



Fibrolipomatous Hamartoma of Median Nerve

KEYWORDS

Median nerve, fibrolipomatous hamartoma, magnetic resonance imaging

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ABSTRACT A 24 years old male patient presented with a swelling over thenar aspect of left palm, which was gradually increasing since 4 years with pain on and off and numbness in the middle finger. Radiological imaging findings were suggestive of fibrolipomatous hamartoma of nerve. The patient was operated and the carpal tunnel was decompressed. fibrolipomatous hamartoma is a rare benign tumour that most commonly affects the median nerve.

A 24 years old male patient presented with a swelling over thenar aspect of left palm (Figure 1), which was gradually increasing since 4 years with pain on and off and numbness in the middle finger. There was no history of trauma. On examination mass was palpable in the thenar aspect which was soft and non tender Radiograph of the left hand revealed a soft tissue swelling in the thenar eminence region of the left palm. (Figure 2 a). CT scan revealed fat containing tissue within the lesion (Figure 2 b.) MR finding revealed an 3.4 x 2.5 cm (anteroposterior x transverse) T2 hyperintense well-defined, round to oval lesion extending from distal forearm into the thenar region (Figure 3). Coaxial cable-like appearance was seen on axial images. The lesion consisted of longitudinally oriented cable like structures (Figure 3) that had low signal on T1-weighted images, intermediate to low signal on T2-weighted images with fat saturation. These features represented the thickened neural fascicles. Fatty tissue was interspersed in between the nerve fascicles, which demonstrated high signal in T1-weighted and T2-weighted images appearing hypointense on fat saturation images suggestive of fibrolipomatous hamartoma of nerve. No significant post contrast enhancement was seen. The patient was operated and the carpal tunnel was decompressed. The median nerve was not identified separate from the mass lesion. The nerve was followed in the distal forearm and was seen in continuity. It was infiltrated with fat for approximately 12 cm in longitudinal extent. As the tumor could not be excised without damage to the median nerve no attempt was made to excise it. Biopsy was carried out with decompression of the carpal tunnel. Post operative period was uneventful with patient having normal motor and sensory function with pain less than the preoperative period. Patient was followed on outpatient basis. Histopathological examination revealed islands of fat cells with nerve tissue interspersed in between confirming the diagnosis of fibrolipomatous hamartoma of median nerve.

Discussion

Fat infiltration of the nerve is a rare benign lesion. The World Health Organization tumour classification describes fibrolipomatous hamartoma as lipomatosis of the nerve. The condition has also been designated as fibrolipomatous nerve enlargement, lipofibromatous hamartoma, lipofibroma, fibro-fatty overgrowth, fatty infiltration of nerve, fibro-fatty nerve enlargement, and neurolipoma. Etiology of this lesion by some authors is considered to be a congenital tumour [1] while others believe to be nerve irritation, inflammation, or prior trauma. [2] There is usually a long history of a painless mass since childhood. [3]

More than 80% of fibrolipomatous hamartomas arise exclusively in the median nerve. [1] Other nerves, including the

ulnar, radial, axillary, musculocutaneous, brachial plexus, and cranial nerves, and nerves in the lower extremity can be affected in 4 to 22% of patients.[4] In 27% to 67% of cases, fibrolipomatous hamartoma is associated with macrodystrophica lipomatosa.[4] The reason for the predilection for the median nerve is not certain. However, the median nerve may easily become symptomatic due to encroachment of the flexor retinaculum. Men and women are equally affected and there is no familial predisposition. [4] The upper extremity is involved in 78 to 96% of patients, and there is a marked predilection for the median nerve.[5] Patients with fibrolipomatous hamartoma typically present in the third to fourth decades of life, with signs and symptoms associated with nerve compression in the distribution of the affected nerve.

Histologically, perineural and endoneural fibrosis cause thickening of the neural fascicles. The interfascicular connective tissue is infiltrated by mature fat cells. [5] The perineural and endoneural fibrosis account for the low signal intensity of the thickened nerve fascicles, which appear as cable like structures longitudinally oriented along the nerve. On axial images, the enlarged nerve, with its thickened nerve fascicles interspersed with infiltrating fat, has a cable like appearance and this is pathognomonic of fibrolipomatous hamartoma. In fact, tumour growth following the nerve is a characteristic pathological feature of fibrolipomatous hamartoma (Figure 3).

In this patient, the neural fascicles were evenly interspersed by fat at the level of the carpal tunnel and distal to the carpal tunnel the distribution of fat became eccentric and the amount of fat noted was also more abundant. This can be explained by the fact that the fibro osseous carpal tunnel was restricting the distribution of fat in the nerve. Once outside the confinement of the carpal tunnel, the fatty tissue had more space to proliferate and distribute freely. This is consistent with the imaging findings.

The differential diagnoses of a median nerve mass include intraneural lipoma, ganglion cyst, traumatic neuroma, schwannoma, tenosynovitis, and vascular malformation in which the signal void areas can mimic the low-signal fibrotic neural fascicles. The presence of mature fat within the lesion virtually excludes all other diagnostic considerations except for intraneural lipoma. In the case of intraneural lipoma, the fat content arises from fatty tissue within the epineurium, so this condition will present as a focal mass separate from the neural fascicles, instead of infiltrating in between and separating the neural fascicles. MRI readily differentiates between the two entities. Plain X-rays for patients without macrodystrophica lipomatosa are usually of normal appearance or show only a soft-tissue mass as in this case. When there is macrodys-

trophica lipomatosa, characteristic osseous and soft tissue changes are obvious.

The treatment of fibrolipomatous hamartoma is controversial and depends on the extent of the nerve involvement. Treatment involves carpal tunnel decompression by excising the transverse carpal ligament, followed by biopsy of the enlarged nerve [6] as was done in this case. This procedure results in clinical improvement in 60% of patients. [7] Debulking of the tumour may compromise the vascularity of the nerve and provokes an intense healing response that may jeopardise neurological function. [2] For complete resection, both satisfactory results and catastrophic motor and sensory deficit have been reported.[1]

In conclusion, fibrolipomatous hamartoma is a rare benign tumour that most commonly affects the median nerve. The MRI features, especially the coaxial cable-like appearance on axial images are considered pathognomonic and should enable accurate diagnosis and differentiation from other possible diagnoses. The tendency of tumour growth to follow the nerve and the variation in distribution of mature fat within the lesion are also characteristic pathological and imaging features.

Conflicts of Interest None identified



Figure 1. Swelling over the thenar eminence of the left palm (arrows).

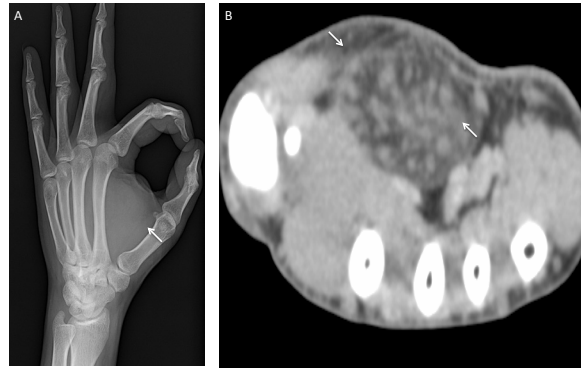


Figure 2. Figure 2a Oblique radiograph the left hand of patient showing soft tissue mass lesion in the thenar aspect of the left hand (arrows) and b CT scan axial image of hand shows a well defined heterogenous lesion in ventral aspect anterior to the flexor tendons having fat density within it with cable like appearance.

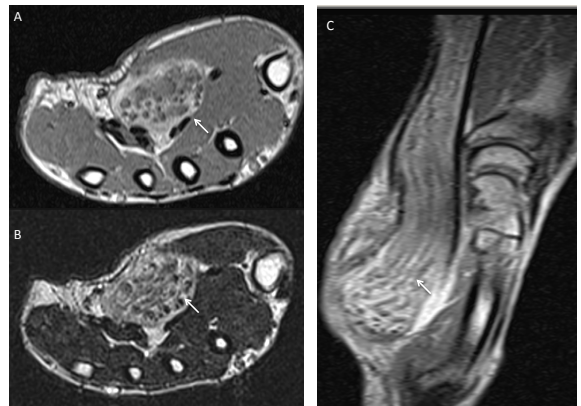


Figure 3. Figure 3a, b and c. MRI T1W and T2W axial images of the left hand and T2 W sagittal image of the left distal forearm and hand shows a well defined fat containing lesion with linear hypointense structure representing the nerve fascicles.

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