

An uncommon presentation of a common disease – Pure neural Hansen’s disease with sinus on skin

Dear Editor,

Leprosy/Hansen’s disease is a chronic granulomatous infection caused by *Mycobacterium leprae* and *Mycobacterium lepromatosis*. It commonly involves peripheral nerves and skin. Occasionally, isolated nerves are involved without skin lesions, called pure neuritic Hansen’s disease. The original definition of pure neural leprosy according to the Indian Association of Leprologists (IAL) five type classification (1982) is absence of any skin lesions. Single or multiple larger nerve trunks/branches may be enlarged and there is sensory loss along the distribution of affected nerves. Slit skin smears are negative. Lepromin reaction is generally positive. Nerve histology could be of tuberculoid, borderline or non-specific type.¹ Neuritis in leprosy is a sub-acute, demyelinating and non-remitting event involving cutaneous nerves and larger peripheral nerve trunks. This presents as dryness, hypoaesthesia, tingling and numbness in the area supplied by the involved nerve, atrophy of the involved muscle, abscess of the nerve and neuralgic pain. Diagnosing such cases is challenging and needs exclusion of other common diseases like tuberculosis, diabetes mellitus, hypertension-induced peripheral neuropathy and neural tumours.² We present a rare case of pure neural Hansen’s disease with sinuses on the skin. A 13-year-old boy presented to our clinic with oozing from the lower arm for 4 months and occasional pain. He complained of tingling in the left hand and weakness of fingers. There was no history of preceding trauma, fever, weight loss or cough. The general condition of the child was good. There was no epitrochlear or axillary lymphadenopathy. Systemic examination was normal. Cutaneous examination revealed puckered plaques with seropurulent discharge and crusting on the medial aspect of the left lower arm [Figure 1]. The left ulnar nerve was beaded and thickened with grade 1 tenderness. There was hypoaesthesia, partial/ulnar claw hand with decreased power and atrophy of hypothenar muscles. Pure neural Hansen’s disease with ulnar nerve abscess and ulnar nerve tuberculoma were the differentials considered. Complete blood picture, erythrocyte sedimentation rate and G6PD levels were normal.



Figure 1: Puckered plaques with crust on the medial aspect of the left lower arm.

Slit skin smear, Mantoux test, Tb Quantiferon gold and pus for culture sensitivity was negative. Patient’s parents didn’t consent for nerve biopsy. A nerve conduction study showed decreased amplitude and increased latency of the ulnar nerve. High Resolution Ultrasound (HRUS) of the left ulnar nerve revealed multiple linear hypoechoic tracts (maximum 10 × 3 mm) extending into the adjacent perineural space and sinus on skin [Figures 2 and 3]. Diagnosis of pure neural Hansen’s disease with sinus was considered and treatment was initiated

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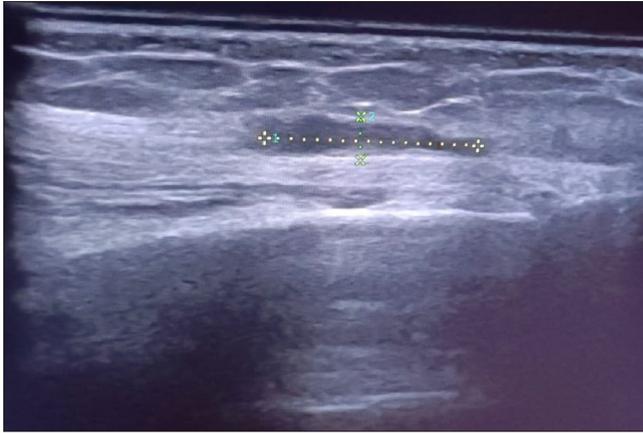


Figure 2: Multiple linear hypoechoic tracts (maximum 10 × 3mm) extending into perineural space along the nerve.

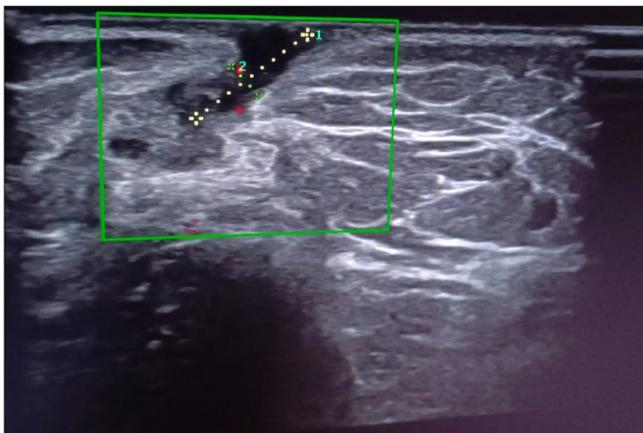


Figure 3: Hypoechoic tract (sinus) extending onto the skin surface (Green square highlights the selected area).

with paucibacillary multi-drug regimen for 6 months. Claw hand improved, discharge subsided and plaques healed with scarring [Figure 4].

Ulnar neuritis with disability frequently occurs in Hansen's disease. Pure neural Hansen's is a diagnostic challenge. Depending upon the patient's immunity, it varies from tuberculoid to lepromatous. Young children and teenagers account for most cases. Ulnar, posterior auricular, peroneal and posterior tibial nerves are most commonly affected.³ However, Mittal *et al.* reported involvement of brachial plexus. As skin lesions are absent in pure neural Hansen's cases, other non-lepromatous aetiologies like compression, trauma,⁴ tuberculosis, diabetes, hypertension, nerve sheath tumours² and rarely neurofibromatosis, syphilis and amyloidosis⁵ must be ruled out. Nerve conduction studies, magnetic resonance imaging, HRUS-guided fine needle aspiration cytology and nerve biopsy with acid fast/Fite-Faraco staining helps in the diagnosis. Nerve abscesses are usually observed in tuberculoid spectrum of pure neural Hansen's disease,¹⁻⁶ but, sinus over skin is very rare. In these



Figure 4: Healed lesions with scar.

cases, ulnar nerve tuberculoma⁷ must be ruled out. Loss of sensation on the left hand, partial claw hand, thickened ulnar nerve and HRUS corroborated the diagnosis in our patient. Early diagnosis is essential to preserve the nerve functions and avoid permanent disability. Multi-drug therapy, oral corticosteroids, nerve decompression, drainage of abscess if needed, splints and physiotherapy are the possible treatment options for neuritis. Lack of histopathology is a limitation in this case.

In a country like India which still accounts for over 50% of the global burden of leprosy cases, any case of hypoesthesia or muscle weakness with thickened nerves must be evaluated in detail to rule out pure neuritic leprosy. We present a case of pure neural Hansen's disease with sinuses on the skin which improved with multi-drug therapy. Partial claw hand improved with splint and physiotherapy subsequently.

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