FIBROLIPOMATOUS HAMARTOMA OF THE MEDIAN NERVE AN UNCOMMON CONGENITAL DISORDER

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HOW TO CITE THIS ARTICLE: Devineni BC, Satbir Singh. Fibrolipomatous hamartoma of the median nerve an uncommon congenital disorder. J. Evid. Based Med. Healthc. 2018; 5(12), 1103-1105. DOI: 10.18410/jebmh/2018/228

PRESENTATION OF CASE

We present a case of fibrolipomatous hamartoma of the median nerve. The patient is a 17-year-old female with painless swelling of the mid-palm for 1 month. The involved sites included the left palm, wrist and forearm. Magnetic resonance imaging demonstrated a typical fibrolipomatous hamartoma with high signal intensity of fat on both T1-weighted and T2-weighted images, characteristic coaxial cable appearance on axial images, and spaghetti appearance on sagittal images.

CLINICAL DIAGNOSIS

Swelling or mass on the volar aspect of the wrist or distal forearm.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis includes fibrolipomatous hamartoma, intraneural lipoma, ganglion cyst, traumatic neuroma and vascular malformations.

Imaging Features

Macrodactyly of the involved part can be present in ~ 66% of all cases of fibrolipomatous hamartoma of the median nerve.

Ultrasound

A characteristic hypoechoic coaxial cabling encased by an echogenic substratum may be seen.

MRI

MRI features are often pathognomonic and typically shows a coaxial cable-like appearance on axial images and a spaghetti-like appearance on coronal images.

- T1 - the neural bundles were hypointense to muscle and the surrounding substratum was isointense to muscle.
- T2 - fat components are high signal and fibrous components are low signal.

Financial or Other, Competing Interest: None.
Submission 27-02-2018, Peer Review 03-03-2018,
Acceptance 18-03-2018, Published 19-03-2018.
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DOI: 10.18410/jebmh/2018/228

PATHOLOGICAL DISCUSSION

Fibrolipomatous hamartoma of the nerve is a benign tumour-like mass of fibrofatty tissue that infiltrates and encases involved nerves and its branches but does not involve the surrounding tissues. Pathologically, the mass consists of benign fibrous and fatty elements around and in between affected nerves, but with no axonal proliferation. Most cases have been reported as occurring in the median nerve,[1] involving the volar aspects of the hands, wrist, and distal forearms, but uncommon locations involve the ulnar nerve, the radial nerve, the nerves of either side of the toes, cranial nerves, and the brachial plexus.

It occurs most often at birth or infants and less commonly in children and young adults. There is no known cause or hereditary predisposition for this lesion, although hypertrophy of mature fat and fibroblasts in the epineurium has been postulated. The clinical manifestation is a slowly growing mass at the wrist, hand or forearm. Pain and neurological symptoms including carpal tunnel syndrome may be associated lately with lipomatosis of the median nerve. Macrodactyly is seen in 27% - 67% and has been referred to as macrodystrophia lipomatosa.[2,3,4]

Plain radiographs of involved areas often are negative or show non-specific soft tissue swelling.

MRI best characterizes this lesion, which is low on T1WI with high-intensity fat signal around and in between the low-intensity nerve axons. It remains low-to-intermediate on T2-fat-sat-WI, and does not demonstrate enhancement with gadolinium. On axial images the lesion has a very characteristic co-axial cable appearance of low-intensity axons surrounded and interdigitated by fibrofatty signal.[5]

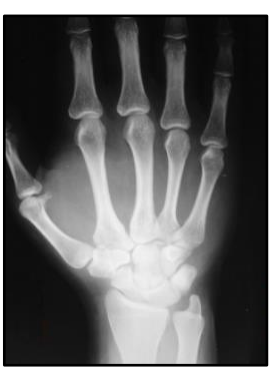

Figure 1

X-Ray Left Hand (AP view) showing a soft tissue mass in the palm.

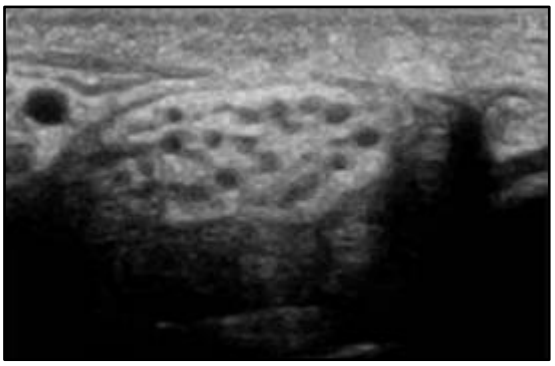

Figure 2

Ultrasound

Transverse image of the median nerve proximal to the carpal tunnel. The nerve is massively thickened with normal and slightly thickened hypoechoic fascicles interspersed with hyperechoic material. A characteristic hypoechoic coaxial cabling encased by an echogenic substratum is seen.

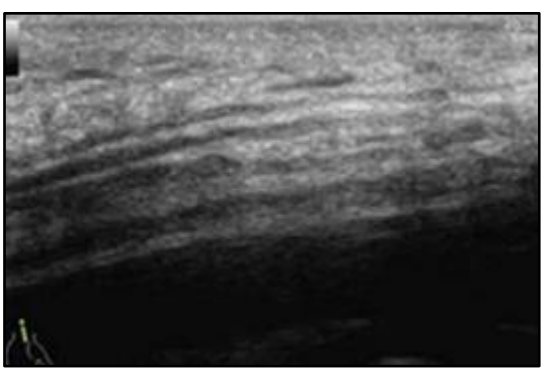

Figure 3

Ultrasound

Longitudinal image of the median nerve proximal to the carpal tunnel showing the massively thickened nerve with normal and slightly thickened hypoechoic fascicles interspersed with hyperechoic material.

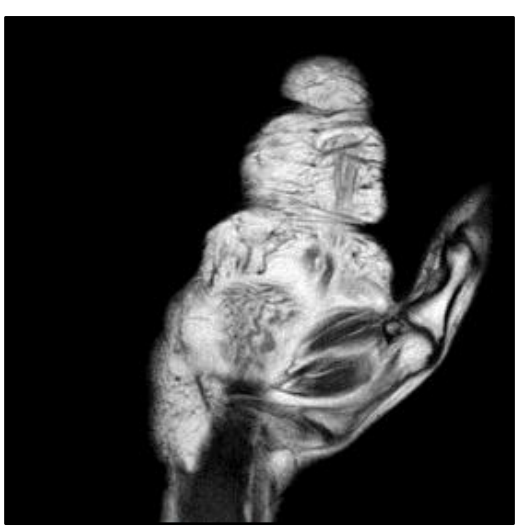

Figure 4

MR

T1 - the neural bundles were hypointense to muscle and the surrounding substratum was isointense to muscle. This coronal image typically shows a 'spaghetti-like' appearance.

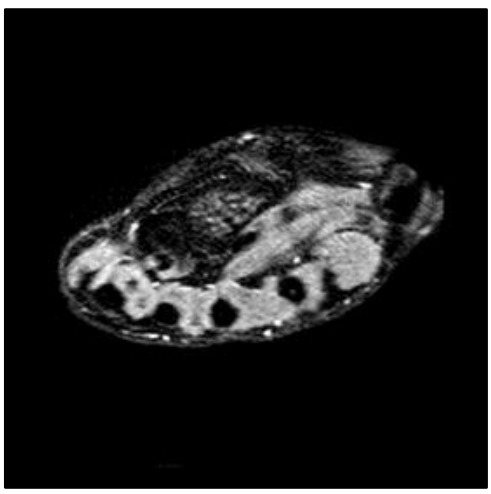

Figure 5

MR

T2 - fat components are high signal and fibrous components are low signal. This axial image typically shows a 'coaxial cable-like' appearance.

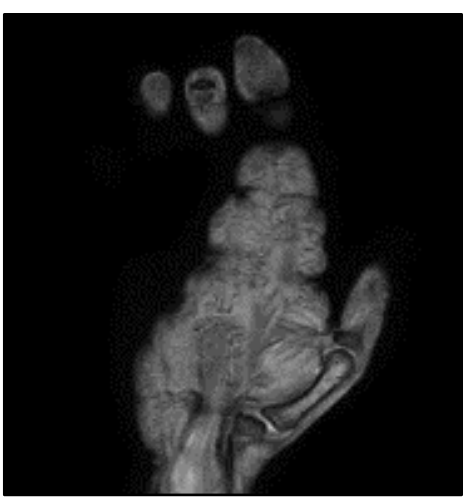

Figure 6

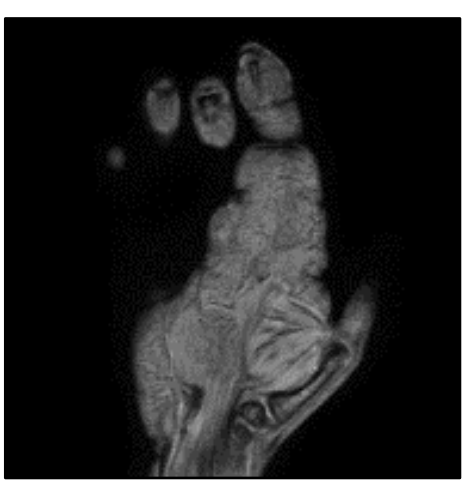

Figure 7

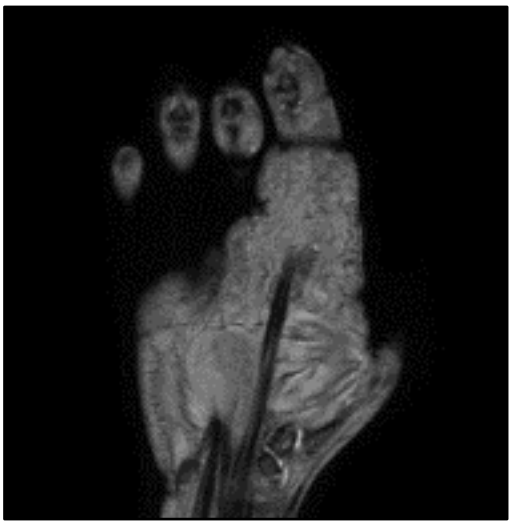

Figure 8

MR
STIR images shows suppression of fatty soft tissue around median nerve fascicles.

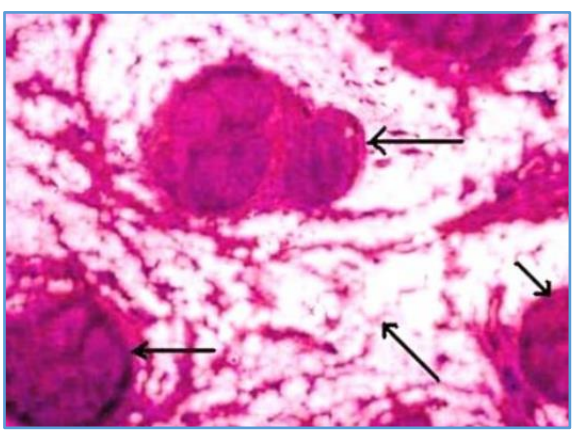

Figure 9

Histology, showing the nerve trunk being surrounded and infiltrated by fibrofatty tissue.

DISCUSSION OF MANAGEMENT

Treatment includes carpal tunnel release. Surgical excision is controversial, motor and sensitive deficits have been reported.[6,7]

Treatment is controversial as excision can sometimes leave the patient with debilitating nerve deficits. Furthermore, an intense healing response after resection has been reported, leaving the patient with worse pain than prior to the surgery. Therefore, the decision to resect is on a case by case basis.

MRI is the gold standard test for the diagnosis of fibrolipomatous hamartoma, with pathognomonic imaging features. The knowledge of radiologic findings may avoid unnecessary biopsies.

FINAL DIAGNOSIS

Fibrolipomatous hamartoma of the median nerve.

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