CASE REPORT

Nerve abscess in pure neural leprosy mistaken for peripheral nerve sheath tumour with disastrous consequence: what can we learn?

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SUMMARY

A 34-year-old Indian man presented to an orthopaedician with gradually progressive hypoesthesia affecting his right lower limb and an ipsilateral common peroneal nerve swelling around the knee. The nerve swelling was diagnosed as a peripheral nerve sheath tumour based on MRI findings and was excised, only to be revealed as leprous nerve abscess on histopathology later. The patient developed right foot drop as a result of common peroneal nerve biopsy. This case presents several learning points in the diagnosis of pure neural leprosy.

BACKGROUND

Pure neural leprosy often poses a diagnostic challenge. As skin lesions are absent, other causes of peripheral neuropathy are also considered in the differential diagnosis. The diagnosis of pure neural leprosy rests on demonstrating the characteristic histopathological changes and/or acid-fast bacilli within the affected nerve. We describe an unusual presentation of

pure neural leprosy, which was misdiagnosed as a peripheral nerve sheath tumour. The nerve swelling was excised, resulting in motor deficit.

CASE PRESENTATION

A 34-year-old, otherwise healthy, man from North India developed spontaneous 'pins-and-needles' sensation and reduced sensation over the dorsum of right foot 5 months back, which gradually progressed proximally to involve his right leg. There was no history suggestive of motor weakness. After a few months, he noticed an asymptomatic swelling on the lateral aspect of his right knee for which he consulted an orthopaedician. An MRI of the right knee showed an elongated fusiform soft tissue swelling (approximately 1.5×2 cm) in continuity with the common peroneal nerve. The swelling was isointense to muscle on T1-weighted (T1W) image and hyperintense on T2-weighted (T2W) image (figure 1). These findings were interpreted as suggestive of

Figure 1 MRI shows thickened common peroneal nerve appearing as an isointense swelling (arrows) to muscle on axial T1-weighted (A) image and hyperintense swelling (arrows) on T2-weighted (B) image.

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a peripheral nerve sheath tumour, and the swelling was surgically excised and sent for histopathological examination.

INVESTIGATIONS

The histopathology showed multiple necrotising epithelioid cell granulomas within the nerve as well as in the surrounding connective tissue (figure 2). Occasional rod-shaped bacilli were identified on auramine-rhodamine stain, but not on Ziehl-Neelsen stain. With a revised diagnosis of leprosy, the patient was referred to us for further management.

TREATMENT

The patient reported sudden-onset weakness in dorsiflexing his right foot noticed after the nerve biopsy. At presentation, he had a 'high-stepping' gait. Complete skin examination did not reveal any lesions suggestive of leprosy. No peripheral nerves were enlarged or tender. There was about 70%–100% reduction in sensations to touch, pain and temperature over the lateral side of right foot dorsum, extending up to the lower one-third of the shin. Apart from the right foot drop, there was no other motor deficit. Slit-skin smear examination was negative. The patient was treated with 6 months of paucibacillary multidrug treatment for pure neuritic leprosy along with physical rehabilitation.

OUTCOME AND FOLLOW-UP

There was no further progression of neurological impairment. The patient is now planned for posterior tibial tendon transfer to correct the foot drop.

Figure 2 H&E stained section of the excised nerve swelling shows well-defined collections of epithelioid cells with few giant cells within the nerve (A, 100X). Foci of necrosis can also be seen (B, 400X).

DISCUSSION

Leprosy remains a common cause of mononeuritis especially in endemic areas, even though it has been 'eliminated' globally. When neuropathy is accompanied by classical cutaneous manifestations of leprosy, its diagnosis is usually straightforward. However, correctly diagnosing pure neural leprosy, where the skin lesions are absent, can be sometimes challenging. Occasionally, as in our patient, the diagnostic dilemma is compounded when the patient presents with only a nerve abscess without thickening of peripheral nerves.¹ As nerve abscesses in leprosy are usually 'cold', they are relatively painless and can be mistaken for a soft tissue mass, a lymph node or a nerve tumour.^{2–4} The gold standard for confirmation of pure neuritic leprosy is nerve biopsy; however, it is limited by sampling errors, low sensitivity and risk of permanent nerve damage.⁵ Though a pure sensory nerve can be biopsied safely, biopsy of a mixed nerve or major nerve trunk can result in motor deficit. Fine needle aspiration of the affected nerve for cytological examination can be a useful 'nerve sparing' alternative. It is safe, less invasive and less time consuming as compared with nerve biopsy, but with a caveat that a negative aspirate does not rule out leprosy.^{6,7} Detection of serum anti-phenolic glycolipid-1 antibodies may also be useful in some patients.⁸ Recently, imaging techniques like ultrasonography and MRI have gained popularity in evaluating nerve involvement in leprosy.^{2,9} As longer nerve segments can be probed with ultrasonography and it is more cost-effective than MRI, high-resolution ultrasonography is preferred initially. Nerves in leprosy are enlarged and exhibit structural abnormalities such as fusiform swelling, loss of fascicles, oedema and increased vascularity on sonography.¹⁰ Ultrasound imaging in a nerve abscess shows diffusely thickened nerve housing a hypoechoic cystic lesion with internal debris,² whereas peripheral nerve sheath tumours appear as well-defined ovoid homogenous hypoechoic lesions with nerve entering and exiting from them.¹¹ MRI may show diffuse oedema and swelling of the involved nerve due to neuritis. Nerve abscess as well as peripheral nerve sheath tumours can appear as a T1 iso-hypointense lesion and T2 hyperintense lesion. Sometimes, peripheral nerve sheath tumours, especially neurofibromas, may show a 'target appearance', as central low intensity surrounded by peripheral high intensity, on T2W images. Postgadolinium sequences can help in differentiating nerve abscess from nerve sheath tumours: a nerve abscess shows peripheral rim of enhancement with a central non-enhancing necrotic region, similar to abscess elsewhere in the body,^{2,3,12} while nerve sheath tumours can have a variable pattern of enhancement: homogenous, heterogeneous or central.¹³

In our patient, postgadolinium contrast sequences were not performed; perhaps possibility of leprosy was not considered by the orthopaedic surgeon given the unusual clinical presentation. A patient with pure neural leprosy may first present to a neurologist, orthopaedician or an internist, instead of a dermatologist. With atypical presentations of leprosy becoming commoner, it has become more important to consider leprosy as a cause of peripheral nerve swelling today than ever before. Imaging modalities like ultrasonography and MRI can aid in the differential diagnosis of peripheral nerve swellings. A fine needle aspiration cytology from the common peroneal nerve could have confirmed the diagnosis, while averting the foot drop. To conclude, this case highlights an uncommon presentation of pure neuritic leprosy and its diagnostic pitfalls.

Learning points

- ▶ Though eliminated globally, leprosy is still prevalent in many parts of the world and should be considered in the differential diagnosis of peripheral neuropathy in patients from these endemic regions.
- ▶ Diffuse peripheral nerve thickening is one of the cardinal signs of leprosy. But sometimes, it may be absent and the patient may present with only a small fusiform nerve swelling.
- ▶ Radiological imaging, especially ultrasonography, may be helpful in differentiating leprosy from other causes of nerve swelling, and their role should be further explored in such settings.
- ▶ Fine needle aspiration cytology is a useful alternative to nerve biopsy in case of motor or mixed nerve involvement, but has a poor sensitivity.

Contributors All the authors were involved in the care of the patient. The manuscript has been drafted by VG and TD, and has been edited by NK. The radiology images were reviewed by CJD. The final manuscript draft has been seen and approved by all the authors.

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