Case Report

Imaging features on sonography and MRI in a case of lipofibromatous hamartoma of the median nerve

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Abstract: Lipofibromatous hamartomas are rare benign tumours of Peripheral nerves. Median nerve is most common affected nerve involved in about 80 percent of the cases. Approximately 92 cases have been reported so far. We present a case of lipofibromatous hamartoma of median nerve diagnosed on sonography and magnetic resonance imaging (MRI). These rare lesions are an important entity to be known to radiologists because their imaging features are quite pathognomonic and allow for confident diagnosis negating the need for biopsy.

Keywords: Lipofibromatous hamartoma; median nerve; sonography; magnetic resonance imaging (MRI)

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Introduction

Lipofibromatous hamartomas (also known as intraneural lipoma, perineural lipoma, fibrolipoma, fibrofatty overgrowth) are rare congenital lesions that most commonly affect the median nerve at the level of wrist (1). We present another case of this uncommon lesion. There are case reports with emphasis on magnetic resonance imaging (MRI) features but very few have described sonographic findings as well which is an equally useful and very convenient modality for the diagnosis.

Case report

A 19-year-old man presented with painless swelling on volar aspect of right wrist since birth which is gradually increasing in size and became painful since last five months. On physical examination there was atrophy of thenar eminence with loss of flexor pollicis longus muscle along with a smooth tender lump on volar aspect of wrist (*Figure 1*). There was decreased grip strength. No other neurological signs of sensory or motor dysfunction were seen. MRI of left wrist with hand was advised which showed thickened hypointense nerve fascicles interspersed within hyperintense fibroadipose tissue on both T1 weighted and T2 weighted images giving characteristic coaxial cable like appearance on axial images (*Figure 2*) and spaghetti string appearance on coronal images (*Figure 3*). Lesion measured 2.7 cm × 2.2 cm × 14.2 cm in size and shows suppression of hyperintense signal more pronounced in distal part of the lesion on STIR coronal image suggestive of fatty tissue (*Figure 4*). Ultrasound was done in retrospect after MRI by our team and showed pathognomonic findings of thickened hypoechoic axonal bundles surrounded by echogenic fatty substratum (*Figure 5*). Color doppler showed no evidence of flow in the mass (*Figure 5*).

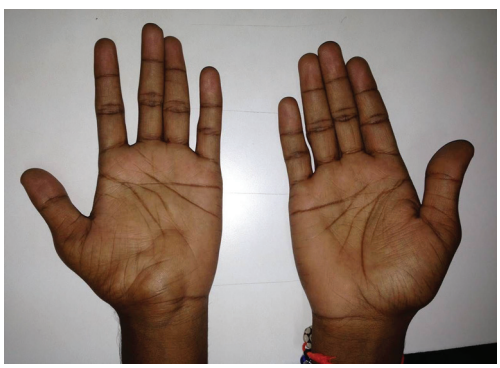

Figure 1 Photograph of both wrists with hands shows lump on volar aspect of left wrist extending in left hand and thenar muscles atrophy on left side (compared from right hand).

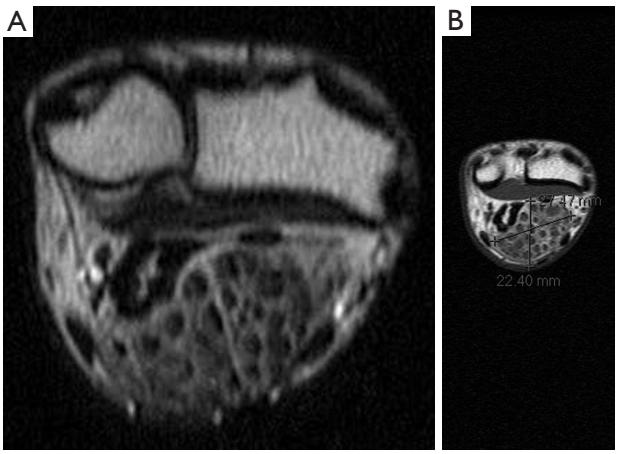

Figure 2 Axial T1 (A) and Axial T2 (B) weighted image of wrist shows thickened hypointense nerve fascicles surrounded by hyperintense fibroadipose tissue giving "coaxial cable" like appearance. Lesion measures 2.7 cm × 2.2 cm.

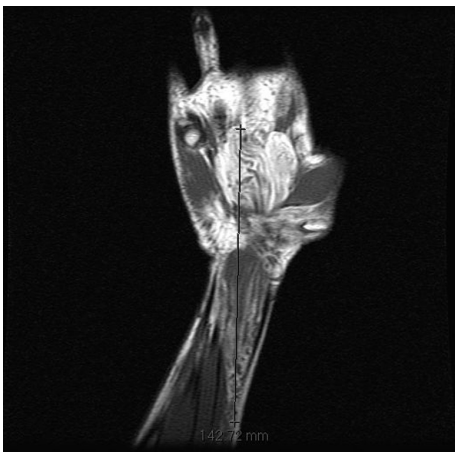

Figure 3 Coronal T2 weighted image shows "spaghetti" like appearance of the lesion. Lesion measures 14.2 cm in craniocaudal dimension.

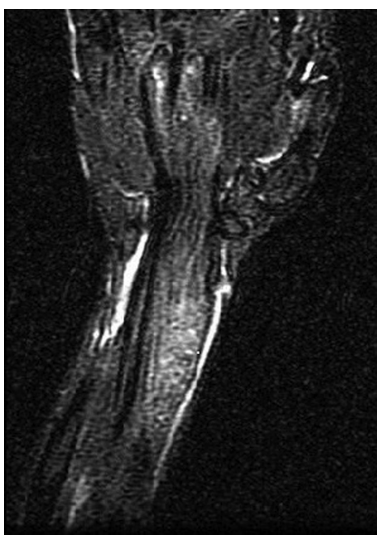

Figure 4 Axial STIR image shows suppression of signal in hyperintense fibroadipose tissue more pronounced in distal part of the lesion suggestive of variable fatty component.

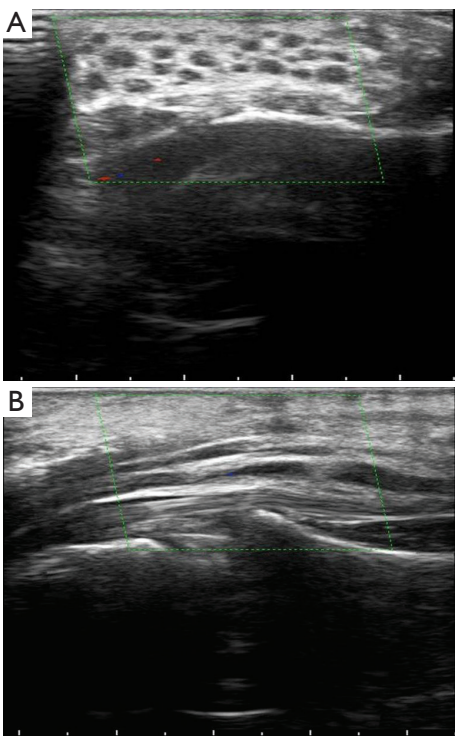

Figure 5 Axial (A) and longitudinal (B) images of ultrasound with color doppler show hypoechoic thickened axonal bundles interspersed in echogenic fatty tissue with no intralesional flow.

Diagnosis of lipofibromatous hamartoma of median nerve was made. Patient was kept on conservative management.

Discussion

Lipofibromatous hamartoma is an uncommon benign lesion that arises due to overgrowth of fibroadipose tissue causing infiltration of endoneurium, perineurium and epineurium resulting in fusiform nerve enlargement. The World Health Organisation tumour classification has described these lesions as lipomatosis of the nerve. The etiology is poorly

understood but they are believed to be of congenital origin. Fewer than 100 cases have been reported previously in the literature (2-4).

The most commonly affected nerve is median nerve followed by ulnar nerve, radial nerve, nerves on dorsum of foot, brachial plexus and cranial nerves. Patients mostly present with soft, slowly enlarging, painless mass on volar aspect of wrist or forearm since infancy. The neural symptoms of pain, paresthesias, weakness, carpal and cubital tunnel syndrome develop gradually due to nerve compression (5,6). It is frequently associated with macrodystrophia lipomatosa in approximately 20 to 66 percent of the cases (7).

The differential diagnosis based on clinical examination includes ganglion cysts, neuromas, lipomas and vascular malformations (2).

Diagnosis is made on the basis of characteristic features on sonography and MRI without the need for biopsy. Sonographic findings show smooth, rounded, thickened hypoechoic or anechoic fascicles surrounded by echogenic fatty tissue corresponding to histological findings. No intralesional flow is seen on color doppler. MRI shows fusiform enlargement of nerve containing thickened axonal bundles encased in epineural fibrous tissue giving "coaxial cable" like appearance on axial images and "spaghetti" like appearance on coronal images (3). We suggest with our experience that as sonography is faster and easier to perform and cheaper modality as compared to MRI, familiarity with characteristic features on sonography may obviate the need for MRI and biopsy.

Management in most cases is conservative. Surgical treatment is only recommended for symptomatic management (1).

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