

A Case of Fibrolipomatous Hamartoma of the Digital Nerve without Macroductyly

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Abstract

Fibrolipomatous hamartoma of nerves without macroductyly is a rare lesion characterized by fibrofatty proliferation causing epineural and perineural fibrosis with fatty infiltration around the nerve bundles. We report an unusual case of fibromatous hamartoma of the ulnar digital nerve of the thumb in a 43-year-old woman. Magnetic resonance imaging revealed a large fusiform mass along the nerve. The findings were unusual and pathognomonic and included a coaxial cable-like appearance on axial sections and a spaghetti-like appearance on coronal sections on both T1- and T2-weighted images; these findings were useful for the diagnosis and preoperative evaluation of this lesion. Surgical exploration revealed a yellow, cordlike mass of the digital nerve enlarged by fat. Gross excision could not be done without extensive damage to the nerve. Therefore, a limited excision with biopsy of the fibrolipomatous tissue around the nerve bundles was performed. The histological appearance was consistent with fibrolipomatous hamartoma. There was no recurrence of the mass and no neurological deficit 3 years after surgery. Some authors have suggested that invasive excision can cause catastrophic sensory or motor deficits because of the extensive fatty infiltration of the nerve fascicles. In conclusion, the recommended treatment for this lesion is limited excision with only biopsy to confirm the diagnosis.

(J Nippon Med Sch 2011; 78: 388–392)

Key words: fibrolipomatous hamartoma, digital nerve, magnetic resonance imaging, treatment, surgery

Introduction

Fibrolipomatous hamartoma of nerves is a rare tumorlike lesion composed of slowly growing fibrofatty tissue surrounding a nerve and infiltrating

the epineurium and perineurium¹, first reported in the English-language literature in 1953². Such lesions have also been reported as lipofibromatous hamartoma^{3,4}, lipofibroma⁵, fibrofatty proliferation of nerve⁶, intraneural lipoma⁷, neurofibroma⁸, and fatty infiltration of nerve⁹. Fibrolipomatous hamartoma

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can occur in isolation, but approximately two-thirds of cases are associated with macrodactyly^{1,3}. In macrodactyly, there is overgrowth of all tissue with digital gigantism, and fibrofatty tissue develops between the nerve fascicles. More than 80% of fibrolipomatous hamartomas arise exclusively in the median nerve with carpal tunnel syndrome^{1,3}. However, extremely rarely, fibrolipomatous hamartoma can occur in a digital nerve alone^{3-7,9}, as in the present case. The current report describes a case of fibrolipomatous hamartoma of the ulnar digital nerve of the thumb.

Case Report

A 43-year-old woman complained of a soft mass of the left thumb. She had had the asymptomatic mass since childhood and had noticed that it had enlarged slowly for several years before the present examination. Past medical and family histories were unremarkable. No evidence of macrodactyly was noted. The patient had sought medical attention mainly because the thumb mass interfered with the normal key pinch. Physical examination revealed a soft, smooth, nontender 5 × 2 × 1-cm mass on the ulnar side of the thumb and extending to the palm (**Fig. 1**). The mass adhered both to the surface skin and to the deep tissue. Sensation and movement of the thumb were normal. The Tinel sign was absent.

Radiological examination showed that the mass pressed on the palmar-ulnar side of the proximal phalanx of the thumb. Magnetic resonance imaging (MRI) demonstrated a large fusiform mass along the ulnar digital nerve of the thumb. Both T1- and T2-weighted coronal images showed fusiform nerve enlargement with thickened nerve bundles



Fig. 1 First physical examination. Physical examination revealed a soft, smooth, nontender, 5 × 2 × 1-cm mass on the ulnar side of the thumb and extending to the palm.

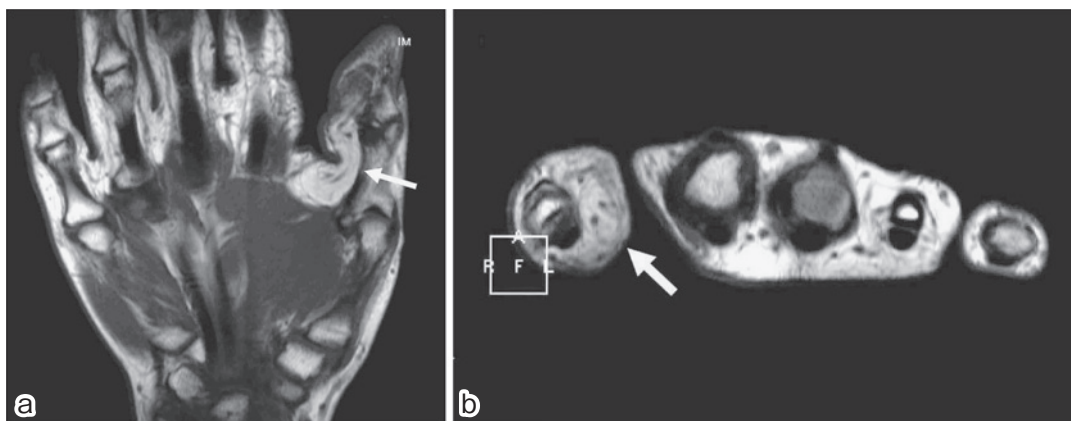


Fig. 2 A T1-weighted MR image of the hand.

- a:** Coronal T1-weighted image of the hand shows a spaghettilike appearance of the digital nerve with hyperintense fat between and surrounding hypointense nerve fascicles (**white arrow**).
- b:** Axial T1-weighted image of the hand shows a coaxial cable-like appearance of the digital nerve with hyperintense fat between and surrounding hypointense nerve fascicles (**white arrow**).

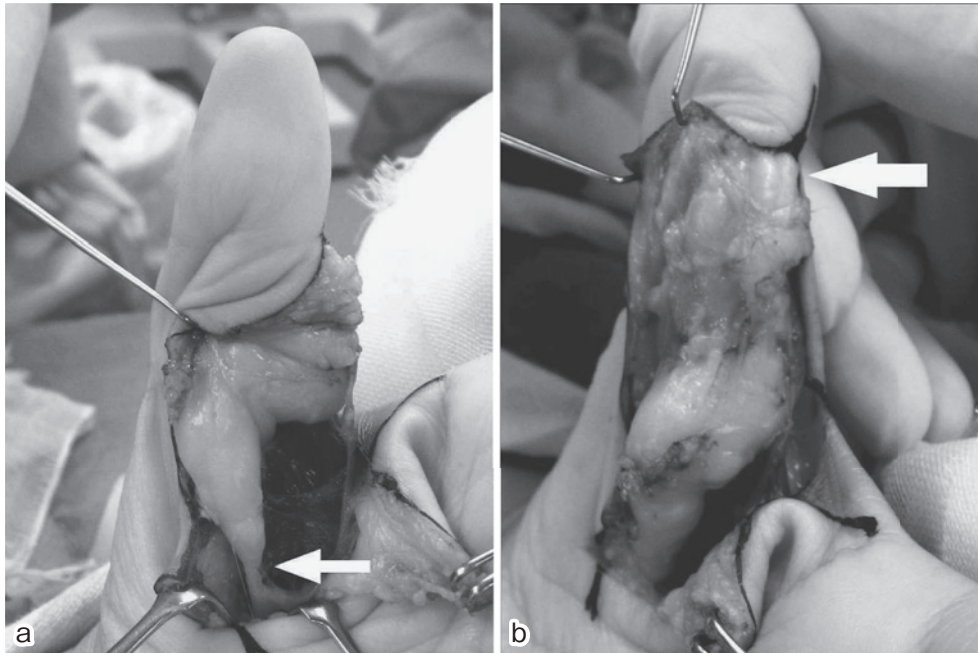


Fig. 3 Intraoperative photograph showing the ulnar digital nerve enlarged by fibrolipomatous tissue in the thumb.

Surgical exposure reveals a soft, yellow, sausage-shaped, fusiform enlargement of the digital nerve by anomalous growth of fibrofatty tissue.

a: White fine arrow: the digital nerve of the thumb.

b: White wide arrow: the digital nerve after limited excision and biopsy of the fibrolipomatous tissue.

appearing as serpentine, low-intensity, tubular structures embedded in the hyperintense fat (**Fig. 2 a**). On both T1- and T2-weighted images in the axial plane, the enlarged nerve appeared as a round, coaxial cable-like structure with hypointense foci representing nerve bundles embedded in hyperplastic fat (**Fig. 2b**). Preoperative needle aspiration biopsy suggested a soft tissue tumor, such as a lipoma.

Surgical exploration revealed a yellow, well-encapsulated, cordlike mass, which proved to be the hypertrophic ulnar digital nerve (**Fig. 3a**). The mass extended from the level of the proximal thumb crease to the tip of the thumb. The digital nerve had a diameter of up to 1 cm at its tip (**Fig. 3b**).

The epineurium of the thickened nerve was incised, because gross excision could not be performed without extensive damage to the nerve. Therefore, a limited excision with biopsy of the fibrolipomatous tissue around the nerve bundles was performed, and the thumb was debulked.

Histological examination of the biopsy specimens

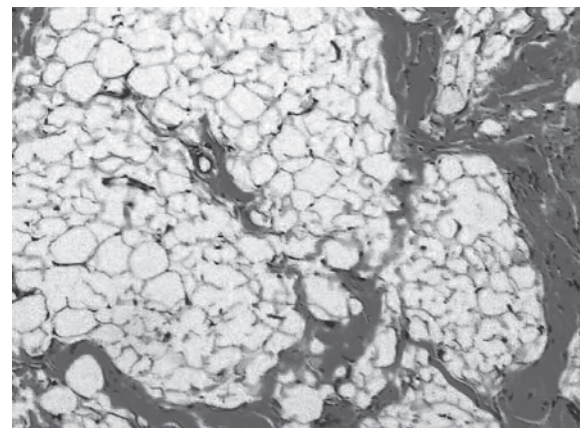


Fig. 4 Histological appearance of the biopsy around nerve fascicles (hematoxylin and eosin stain). The section shows thick fibrous septi of proliferating collagen fibers among the densely accumulating fat cells.

confirmed the presence of mature fat cells interspersed with fibrofatty connective tissue (**Fig. 4**). The nerve enlargement was due to infiltration by fibrofatty tissue with epineural and perineural fibrosis. The histological appearance was consistent

with fibrolipomatous hamartoma.

One year after surgery, the hyperesthesia of the ulnar side of the left thumb which had developed after surgery had resolved. At the 3-year follow-up evaluation, there was no recurrence of the mass and no neurological deficit.

Discussion

Fibrolipomatous hamartoma of nerves is a rare lesion characterized by fibrofatty proliferation causing epineural and perineural fibrosis with fatty infiltration around the nerve bundles; therefore, the abnormality is appropriately described as a hamartoma¹. A variety of descriptions have been applied to this lesion and include lipofibromatous hamartoma^{3,4}, lipofibroma⁵, fibro-fatty proliferation of nerve⁶, intraneural lipoma⁷, neurofibroma⁸, fatty infiltration of nerve⁹, fibrolipoma of nerve¹⁰, and nerve lipoma¹¹. Fibrolipomatous hamartoma of the nerves may occur as an isolated lesion, but two-thirds are associated with macrodactyly, which is a congenital progressive gigantism with increased fibromatous tissue^{1,3}. In macrodactyly, the nerves, phalanges, tendons, vessels, subcutaneous fat, fingernails, and skin are all enlarged^{1,3}. Tsuge and Ikuta have suggested that the similarity of histopathological features between fibrolipomatous hamartoma and macrodactyly indicates that the 2 conditions are a single entity¹². To our knowledge, 122 cases of fibrolipomatous hamartoma of the nerves without macrodactyly have been reported^{1-11,13-15}. In 90 cases (73.7%) it most often affected the median nerve and was associated with carpal tunnel syndrome^{1,3,5-11}. The involvement of other peripheral nerves, such as the radial, ulnar, superficial peroneal, plantar, and sciatic nerves, has been described^{3,13-15}. However, of the reported cases, only 5 had originated in a branch of a digital nerve^{3-5,7,9}.

The MRI appearance of fibrolipomatous hamartoma of the nerve is unique and characteristic¹³⁻¹⁵. Both T1- and T2-weighted MRIs typically show fusiform nerve enlargement with hypointense, thickened nerve fascicles interspersed among hyperintense areas of fatty infiltration¹³⁻¹⁵.

That MRI features are pathognomonic, demonstrating a coaxial cable-like appearance on axial sections and a spaghetti-like appearance on coronal sections¹³⁻¹⁵. The MRI findings can help differentiate fibromatous hamartoma from other nerve sheath tumors, such as traumatic neuroma, lipoma, and neurofibroma, thus allowing a preoperative diagnosis. The MR findings in the present case were similar to those reported previously. However, the most reported MR features is concerned with the lesions originating in the distribution of the median nerve in the wrist. On MRI of the digits, accurate images of thin nerve fascicles in the coronal or axial plane cannot always be obtained. Consequently, it may be difficult to evaluate the typical appearance on MRI and to diagnose fibromatous hamartoma. Therefore, even if a nerve tumor has a characteristic appearance on MRI, a nerve tumor might not be suspected by the clinician, and diagnostic biopsy is still required.

Treatment of this lesion has been controversial¹. Of 5 reported cases affecting a digital nerve, 2 cases were treated with partial tumorectomy^{4,5}, 1 case with partial resection of the affected nerve³, 1 case with total resection of the affected nerve⁷, and 1 case with biopsy⁹. Follow-up of the 4 cases for which biopsy was not done revealed significant sensory loss postoperatively. Surgical resection of the nerve usually results in devastating nerve deficits¹. Catastrophic motor and sensory deficits following nerve excision have been reported¹. In the present case, a limited excision with biopsy of the fibrolipomatous tissue around the nerve bundles was performed. Although the hyperesthesia of the area of the affected digital nerve occurred immediately after surgery, the sensory change of the thumb had resolved by 1 year after surgery. Because of the extensive fatty infiltration of the nerve fascicles, invasive excision may cause catastrophic sensory or motor deficits. Therefore, the recommended treatment for fibrolipomatous hamartoma of the digital nerve is limited excision with only biopsy of the fibrolipomatous tissue around the nerve bundle without interrupting nerve continuity, to confirm the diagnosis, when the tumor exists only as a localized mass.

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(Received, June 7, 2011)

(Accepted, June 22, 2011)