Carpal tunnel syndrome caused by neural fibrolipoma

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DESCRIPTION

A 45-year-old woman presented with progressive right hand numbness in the area of distribution of the median nerve, and inadequate thumb opposition. On physical examination, a soft elastic mass was noted on the distal volar aspect of her forearm (figure 1, dot circle), with an atrophy of the thenar eminence (figures 1 and 2, red circle). Phalen's test

Figure 1 A subcutaneous tumour on the distal volar aspect of forearm, with atrophy of the thenar eminence.

Figure 2 Axial T2-weighted MRI showing thenar muscle atrophy and cable-like appearance due to hyperintense fat interspersed between the hypointense nerve fascicles.

and Tinel's sign were positive. Motor and sensory nerve conduction velocity of the right median nerve were not evoked. MRI revealed the enlarged median nerve extended into the carpal tunnel

Figure 3 Axial T1-weighted MRI showing enlarged median nerve.

Figure 4 Axial T1-weighted MRI showing enlarged median nerve with coaxial cable-like appearance due to hyperintense fat interspersed between the hypointense nerve fascicles.

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Figure 5 Fat suppression proton density weighted axial image showing low signal lesions with suppression of the predominant adipose component.

(figures 2–4). The characteristic ‘coaxial cable-like’ appearance was observed on T1-weighted and T2-weighted axial images (figures 2 and 4), in which the intermingled reticular high signals were decreased in the fat suppression proton density weighted axial image (figure 5), which indicated the lesion included diffuse fatty infiltration. The ‘spaghetti-like’ appearance in the fat suppression proton density weighted coronal image was shown (figure 6). The carpal tunnel syndrome due to neural fibrolipoma of the right median nerve was diagnosed,

Figure 6 Fat suppression proton density weighted coronal image showing hyperintense linear median nerve fibres in hypointense fat, giving a spaghetti-like appearance.

Figure 7 Operative findings showing a fusiform enlarged median nerve.

then a carpal tunnel release operation was performed. On surgical exploration, a 6×2 cm fusiform enlarged median nerve was observed (figure 7). The patient’s condition had improved at 3-month follow-up, and she now uses the hand well in her job as a cashier.

Neural fibrolipoma, also called fibrolipomatous hamartoma, was first described in 1953 by Mason.¹ Although 22–67% of cases were reported to be associated with macrodactyly, or macrodystrophia lipomatosa if it was present at birth,² the patients had no macrodactyly. The relationship between neural fibrolipoma and macrodystrophia lipomatosa is not clear.²

This disease is a comparatively rare benign tumour that most often involves the median nerve, however, we must keep in mind the characteristic MRI findings.^{2,3} The differential diagnosis of fusiform nerve enlargement includes neural fibrolipoma, lipoma within the nerve sheath and plexiform neurofibroma. In this case, MRI revealed characteristic nerve fascicles surrounded and separated by fibrous and fatty tissue within the expanded nerve sheath as typical features of neural fibrolipoma, which could clearly be distinguished from other disease. MRI may lead to accurate diagnosis and obviate unnecessary biopsy.^{2,3}

Treatment is still controversial. Careful preoperative planning is necessary for the optimal treatment based on the clinical and radiological findings.

Learning points

- ▶ MRI of neural fibrolipoma of the nerve has a characteristic 'coaxial cable-like' appearance on axial image and a 'spaghetti-like' display on coronal image.
- ▶ MRI may obviate the need for biopsy for diagnosis.

Contributors DU, TO and KN evaluated the patient, diagnosed the condition and carried out the treatment. All the authors were involved in collecting data, searching the literature and final approval of the article.

Competing interests None declared.

Patient consent Obtained.

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