve with mixed echo-texture. No cutaneous lesions suggestive of leprosy were identified in the patient. No slit skin smears were performed. A provisional diagnosis of nerve abscess was entertained and the patient was posted for surgery. On exploratory surgery, a large segment of the right ulnar nerve was replaced with necrotic tissue (Figures 2a and 2b) and no nerve fascicles were discernible even under an operating microscope.

With a plan to bridge the long nerve gap using cable nerve grafts, the involved segment of nerve measuring 8 cm in length was excised till the mid-arm level proximally and till the cubital tunnel distally. The nerve distal to the cubital fossa was also thickened. However, no suitable nerve fascicle could be identified for cable nerve graft anastomosis and primary nerve grafting was not done. The excised tissue was sent for histopathology. Microscopic examination of the excised nerve specimen however, revealed extensive areas of caseous necrosis within the nerve resulting in destruction of the nerve. A moderately dense lympho-histiocytic infiltrate with epithelioid cell granulomas and multinucleated giant cells were also noted within the nerve (Figure 3a). The stain for lepra bacilli was positive Figure 3b.

A final diagnosis of tuberculoid neuritic leprosy with ulnar nerve necrosis was made. Anti-leprosy treatment (multi-bacillary regimen) was re-started in view of presence of lepra bacilli in the nerve biopsy. Six months later, the patient underwent a four-tailed tendon transfer operation to correct his claw hand deformity. At 1 year of follow-up, the patient has maintained good correction of claw deformity, as well as restoration of transverse metacarpal arch. He has not shown any cutaneous signs of leprosy or any new nerve function impairment.

Figure 1. Soft tissue swelling over the medial arm
Pure neuritic leprosy is characterised clinically by peripheral neuropathy manifesting as motor or sensory impairment of single or multiple nerves; occasionally presenting as a thickened nerve, soft tissue mass or rarely as a leprotic nerve abscess. The natural history of PNL involves formation of granuloma in the nerve fascicles, which eventually leads to formation of a localised nerve abscess. This is more common in the pauci-bacillary and tuberculoid forms of the leprosy. There was extensive necrosis of the ulnar nerve fascicles involving the entire ulnar nerve in our patient. Ulnar nerve abscess following successful completion of multi-drug regimen for indeterminate leprosy however, has been previously reported.

It has been observed that in tuberculoid leprosy, T cells breach the perineurium, and destruction of Schwann cells and axons may be evident, resulting in fibrosis of the epineurium, replacement of the endoneurium with epitheliod granulomas, and occasionally caseous necrosis. Interestingly cutaneous tuberculoid leprosy is never associated with caseation. Our patient revealed both granulomas and caseous necrosis on microscopic examination and hence was probably a case of tuberculoid form of neuritic leprosy.

The diagnosis of leprous nerve abscess may be confirmed by the use of ultrasound with or without the use of fine needle aspiration. A histological examination is mandatory for...
confirmation of diagnosis. Once diagnosed, a leprous nerve abscess requires decompression for possible recovery of neural function. Steroid therapy in addition to multi-drug therapy has also been used successfully to reduce the intra-neural edema and may have been beneficial in this case, if it had been prescribed early enough.

In conclusion, pure neuritic tuberculoid leprosy is an uncommon presentation of Hansen’s disease even in endemic populations. This patient probably represents an extreme of the spectrum of pure neuritic tuberculoid leprosy where a very large segment of the ulnar nerve was involved by the lepra bacilli.

References


