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Case Report

Ultrasonographic Features of Ulnar Nerve Affected by Hansen's Disease

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Leprosy continues to be a major public health problem in some areas of our country. It predominantly afflicts peripheral nerves and skin and may lead to deformities. Social stigma as a result of deformities further plagues the situation. Prompt and early diagnosis coupled with adequate treatment, concurrent rehabilitative strategies if deformities do occur, and health education help to control the problem. Definitive diagnosis of leprosy has traditionally been based on assessment of slit skin smears (SSS) after AFB staining and characteristic histopathology after biopsy of the lesion. However, recently, thickening of the peripheral nerves has been demonstrated by ultrasonography and this can be used as a sensitive tool to assess and measure enlargement of peripheral nerves, which are hallmarks for leprosy especially in clinical settings. In this report, the ultrasonographic findings of ulnar nerve enlargement due to leprosy in a fourteen-year-old male patient are described.

Key words : Ultrasonography, ulnar nerve, leprosy

Introduction

Mycobacterium leprae, the causative organism of leprosy thrives in cooler regions (Park 2015) of body namely skin (except axillae, groins), peripheral nerves, anterior chamber of the eye, upper respiratory tract and testes (Gelber 2008, Park 2015). One of the important signs and diagnostic features of leprosy includes enlargement of nerves. This enlargement of the nerves may be observed in a few hereditary conditions as well (Elias et al 2009). Thickened nerves in leprosy are damaged during the course of the disease and its immunological reactions and are the major cause of deformities including claw hand, foot-drop and trophic ulcers (Gelber 2008). Clinically, thickened nerves may be palpable, however, this may be subjective (Elias et al 2009, Jain et al 2009, Jain et al 2013). Moreover, not all nerves are amenable for palpation and consequently the nerve enlargement may not

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be detected. Ultrasonographic evaluation of thickened nerves is a viable and non-invasive alternative. Moreover, unlike magnetic resonance imaging, sonography is cost effective which make ultrasonography a preferred modality for examination of the peripheral nerves (Elias et al 2009, Jain et al 2009, Jain et al 2013).

Case Report

A 14-year-old male presented with a few hypopigmented, erythematous plaques on the left upper extremity since twelve months. He also complained of tingling and numbness along the ulnar nerve distribution since a few months.

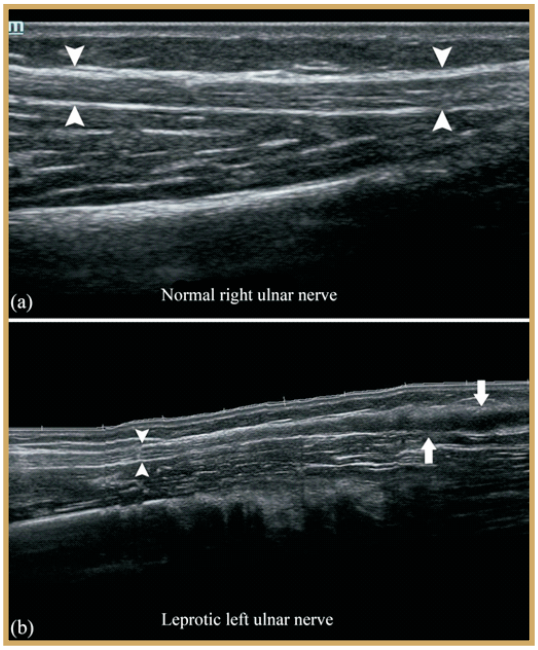

Fig 1 : Ultrasonography panoramic view reveals normal right ulnar nerve with hypoechoic nerve fascicles and intervening hyperechoic perineurium giving the bundle of straws appearance (arrowheads in a). The left ulnar nerve shows fusiform enlargement with loss of fascicular architecture and hypoechogenicity distally (arrows in b; the arrowheads point towards normal segment of the nerve).

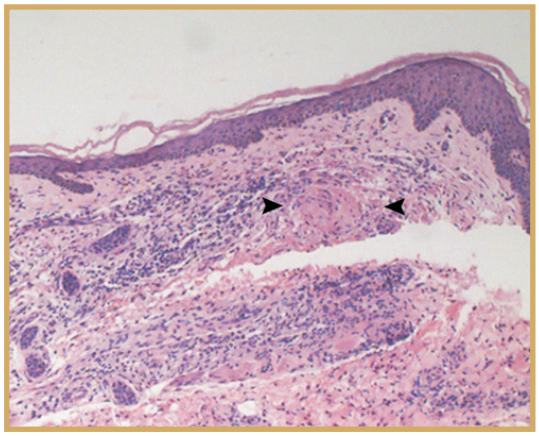

Fig 2 : Photomicrograph (hematoxylin and eosin stain) from the periphery of the hypopigmented patch demonstrates thinning of the epidermis. There is presence of ill-formed granuloma with lymphocytic infiltration in the subepithelial tissue (arrow heads).

The general examination was normal. Local examination revealed weakness and wasting of the hypo-thenar muscles of the left hand. There was also diminution of sensations on the medial aspect of the left hand. On palpation, thickened, cord-like ulnar nerve was palpated behind and above the medial epicondyle. Ultrasonographic evaluation demonstrated fusiform thickening of a long segment of the left ulnar nerve with loss of fascicular morphology (Fig 1). The contralateral nerve was normal. Slit-skin smears from ear lobes and skin lesions were negative for acid fast bacilli. However, histopathology was consistent with leprosy, with involvement of the skin as well as a twig of the peripheral nerve (Fig 2). The patient refused to undertake nerve conduction studies. Based on clinical, pathologic analysis and ultrasonographic features, a diagnosis of tuberculoid leprosy (with involvement of left ulnar nerve and associated wasting of hypothenar muscles) was thus established. The patient

was started on MDT (multidrug therapy) for PB (paucibacillary) leprosy. The regimen included administration of 100 mg of Dapsone daily with 600 mg of Rifampin once a month (supervised) for a total of six months. At four months of follow-up, the patient is compliant with the chemotherapy and there is mild reduction in symptoms.

Discussion

The history of Leprosy is as old as that of mankind and is still not fully understood. It has a long and variable incubation period which on an average is believed to be three to five years but may be as long as twenty years (Park 2015). The organism cannot be grown on artificial culture media, which adds to the lack of definitive tests for diagnosis of the disease. Leprosy is a chronic, non-fatal disease and the bacillus has low pathogenicity. The lepra bacillus is less infectious than tubercle bacillus (Park 2015). Leprosy has been known to be associated with poverty and poor hygienic conditions. Animal models for the disease include armadillos, mangabey monkeys and chimpanzees. These animals, however, are not a threat to transmission of the disease (Gebler 2008 and Park 2015).

Epidemiologically, the disease has a male predilection. No age is exempt from the disease; however, the highest incidence is noted in the second and third decades of life. The youngest detected case was a two-and - a half months old child from South India (Park 2015). Although the exact portal of entry of the bacillus into humans is debatable, higher levels of mucosa associated-IgA in nasal mucosa of close contacts supports the postulate that droplets (containing the *M leprae*) area potential mode of spread. Besides, indirect transmission via moist soil and insect-bite have also been incriminated (Park 2015, Gebler 2008). Once in the host, the Mycobacterium has a propensity to invade Schwann cells (Elias et al 2009). Most of the exposed individuals do not manifest the disease and self heal while a small percentage who lack the specific immunity to it manifest the disease in their lifetime. An interplay of cell-mediated immunity of the host and the organism decide the severity of the disease. In tuberculoid forms, host immunity is relatively preserved with only a few skin lesions and asymmetric involvement of a few peripheral nerves. There can also be complete spontaneous resolution of the disease. On the contrary, a deficient specific cell mediated immunity, malnutrition, inter current illnesses may precipitate the occurrence of multiple anaesthetic skin lesions with multiple nerve involvement which are characteristics of the more severe forms of the disease. The skin lesions are typically hypopigmented and or erythematous, and may be macules, patches, plaques or nodular in configuration with varying degrees of loss of sensation. Affection of autonomic nerve fibres by lepra bacilli may cause the skin lesions to be anhidrotic and scaly. Notably, Pure neuritic form of leprosy, found in Indian subcontinent does not have any dermatological manifestations (Jain et al 2013).

Leprosy is usually diagnosed by demonstrating presence of acid fast bacilli in skin smears from ear lobes, and growing edge of the skin lesions Besides, it is also diagnosed in the skin biopsy by granuloma formation within the nerve bundle (pathognomonic for the disease), macrophages and lymphocytic infiltration and presence of AFB in the specimen. Nerve involvement is assessed by examination of the patient, clinical palpation and sometimes electrophysiological studies. Clinical palpation is however, subjective and a few deep seated nerves may be difficult to palpate. Moreover, nerve conduction studies are painful, expensive and not available routinely

in all hospitals. Keeping in view the limitations of clinical palpation and electrophysiological studies, ultrasonographic evaluation of nerves is a good non invasive, inexpensive, and less time consuming alternative.

Ultrasonographic characterisation of a normal nerve depicts fine hypoechoic nerve fascicles separated by echogenic epi-perineurium. This characteristic appearance of nerves is called 'honey-comb' and 'bundle of straws' on transverse and longitudinal planes, respectively. Leprosy affected nerves tend to become oedematous and thickened. The fascicular morphology is however, preserved. At times, resolution of individual fascicle is not possible and the nerve is transformed into a hypoechoic structure with fusiform thickening (complete loss of internal architecture). Enlargement of nerves can also be seen in amyloidosis, and a few hereditary neuropathies (Elias et al 2009, Jain et al 2009 and 2013). The distinguishing features of leprosy however, are an extensive enlargement along a long segment of the nerve and maximal thickening proximal to osseo-fibrous tunnels. This may be associated with increased vascularity which is a characteristic of leprosy associated neuritis and lepra reactions. Echogenic foci representing fibrosis within the nerve and nerve abscesses may also be seen (Elias et al 2009). Magnetic resonance imaging of the nerve may also be performed but is associated with longer acquisition time, need to perform the scan along the long length of the nerve, increased cost, patient discomfort, and expensive nature of the modality. All this makes ultrasonography a preferred technique for examination and investigation of peripheral nerves in leprosy.

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