J HK Coll Radiol. 2007;10:81-84

CASE REPORT

Fibrolipomatous Hamartoma of the Median Nerve

CK So,[1] KF Tam,[1] CY Lui,[1] CM Lee[2]

[1]Department of Radiology, Kwong Wah Hospital, and [2]Department of Radiology, Princess Margaret Hospital, Hong Kong

ABSTRACT

This report is of a 64-year-old woman with a fibrolipomatous hamartoma of the median nerve who presented with recent enlargement of a right palm mass that had been present for more than 30 years. Magnetic resonance imaging demonstrated pathognomonic features of an enlarged median nerve with thickened individual neural fascicles interspersed with fatty tissue. Magnetic resonance imaging also showed characteristic tumour growth following the branching pattern of the nerve and restriction of tumour fat proliferation at the fibro-osseous carpal tunnel. Debulking of the tumour without complete excision was performed. The pathognomonic magnetic resonance imaging features enable accurate diagnosis and obviate unnecessary biopsy of this tumour.

Key Words: *Hamartoma; Magnetic resonance imaging; Median nerve*

INTRODUCTION

Fibrolipomatous hamartoma is a rare condition that most commonly affects the median nerve. The tumour consists of perineural and endoneural fibrosis causing thickening of the neural fascicles, which are interspersed with proliferative mature fat. The features of fibrolipomatous hamartoma on magnetic resonance imaging (MRI) are pathognomonic and enable accurate diagnosis. Unnecessary biopsy of the tumour may be obviated. Treatment of fibrolipomatous hamartoma is controversial, and depends on the extent of nerve involvement. Catastrophic motor and sensory deficit have been reported following complete resection.[1]

CASE REPORT

A 64-year-old woman presented in 2003 with recent rapid enlargement of a right palm mass associated with right middle finger numbness. The mass had been present for more than 30 years and measured approximately 1 cm in size, but had increased to approximately 7 cm within the previous 2 months. At physical examination, the mass was soft and non-tender. Tinel's sign was positive, with numbness along the right middle finger.

Plain X-rays showed a soft-tissue bulge without calcification between the first and second metacarpal. There were no periosteal reaction or bone remodelling. MRI showed an elongated mass along the segment of median nerve in the carpal tunnel, which had extended to involve the first digital branch of the median nerve. The mass consisted of longitudinally oriented serpentine structures that demonstrated low signal on T1-weighted images, intermediate to low signal on T2-weighted images with fat saturation, and demonstrated enhancement after intravenous contrast injection. These features represented the thickened neural fascicles with endoneural and perineural fibrosis. The fascicles were separated or interspersed by fatty tissue, which demonstrated high signal in T1-weighted images and low signal in T2-weighted images, with fat saturation. Fatty tissue also surrounded the neural fascicles at the periphery of the tumour. On axial images, the tumour had a coaxial cable-like appearance (Figure 1).

At surgery, the median nerve was found to be enlarged and tortuous from the level of the carpal tunnel onwards. The nerve was insinuated with fibrous tissue and fat. The branches to the thenar muscles and the first digital branch of the median nerve were also infiltrated by the

Correspondence: *Dr CK So, Department of Radiology, Kwong Wah Hospital, 25 Waterloo Road, Kowloon, Hong Kong.*
Tel: (852) 3517 5189; Fax: (852) 3517 5454;
E-mail: edwardckso@yahoo.com

Submitted: 29 March 2007; Accepted: 19 October 2007.

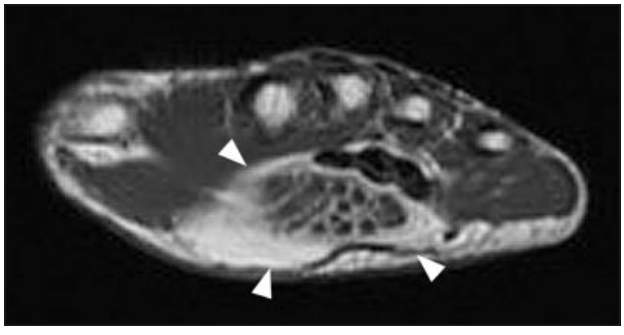

Figure 1. Fibrolipomatous hamartoma of the median nerve. Axial T1-weighted image at the level of the metacarpal base. Note the coaxial cable-like mass (arrowheads) in the position of the median nerve. There were hypointense thickened neural fascicles interspersed by hyperintense fat infiltrate. Mature fat was also found eccentrically in the periphery of the tumour.

tumour, and the branches were thickened. The carpal tunnel was opened and debulking of the tumour without complete excision was performed. Grossly, the tumour appeared pink-yellow in colour. Histological sections of the lesion confirmed the diagnosis of fibrolipomatous hamartoma with mature benign fibro-fatty soft tissue, and a few small branches of the nerve were noted. There was no postoperative neurological deficit.

DISCUSSION

Fibrolipomatous hamartoma is a rare benign lesion. 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. The World Health Organization tumour classification describes fibrolipomatous hamartoma as lipomatosis of the nerve. However, this description does not account for the fibrous element of this tumour.[2] Some researchers consider fibrolipomatous hamartoma to be a congenital tumour,[1] while others believe that it is incited to grow by nerve irritation, inflammation, or prior trauma.[3]

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. There is usually a long history of a painless mass since childhood.[1,4] Men and women are equally affected and there is no familial predisposition.[5] The upper extremity is involved in 78% to 96% of patients, and there is a marked predilection for the median nerve.[5,6] More than 80% of fibrolipomatous hamartomas arise exclusively in the median nerve.[1] Other nerves, including the ulnar nerve, radial nerve, axillary nerve, musculocutneous nerve, brachial plexus, and cranial nerves, and nerves in the lower extremity can be affected in 4% to 22% of patients.[5-7] In 27% to 67% of cases, fibrolipomatous hamartoma is associated with macrodystrophica lipomatosa.[5,6] 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.[3]

Histologically, perineural and endoneural fibrosis cause thickening of the neural fascicles. The interfascicular connective tissue is infiltrated by mature fat cells.[1,4] The perineural and endoneural fibrosis account for the low-signal intensity of the thickened nerve fascicles, which appear as serpentine structures longitudinally oriented along the nerve. On axial images, the enlarged nerve, with its thickened nerve fascicles interspersed with infiltrating fat, has a coaxial cable-like appearance and this is pathognomonic of fibrolipomatous hamartoma. Besides the segment of median nerve in the carpal tunnel, the first digital branch and branches to the thenar muscles were also involved in this patient. In fact, tumour growth following the branching pattern of the nerve is a characteristic pathological feature of fibrolipomatous hamartoma (Figure 2).

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 (Figure 3). This is consistent with the imaging findings for the 2 patients with extensive fibrolipomatous hamartoma of the upper extremity nerves reported by Toms et al.[4] Those 2 patients demonstrated evenly separated neural fascicles by fat in the regions of the carpal tunnel and the cubital tunnel, while the fat was distributed eccentrically and erratically in other areas. This variation in the distribution of tumour fat within the same lesion is another pathological and imaging characteristic of fibrolipomatous hamartoma.

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 serpentine low-signal fibrotic neural fascicles. The

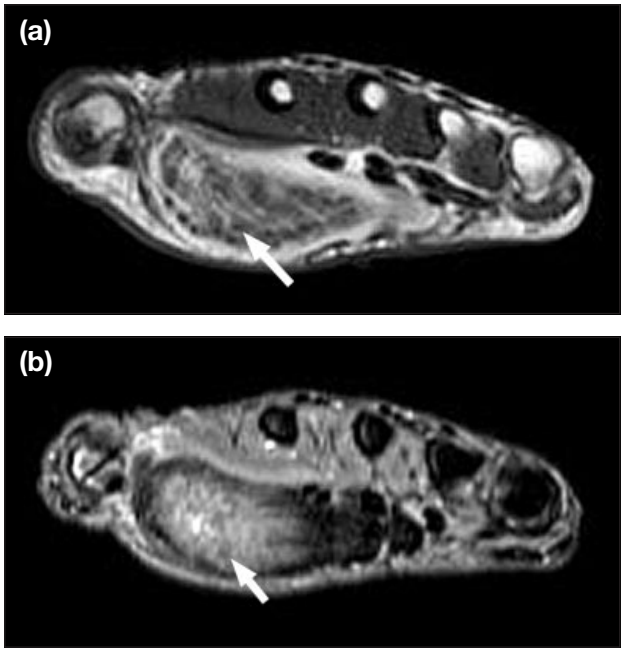

Figure 2. Extension of fibrolipomatous hamartoma of the median nerve laterally into the first digital branch. (a) Axial T1-weighted image at the level of the metacarpal shaft; and (b) axial T2-weighted image, with fat saturation, at the level just distal to (a). Note the signal intensity of the thickened neural fascicles (arrows), which were of low to intermediate signal intensity in both T1-weighted and T2-weighted images with fat saturation.

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 should readily differentiate between the 2 entities.

Sonography of fibrolipomatous hamartoma also shows characteristic hypoechoic coaxial cabling, which corresponds to the thickened fibrotic neural fascicles encased by echogenic fatty substratum.[4] Sonography may be a more convenient and easily available diagnostic tool. Plain X-rays for patients without macrodystrophica lipomatosa are usually of normal appearance or show only a soft-tissue mass. When there is macrodystrophica 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. Conventional treatment involves carpal tunnel decompression by excising the transverse carpal ligament, followed by biopsy of the enlarged nerve.[7] This procedure has resulted in clinical improvement in

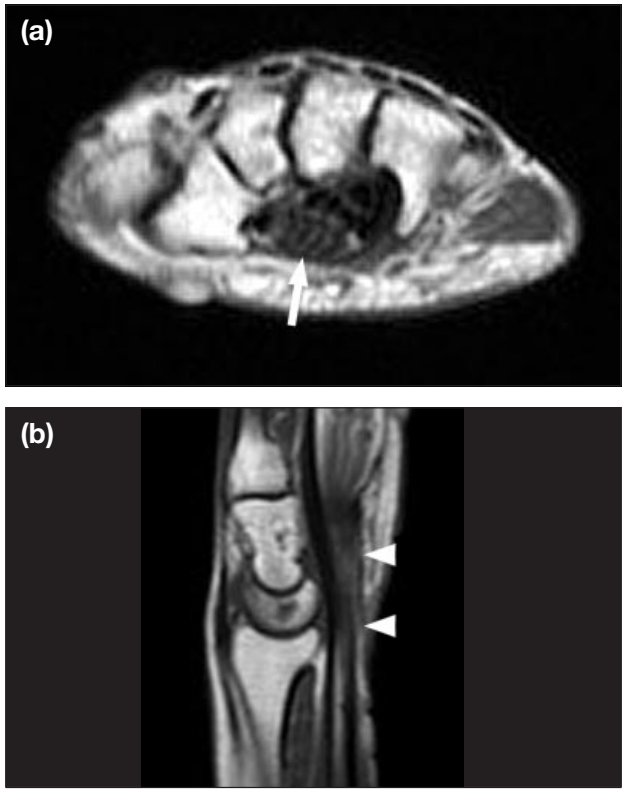

Figure 3. Fibro-osseous carpal tunnel restricting tumour fat proliferation in fibrolipomatous hamartoma. (a) Axial T1-weighted image at the level of the carpal tunnel — note the smaller calibre of the tumour within the carpal tunnel (arrow) compared with Figure 2; and (b) sagittal T1-weighted image — proliferation of tumour fat is restricted at the level of the carpal tunnel (arrowheads).

60% of patients in 1 study.[8] Debulking of the tumour may compromise the vascularity of the nerve and provokes an intense healing response that may jeopardise neurological function.[3] 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. Unnecessary biopsy of the tumour may be obviated. The tendency of tumour growth to follow the branching pattern of the nerve, and the variation in distribution of mature fat within the lesion are also characteristic pathological and imaging features.

REFERENCES

1. Cavallaro MC, Taylor JA, Gorman JD, Haghighi P, Resnick D. Imaging findings in a patient with fibrolipomatous hamartoma of the median nerve. AJR Am J Roentgenol. 1993;161:837-8.
2. Nielsen GP. Lipomatosis of nerve. In: Fletcher DM, Unni KK,

Mertens F, editors. Pathology and genetics of tumours of soft tissue and bone. WHO classification of tumours. Vol 5. Lyon: IARC Press; 2002. p 24-5.
3. Marom EM, Helms CA. Fibrolipomatous hamartoma: pathognomonic on MR imaging. Skeletal Radiol. 1999;28:260-4.
4. Toms AP, Anastakis D, Bleakney RR, Marshall TJ. Lipofibromatous hamartoma of the upper extremity: a review of the radiologic findings for 15 patients. AJR Am J Roentgenol. 2006; 186:805-11.
5. Silverman TA, Enzinger FM. Fibrolipomatous hamartoma of nerve: a clinicopathologic analysis of 26 cases. Am J Surg Pathol. 1985;9:7-14.
6. Amadio PC, Reiman HM, Dobyns JH. Lipofibromatous hamartoma of nerve. J Hand Surg. 1988;13:67-75.
7. Giuliano V, Outwater EK, Mitchell DG, Burke MA. Median nerve hamartoma: MR imaging using chemical shift techniques. Magn Reson Imaging. 1997;15:1091-4.
8. Warhold LG, Urban MA, Bora FW Jr, Brooks JS, Peter SB. Lipofibromatous hamartoma of the median nerve. J Hand Surg. 1993; 18:1032-7.