Chapter 13
Annular Erythematous Plaque on the Right Hand

P. K. Ashwini

Abstract Leprosy is an infectious disease primarily involving the skin and peripheral nerves. Most important consequences of leprosy are due to nerve involvement. Neuritis is defined as inflammation of the nerves. The terms neuropathy and neuritis are used interchangeably. Neuritis is defined as pain in the neuro-anatomical area but in conjunction with motor impairment and/or sensory signs in the correspondent nerve, in addition to confirmed demyelinating signs demonstrated in the electrophysiological examination results. Neuropathic pain is defined as pain distribution in a neuro-anatomically plausible area with confirmed negative or positive sensory signs (i.e., hypoesthesia, hyperesthesia, hypoalgesia, hyperalgesia, or allodynia). Neural pain is a common complaint when a leprosy patient seeks medical consultation**.** Neuropathy in leprosy starts with the entry of *Mycobacterium leprae* into the Schwann cells of the small nerve fibers. Clinical examination focusing on the enlargement of peripheral nerves and also the triggered pain by nerve palpation are the clinical signs to predict neuritis. A detailed clinical examination is needed to identify the signs, which can otherwise be missed if the only presenting complaint of the patient is a vague neurological symptom. We hereby present a case who presented with such a trivial neurological symptom which on further clinical examination and evaluation revealed the presence of Hansen's disease. The relevance of clinical examination giving importance to signs is what is being highlighted.

Keywords Leprosy · Neuritis · Neuropathic pain

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S. Pradhan, P. Kumar (eds.), *Clinical Cases in Leprosy*, Clinical Cases in Dermatology, https://doi.org/10.1007/978-3-031-08220-7_13

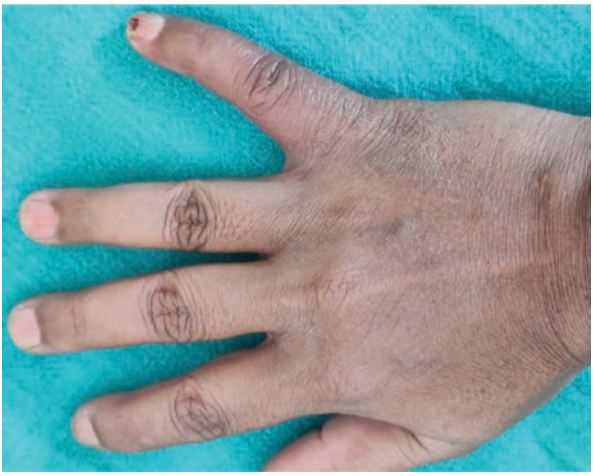

Fig. 13.1 Ill-defined dry erythematous patch over the right little finger extending into the fourth web space

Clinical Presentation

A 37-year-old female presented with a complaint of a vague dragging pain on the right-hand medial aspect for 4 months. She gave history of the low-grade pain, being present at the right elbow and radiating up to the little finger. She had noticed an asymptomatic reddish lesion on the right hand, to which various topical medications were prescribed earlier. There were no known medical comorbidities. On examination there was a solitary ill-defined dry erythematous patch over the right little finger extending into the fourth web space (Fig. 13.1). The patch had no sensory impairment. The right ulnar nerve was thickened and tender. No other peripheral nerve was thickened. Wartenberg's sign positivity was appreciated (Fig. 13.2). Systemic examination was normal.

What Is Your Diagnosis?

1. Resolving cellulitis.
2. Leprosy.
3. Dermatophytosis.

Investigation

- Slit skin smear was negative for acid fast bacilli.
- Skin histopathological examination revealed presence of multiple granulomas composed of epithelioid cells, lymphocytes, and Langhans giant cells around

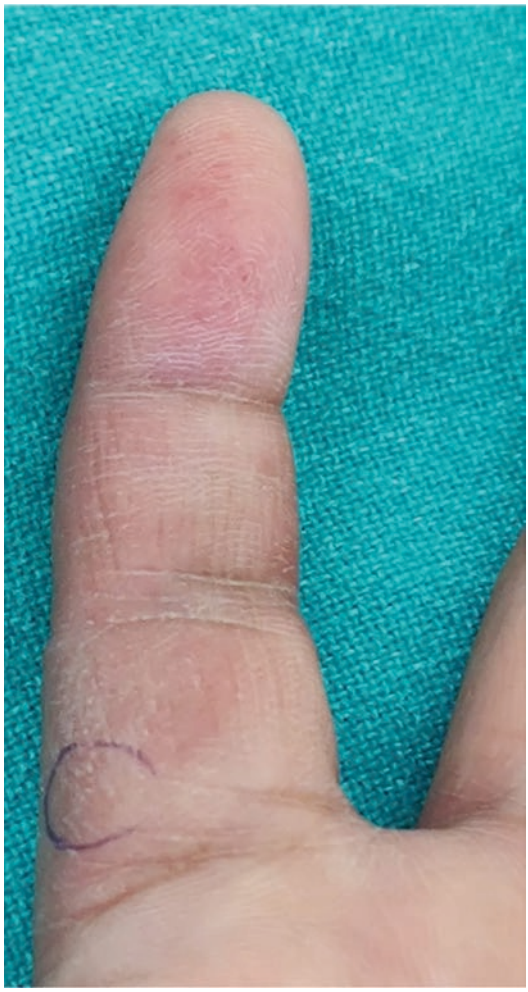

Fig. 13.2 Positive Wartenberg's sign

neurovascular bundles in the superficial and deep dermis. Fite-Faraco stain was negative for lepra bacilli. Features suggested borderline tuberculoid leprosy.

- Ultrasound of the thickened nerve using a 8 MHz probe revealed thickening of the right ulnar nerve 8 mm (Figs. 13.3 and 13.4) compared to the left ulnar nerve 2 mm (Fig. 13.5).
- Nerve conduction studies performed for both upper limbs affirmed abnormalities that were suggestive of sensorimotor axonopathy of the right ulnar nerve. Right ulnar nerve showed reduced compound muscle action potential and sensory nerve action potential amplitude with normal distal latency and conduction velocity.

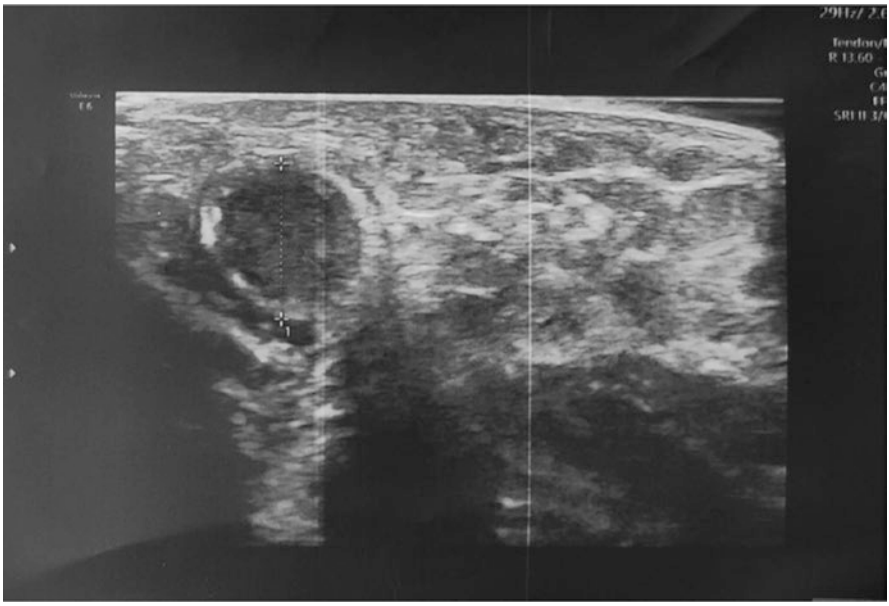

Fig. 13.3 High-resolution USG right ulnar nerve

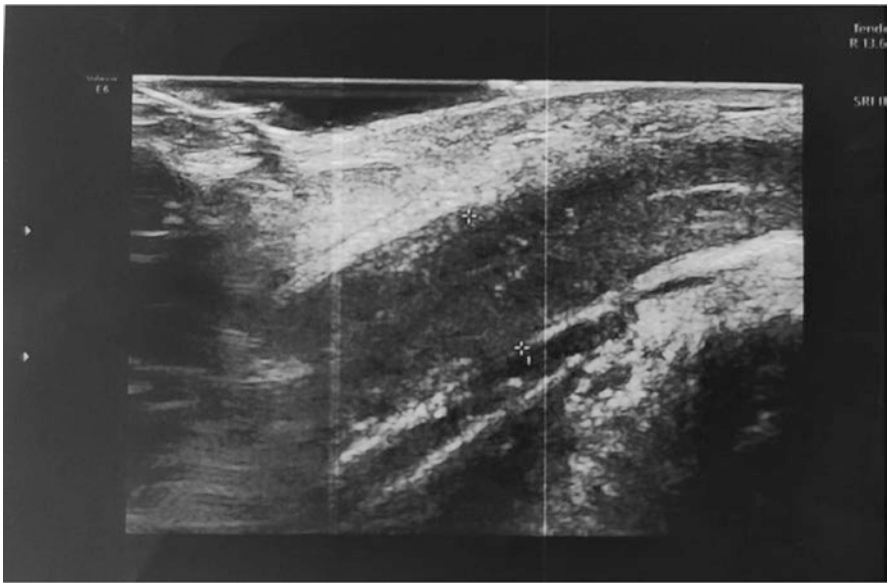

Fig. 13.4 High-resolution USG right ulnar nerve

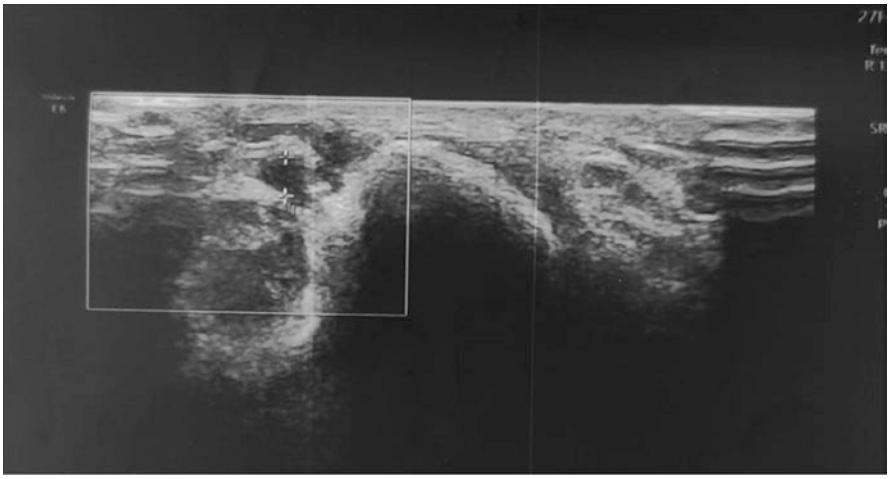

Fig. 13.5 High-resolution USG left ulnar nerve

Final Diagnosis

Borderline tuberculoid leprosy with type 1 reaction with right ulnar neuritis.

Discussion

The involvement of nerves is observed across the clinical spectrum of leprosy, and such cases are said to be having Hansen's neuritis (HN). HN can broadly be classified into four clinical types based on their period of occurrence and type of presentation. However, it should be noted that they are not mutually exclusive and can overlap one another:

1. Neuritis associated with the disease, which is usually chronic and low grade
2. Neuritis associated with reactions, which is usually acute and severe
3. Silent neuropathy or quiet nerve paralysis paralysis
4. Neuropathic pain in leprosy

Neuritis in leprosy is usually a subacute, demyelinating, and non-remitting event involving cutaneous nerves and larger peripheral nerve trunks. Invasion of Schwann cells and axons by *Mycobacterium leprae* leads to demyelination and axonal degeneration. Some of the most significant complications of leprosy occur as a result of the involvement of the peripheral nervous system [1].

One of the earliest nerves to be involved in Hansen's disease is the ulnar nerve. In case of ulnar palsy, the patient is unable to keep all the fingers straight and together, and the little finger tends to stay abducted from the ring finger and may

also be slightly bent or clawed. This is called Wartenberg's sign [5]. The muscle involved is the abductor digiti minimi.

Nerve involvement can be appreciated clinically in most cases. However, in some early cases, other modalities such as nerve ultrasound, nerve conduction studies, and nerve biopsies may be warranted.

The hallmarks of leprosy are nerve enlargement and inflammation. High-resolution ultrasonography (HRUS) can be used for imaging of nerves. Use of high resonance frequency (15–20 MHz) has made it very effective to visualize nerves. HRUS is efficient, user-friendly, and economical. Additional features such as compound imaging and panorama view make high-resolution ultrasonography a superior modality for imaging of nerves. High-resolution ultrasonography demonstrates nerve enlargement, even if subclinical. Inflammation can be detected by color Doppler study of involved nerves which show increased blood flow signals of endoneural and perineural vessels [6].

Nerve damage in leprosy may present as silent neuropathy without overt signs and symptoms or clinically manifest with weakness, atrophy, or contracture. Common methods used to detect sensory nerve function impairment are monofilament testing and ballpoint testing. For detection of motor function impairment, voluntary muscle testing is performed.

Functional derangement of nerves can be detected by nerve conduction studies before the appearance of clinical signs and symptoms. Disability and deformity could be minimized if nerve function impairment is detected and treated early. Nerve conduction studies involve the recording, display, measurement, and interpreting of action potentials arising from the peripheral nerves.

Nerve is stimulated through the skin with a surface stimulator or through a needle placed close to a nerve or a nerve root. Motor studies are performed by electrical stimulation of a nerve and recording the compound muscle action potential (CMAP) from surface electrodes overlying a muscle supplied by that nerve.

Conduction velocity (CV) is calculated by dividing the length of the nerve segment between the two stimulation points by the difference between the proximal and distal latency. Latency is the time from stimulus artifact to the onset of the response. The sensory nerve action potential (SNAP) is obtained by electrically stimulating sensory fibers and recording the nerve action potential at a point further along that nerve [7].

The electro-neuro-myographic pattern of leprosy neuropathy is described as the impairment of conduction of nerve impulse and decreased amplitude of sensory-motor potentials. Along with reduction in nerve conduction velocity, changes in latency are also reported. The slowing of sensory conduction velocity might show no difference between tuberculoid and lepromatous patients. In addition, a significant slowing of nerve conduction has also been reported in clinically normal nerves in leprosy. However, studies have reported the absence of correlation between neurological symptoms and electroneurographic studies in leprosy patients. A comprehensive electrophysiologic, ultrasonographic, and histological evaluation may be helpful in establishing a diagnosis of leprosy, where the presentation is more with neural symptoms.

In our case, multibacillary multidrug therapy (MB-MDT) was started. NSAIDs were given for neuritis. After 4 weeks of MB-MDT, she presented fever, fatigue, and reddish rashes over the body. On examination, there were erythematous maculopapular eruptions involving the face, trunk, and extremities. Drug hypersensitivity syndrome to dapsone was considered. Investigations confirmed the same with altered liver enzymes and eosinophilia. She improved with discontinuation of dapsone and injectable steroids. Patient was then started on ROM therapy and is being followed up.

Conclusion

Neurological symptoms commonly accompany cutaneous lesions in leprosy. Occasionally there can occur only neuritis at time of presentation without any overt clinical signs of cutaneous involvement. This may lead to significant delay in the diagnosis. Most deformities observed in leprosy are a consequence of delayed diagnosis of underlying nerve damage. The importance of prompt clinical and neurological examination is highlighted in the case.

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