Case Report

Fibrolipomatous hamartoma of median nerve: A rare cause of carpal tunnel syndrome and macrodactyly

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ABSTRACT

Neurogenic fibrolipomatous hamartoma (FLH) is a benign lesion that can affect any of the peripheral nerves, causing significant enlargement. Though the pathology is a rare one, median nerve is the most common nerve in the body to be affected. The most common segment of the nerve to be affected is that around the wrist. We present a case of carpal tunnel syndrome caused by FLH of the median nerve associated with macrodactyly.

Keywords: *Carpal tunnel syndrome, fibrolipomatous hamartoma, median nerve*

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Introduction

Neural fibrolipoma, also known as fibrolipomatoushamartoma (FLH), is a rare, slow-growing, and benign tumor of a peripheral nerve, most often occurring in the median nerve of young patients.[1] The pathology affects the epineurium and perineurium, which are thereby are thickened and distorted due to proliferation of fibrofatty tissues. The hamartoma so formed compresses the neural elements circumferentially as well as infiltrates between them, thereby causing a sort of compartment syndrome within the nerve. According to the new World Health Organization (WHO) classification of tumors, the pathology is designated as nervous lipomatosis.[2] FLH of a nerve has also been reported as lipofibroma, fibrolipomatous nerve enlargement, lipomatosis, fibrofatty overgrowth, fatty infiltration of the nerve, and neurolipoma. The tumor is sometimes associated with macrodactyly and lipomatousmacrodystrophy of muscles and subcutaneous fat in the region supplied by the affected nerve.[3,4]

Case Report

A 21-year-old right-handed lady was referred to our department, formagnetic resonance imaging (MRI), with complain of paresthesias in the right hand, in the distribution of the median nerve, with macrodactyly. Findings on physical examination revealed a palpable mass on the distal volar aspect of forearm, atrophy of the thenar eminence with macrodactyly [Figure 1]. Tinel's sign and Phalen's test were positive all over the mass. Neurological deficits consist of paresthesia of the middle and ring finger at the time of compression of the mass, and the two-point discrimination test was about 4 mm in the median territory (at the pulp). No lymphadenopathy was detected. MRI showed generalized bulkiness of the right median nerve with increased signal intensity on short TE/TR sequences suggesting a fatty infiltration of the nerve [Figure 2a]. The fat suppressed sequence with long TE/TR; however, revealed compressed nerve bundles in between the fibrofatty elements [Figure 2b-d]. No significant enhancement was noted on administration of intravenous gadolinium. A sonography done for correlation revealed extensive proliferation of abnormal soft tissue having intermediate echogenicity [Figure 3a and b]. The normal nerve fibrils were interspersed in between the abnormal soft tissue and looked compressed. The pathology involved the entire length of the median nerve, in the distal third of the right forearm, dissociating the nerve fibers. A possibility of FLH was kept and the patient was taken up for fasciotomy, which relived her symptoms. A biopsy specimen taken during surgery confirmed the possibility given on imaging, as there was presence of abnormal fibrofatty tissue within the nerve sheath on histopathology.

Discussion

FLH is a benign tumor composed of hypertrophied fibrofatty tissue

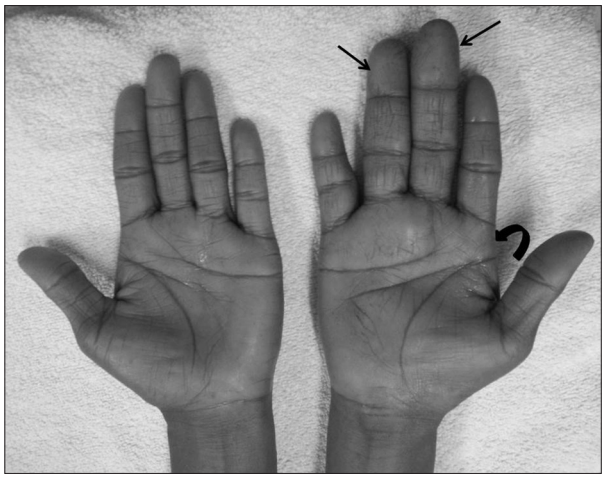

Figure 1: Frontal view of the palms shows atrophy of the thenar muscle, characterized by loss of thenar muscle eminence (curved arrow). Associated soft tissue hypertrophy is seen along the distribution of median nerve in right hand (straight arrows)

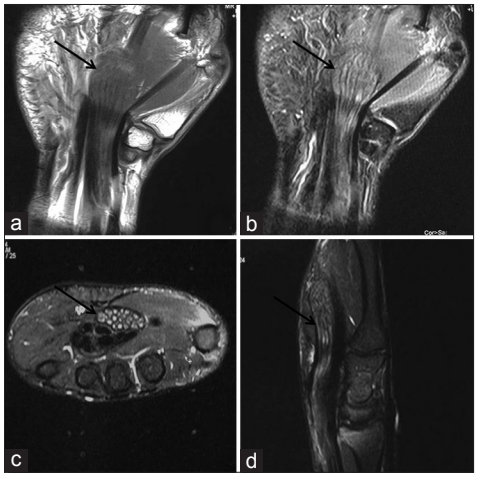

Figure 2: Magnetic resonance imaging of the patient shows hypertrophied soft tissue with intermediate signal and intervening areas of bright (fat) signal of coronal short TE/TR images (a). Coronal, axial, and sagittal fat suppressed long TE/TR images (b-d) shows suppression of the abnormal intraneural soft tissue with compressed bright nerve fibrils giving a typical "spaghetti appearance of gigantic median nerve"

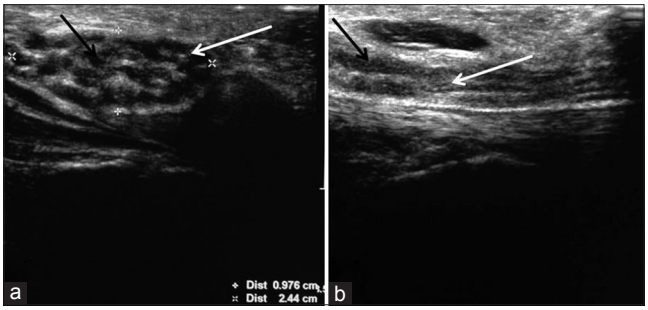

Figure 3: Transverse (a) and sagittal (b) high resolution sonography at the level of wrist showing enlarged median nerve with brighter hypertrophied abnormal intraneural soft tissue (white arrow). Note the compressed nerve fibrils showing relatively lower echogenicity within (black arrow)

intermixed with neural tissue. The condition was first described by Morley in 1964 as intraneurallipoma.[5] The median nerve and its branches are the most commonly involved, followed by the radial nerve, ulnar nerve, nerves at the dorsal aspect of the foot, and brachial plexus.[3] The patient in the present case had involvement of median nerve segment at wrist, which has been mentioned as the most commonly affected segment in most cases.[6,7] The cranial nerves may rarely be affected as well, with symptoms of compression.[3,7] As in the present case, the most common clinical features consist of sensory and motor symptoms, resulting from compression of neural elements.[5,6] Focal gigantism of the involved anatomical part may also be seen, as was evident by the presence of macrodactyly, in the present patient. Macrodactyly is seen into two-thirds of the cases with involvement of median nerve.[3,8] The differential diagnosis of unilocal fusiform nerve enlargement comprises FLH, lipomas within the nerve sheath, and segmental or plexiform neurofibromatosis. In our patient, MRI disclosed serpentiform nerve fascicles surrounded and separated by fibrous and fatty tissue within the expanded nerve sheath as typical features of FLH.[4,6] FLH can clearly be distinguished from lipomas within the nerve sheath, which are characteristic focal masses that dislocate and compress the normal nerve bundles,[8] and from segmental and plexiform neurofibromatosis, in which the neurofibroma has MRI signal characteristics of soft tissue and not of fat.

Apart from documentation of the imaging appearances of this rare entity, the present case emphasizes the importance of systematic differentiation of hamartomatous neural lesions. This remains important because focal lesions like lipomas are treated by enucleation, while systemic conditions like neurofibroma are observed unless causing gross compression. Tenotomy and fasciotomy remains the treatment of choice for FLH.

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