Neuritic Leprosy; An Intriguing Re-visit to a Forbidden Ailment

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ABSTRACT

Leprosy is a chronic infectious disease that presents with varied manifestations. Pure neuritic leprosy is one of the rarest forms of the disease which is characterized by nerve involvement without the characteristic cutaneous stigmata.

Eleven year old, healthy male presented with progressively increasing painful swelling at the medial aspect of the arm near to the right elbow joint with difficulty in extending right ring and little fingers at interphalangeal joint and numbness in the same region for last 1 year with no cutaneous abnormalities. Physical examination revealed 6x3 cm firm, tender lesion 3 cm proximal to the right elbow joint with positive tinel's sign, without signs of inflammation, along with characteristic claw hand deformity of right hand and atrophy of hypothenar and interossei muscle. Electro-diagnostic testing revealed findings consistent with a right ulnar axonal neuropathy above the elbow. Magnetic resonance imaging revealed well defined heterogeneously hyper intense linear lesion along the course of thickened ulnar nerve in the distal arm extending posterior to the medial condyle. It also showed an oval shaped lesion (2.1x1.0 cm) arising from the same segment of the nerve, without any bony or muscular involvement of that area. The patient underwent surgical exploration and ulnar nerve decompression with biopsy. Pathology revealed necrotizing granulomatous inflammatory acid fast bacilli stain negative lesion, which was histologically consistent with caseous abscess caused by tuberculoid leprosy, pathognomonic for Hansen's disease. He has been started on antibiotic therapy and is referred to leprosy center for further course of management.

Pure neuritic leprosy, though rare, should be considered as differential diagnosis in cases presenting with peripheral neuropathy at leprosy-endemic areas. Prompt diagnosis and treatment is imperative to prevent permanent neurological injury.

KEY WORDS

Leprosy, Neuropathy, Ulnar nerve

INTRODUCTION

Leprosy was first described in 1873 by Dr. Gerhard Armauer Hansen.[1] The causative organism 'Mycobacterium leprae' an acid-fast bacterium, spreads by airborne droplets. It primarily infects Schwann cells in the peripheral nervous system as well as histiocytes in the dermis.[2] Manifestation of the disease varies as it depends on the host immune response. Leprosy involves major nerves at and above locations surgeons commonly address for compressive neuropathy such as the ulnar nerve at the elbow, tibial nerve at the ankle, median nerve at the wrist, and peroneal nerve at the knee.[2] In the pure neuritic form of leprosy, there are no significant cutaneous manifestations, making diagnosis particularly challenging.[3]

CASE REPORT

An 11-year-old, healthy male with progressive painful right ulnar nerve swelling presents with disability of not being able to properly extend the ring and little finger of the same hand. He also gives history of progressive wasting of his hand muscles and also associated paresthesias followed by numbness over the little finger and medial half of ring finger on supinated forearm. He had no history of neck or radicular pain, denied recent trauma or viral illness, and had no family history of neurological disease. He was delivered at term by normal delivery and lives in the terai region of Nepal. His medical history was otherwise unremarkable, and his immunizations were up to date.

Physical examination revealed 6x3 cm firm, tender lesion with positive tinel's sign, without signs of inflammation, 3 cm proximal to the right elbow joint along with characteristic claw hand deformity of right hand and atrophy of hypothenar and interossei muscles. The ulnar nerve was palpably thickened proximal and distal to the area of interest. He had loss of pin prick and temperature sensation in the ulnar distribution of the hand. Motor exam revealed ulnar palsy with minimal function of intrinsic hand muscles innervated by the ulnar nerve and significant weakness in the flexor digitorum profundus to the fourth and fifth digits with associated clawing. Median and radial motor function was normal. (fig. 1 a, b, c, d)

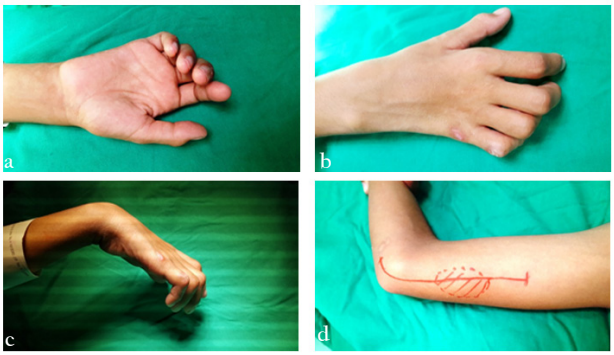

Figure 1. (a-c) Shows wasting of hypothenar muscles, hollowing of the areas on dorsal aspect of the hand signifying wasting of interossei muscles with significant clawing of the hand respectively. (d) Shows location of the swelling and proposed marking of the exploration of the nerve.

On MRI studies, it revealed well defined heterogeneously hyperintense linear lesion (8.6 cm in length) along the course of thickened ulnar nerve in the distal arm proximal and lateral to the olecranon. There is an oval shaped 2.1x1.0 cm lesion arising from the same segment of nerve. It was hypo-isointense to muscle on T1WI, intermediate to hyperintense in T2WI and hyperintense on STIR sequences. Coronal STIR sequences shows nerve thickening and edema in the proximal and distal segment of the nerve with respect to the lesion. Axial DWI shows diffusion restriction in the lesion and focal areas of thickened nerve. The lesion appears separate from bone and surrounding muscles. (fig. 2 a, b, c, d)

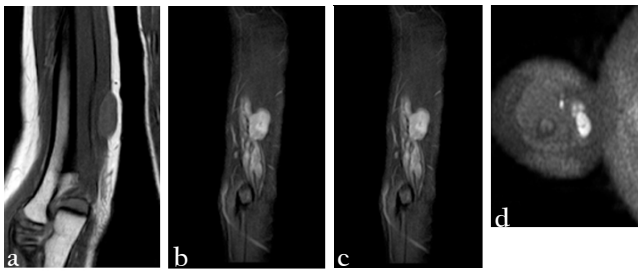

Figure 2. (a) Coronal T1WI shows well marginated ovoid lesion along the course of ulnar nerve proximal and lateral to olecranon. The lesion displays isointensity signal in T1WI and intermediate to hyperintensity signal in T2WI. (b) Nerve thickening and edema is seen in segment of Ulnar nerve proximal and distal to the lesions in coronal STIR section. (c) The lesion shows bright signal in axial STIR. (d) Axial DWI shows diffusion restriction in the lesion and focal areas of thickened nerve.

The ulnar motor potential at the abductor digit minimi (ADM) muscle was significantly reduced in amplitude at 0.2 mV (left 10.8 mV). The take off latency for wrist stimulation considering stimulation to ADM muscle was increased. However peak amplitude and take of velocities were reduced for wrist stimulation as well as stimulation of above and below elbow segment of right ulnar nerve. The right ulnar as well as left radial sensory potential were found to be outside the specified normal range for peak amplitude upon stimulation of forearm. The electrophysiological studies result was consistent with right ulnar axonal neuropathy.

With the background of no cutaneous manifestation pointing us towards leprosy and radiological evidence suggesting the diagnosis of ulnar schwannoma, surgical exploration and excision of tumor of ulnar nerve was planned. However with the clinical findings of ulnar nerve thickening, leprosy was kept into consideration as the possible differential diagnosis.

At surgery, the ulnar nerve above the elbow was thickened and firm with exophytic lesion of about 2x2 cm arising from the medial aspect of the thickened portion of the nerve. A relative constriction at the cubital tunnel was also identified and released. Using electrophysiological monitoring and microscopic dissection, we removed the exophytic lesion along with the caseous abscess within it and also a small nonfunctioning fascicle for pathological evaluation. (fig 3 a, b, c, d)

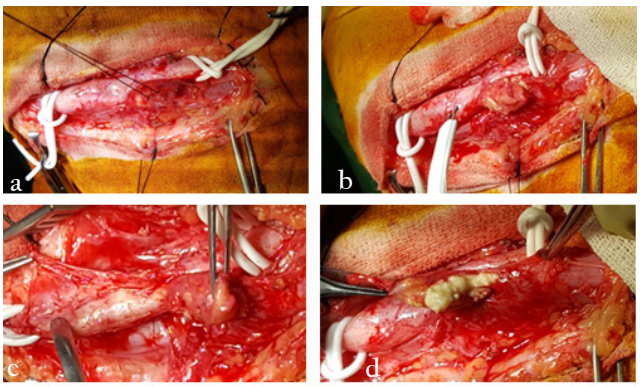

Figure 3. (a,b,c,d) Showing delineation of the lesion alongside the thickened ulnar nerve, neuro-monitoring with nerve stimulation, neurolysis along with dissection of the lesion at interest and evacuation of the cheesy pus material from the lesion of interest respectively.

Histopathological findings of specimen revealed large expanse of caseous necrosis bordered by a rim of diffuse lymphohistiocytic infiltrate along with frequent arrangement of small epitheloid cell granuloma sprinkled with and surrounded by lymphocytes. However no mycobacteria were noted by both ZN as well as modified fite-faraco stains. Hence in the background of necrotizing granulomatous inflammatory lesion though acid fast bacilli stain negative, the result was histologically consistent with caseous abscess caused by tuberculoid leprosy. (fig. 4 a,b)

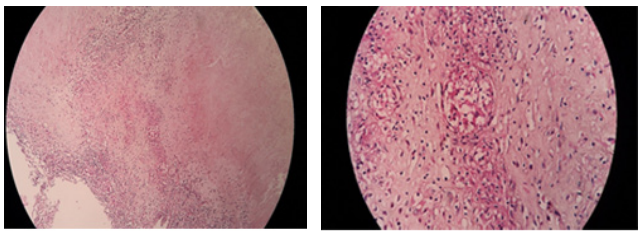

Figure 4. (a) Large expanse of caseous necrosis bordered by rim of diffuse lymphohistiocytic infiltrate along with small epitheloid cell granuloma sprinkled with and surrounded by lymphocytes. (b) Characteristic small epitheloid cell granuloma sprinkled with and surrounded by lymphocytes.

After rendering a diagnosis, we planned to start the patient on a 2-drug regimen including clofazimine and dapsone for required duration of therapy. Postoperatively, the ulnar distribution numbness had resolved with no improvement in clawing. However, mild improvement was noted in the flexor digitorum profundus and ulnar interosseous muscle weakness. Patient was referred to the leprosy center for further medical treatment and management. He is in continued follow up at our institute for any post surgical issues.

DISCUSSION

Nepal has achieved the milestone in elimination of leprosy as a public health problem in December 2009 and declared its elimination in 2010. Yet, few regions from terai belt have been identified as high prevalence area like Jhapa, Morang, Parsa, Dhanusa and Kapilvastu till date. Though, there are programs running with the aim to reduce the incidence of new case and prevalence rate there has been an increasing trend in detection of new leprosy cases during fiscal year 066/67 to 070/71 with more than 3000 new cases being detected each year.[4]

Concerning this case, the diagnosis of leprosy seems not obvious; however, the pure neuritic form of the disease can come into view with multitude of presentation which may complicate the diagnosis. Even when patient was residing at the endemic region, his father notes that the patient had no known contact with any infected individual. Electrodiagnostic studies were helpful in delineating the extent of nerve involvement but were not specific to any particular diagnosis. The imaging studies were interpreted as being consistent with Schwannoma, hence patient's family was counseled in line for surgical intervention.

It has been a topic of controversy and continuous debate that whether neurolysis or any form of decompressive surgery has any role as treatment in leprosy.[5] With predilection of mycobacterium leprae to larger nerves in cooler location and ulnar nerve being the most common nerve involved in tuberculoid leprosy, features of ulnar nerve compression neuropathy is not a rare occurrence in patients with leprosy.[6]

In cases of pure neuritic form of leprosy, without any specific distinguishing feature to differentiate it from compressive neuropathy and also owing to lack of any specific non invasive test to guide further management in line of leprosy, biopsy of the involved nerve remains as only tool to establish the histological diagnosis.[7,8]

Considering pathophysiology in pure neuritic leprosy, fibrotic epineurium along with external compression to the nerve creates relative venous obstruction, capillary stasis, edema, and ischemia leading to features of focal neurological deficit. Thus, external decompression and epineurotomy had been an effective tool to neurosurgeons to choose in order to improve the overall outcomes in patients with leprosy.[9] We also decompressed the ulnar nerve at the time of biopsy in our case to alleviate any contribution of compression to his deficit, to prevent potential worsening compression in the face of swelling related to the biopsy, and to theoretically maximize his neural regeneration.[10,11]

In our patient, misled by current diagnostics as ulnar nerve schwannoma, surgical exploration and biopsy meant critical in making the diagnosis and initiating definitive medical treatment. The lesson learnt was not to undermine one's clinical appraisal of the disease in this technology led era of diagnostic medicine. Had we been able to make the diagnosis of leprosy in this case without the biopsy, we would have recommended initial treatment with multidrug therapy and steroids and would not necessarily have intervened surgically unless he had pain and other symptoms would have deteriorated further even with medical therapy.

Even when there are various reports suggesting various benefits of surgical decompression, including improvement in neurological status, less deformity, and alleviation from pain, the true effectiveness of surgery is yet to be proven.[9] The literature relating to this issue has been summarized in a Cochrane review last updated in 2012. In that review no clear statistical benefit from surgical decompression could be demonstrated based on the existing literature, and recommendations have been made for careful randomized controlled trials.[12]

The multitude in nature of the host response, spontaneous recovery without treatment, clinical worsening during and even months after successful treatment, variable number of nerves involved with their ability to recover from injury and constraints with our ability to obtain meaningful outcome measures even when we provide care to these disease scenario in numbers makes the likelihood of obtaining a legitimate answer regarding surgical decompression in a randomized multicenter trial is essentially nil. Hence till the concluding answers are out, treatment of the individual cases are to be based on current evidences and case to case basis for the betterment of the patient's symptom.

In this technology led era of diagnostic medicine one should not undermine one's clinical appraisal of the disease. Basic core of medicine (history and examinations) are the most efficient first line tools when applied with utmost sincerity always to leads to the diagnosis. Considering, role of surgical decompression as treatment of neuritic leprosy, no definitive literatures are available to guide treatment. However with the background theoretical benefits of alleviating deficit and maximizing neural regeneration, role of surgical decompression cannot be totally ruled out. Even when medical management is the first line of treatment, surgical measure is to be considered wisely as individual case to case based judgment for the betterment of the patient.

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