

Fibrolipomatous Hamartoma of Digital Branch of the Median Nerve without Macrodystrophy: Two Case Reports with Magnetic Resonance Imaging Findings¹

정중신경의 손가락 분지에 발생한 부분거대증을 동반하지 않은
섬유지방종성 과오종: 자기공명영상 소견을 포함하는 2증례¹

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Fibrolipomatous hamartoma (FLH) of the nerve is a rare, benign tumor that most commonly originates from the median nerve of the hand. Fibrofatty tissue proliferates around the nerve and infiltrates the epineurium and perineurium. We present two cases of pathologically proven FLH of a digital branch of the median nerve, without macrodystrophy with magnetic resonance imaging, surgical and pathologic findings. Magnetic resonance images of both cases show well-circumscribed mass with fat signal intensity around an enlarged digital branch of the median nerve and characteristic coaxial-cable-like appearance on axial images and spaghetti-like appearance on coronal images.

Index terms

Fibrolipomatous Hamartoma
Median Nerve
Magnetic Resonance Imaging
Hand

Received August 6, 2012; Accepted October 3, 2012

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INTRODUCTION

Fibrolipomatous hamartoma (FLH) of the nerve is a rare, benign tumor that is characterized by fibrofatty enlargement of the nerve with epineural and perineural fibrosis, and most commonly originates from the median nerve of the hand (1). Patients commonly present with a long history of painless mass and may have associated with macrodactyly of the involved region (1, 2). Characteristic magnetic resonance (MR) imaging findings, which are coaxial cable-like appearance on axial images and spaghetti-like appearance on coronal images have been described in several previous reports (2, 3). We present here two cases of pathologically proven FLH without macrodystrophy of digital branch of the median nerve with MR imaging findings.

CASE REPORT

Case 1

A 39-year-old man was referred to our hospital due to a soft lump in the palm of the second metacarpal joint area of the right hand and a radial aspect of the right second finger. The lump had been present since several months ago. Clinical examination revealed a soft mass and mild tenderness to palpation. He had no symptoms and signs of nerve compression. Radiograph showed soft tissue bulging in radial aspect of the right second finger at the level of proximal phalanx, without bony abnormalities (Fig. 1A). MR imaging of the right hand showed a well-circumscribed mass with fat signal intensity around an enlarged proper digital branch of the median nerve from a

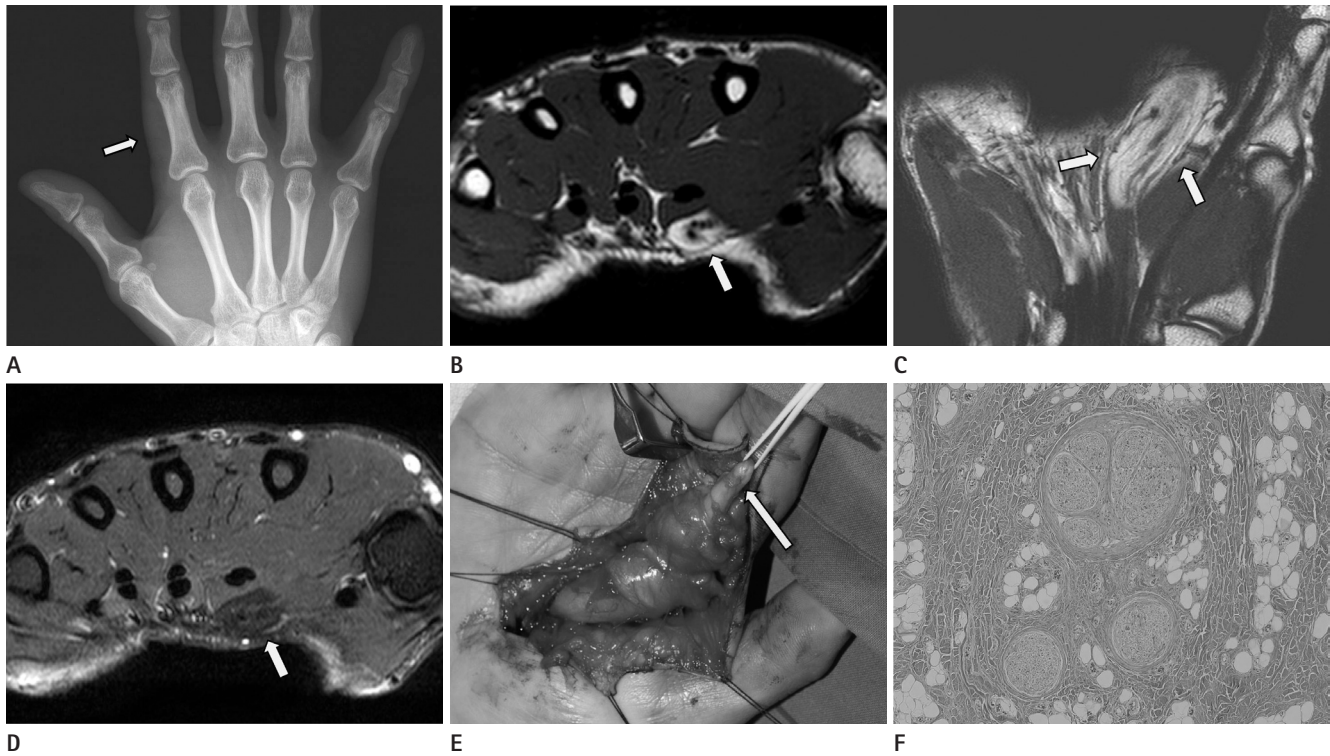


Fig. 1. A 39-year-old man with pathologically proven fibrolipomatous hamartoma involving digital branch of median nerve at right second finger. Radiograph (**A**) shows soft tissue bulging in radial aspect of right second finger (arrow) without bony abnormality. Axial T1-weighted (**B**) and coronal T2-weighted images (**C**) show enlargement of the digital branch of the median nerve and hypointense nerve bundles embedded within fat (arrows). Axial fat-suppressed T1 weighted image with gadolinium enhancement (**D**) shows non-enhancing hypointense fat tissue (arrow) around the digital branch of the median nerve. Intraoperative photograph (**E**) shows fusiform shaped mass and digital nerve fiber (arrow) at the distal ends of the mass. Photomicrograph (**F**) shows fibrofatty tissue growth surrounding the nerve trunk. Septation of nerve fascicles and concentric perineural fibrosis are noted (Hematoxylin & Eosin staining, $\times 100$).

metacarpal shaft to proximal phalangeal shaft level of the second finger (Fig. 1B-D). T1- and T2-weighted images showed that serpentine nerve bundles of low signal intensity were embedded within excessive fatty tissue, which appeared hyperintense. Fat-suppressed T1 weighted images, with gadolinium enhancement, show non-enhancing hypointense fat tissue around the nerve.

He underwent a debulking surgery. Intraoperative findings showed a yellowish mass with about 8 cm in size of the digital nerve. Digital nerve fibers were identified at the proximal and distal ends of the mass (Fig. 1E). The mass was partially removed from the digital nerve, and the nerve fibers were preserved. The histologic examination showed that the mass contained fibrous and lipomatous components, and confirmed the diagnosis of a FLH (Fig. 1F). The patient recovered well and there were no neurologic deficits postoperatively.

Case 2

A 31-year-old man was presented with a mass on the left hand,

at the level of the second metacarpal joint area. He had noticed the mass 1 month ago. Upon a physical examination, a soft mass was located in the volar and radial side of the second metacarpal joint area of the left hand, and there was no abnormal finding in a neurologic examination.

MR imaging of the left hand showed a well-circumscribed, fusiform shaped mass, along the digital branch of the median nerve (Fig. 2A, B). The mass extended from volar aspect of the second metacarpal bone to the radial side of the second proximal phalangeal bone. The low-intensity nerve fibers were interspersed with the high-intensity fatty component, and the nerve fibers within the mass were mildly enlarged. Lipomatous hypertrophy of the soft tissue and overgrowth of the bone were not found in MR images.

He underwent a debulking surgery. Intraoperative findings revealed that the mass was yellowish, fatty in consistency and infiltrated the digital branch of the median nerve (Fig. 2C, D). The mass was pathologically confirmed as FLH (Fig. 2E).

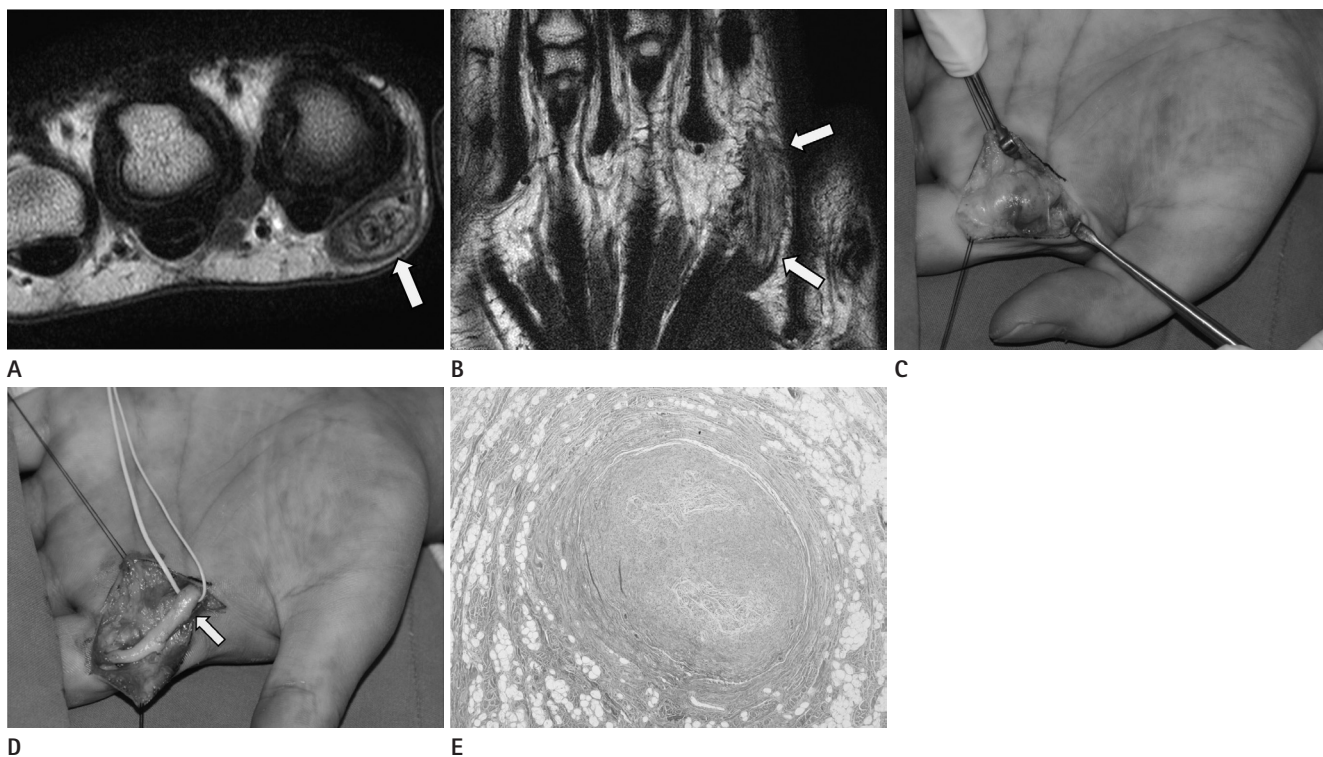


Fig. 2. A 31-year-old man with pathologically proven fibrolipomatous hamartoma involving digital branch of median nerve at left second finger. Axial T2-weighted images **(A)** shows marked enlargement of the digital branch of the median nerve (arrow) surrounded by prominent fat tissue. Coronal T2-weighted images **(B)** shows hypointense nerve bundles embedded within fat (arrows). Intraoperative photograph **(C)** shows a yellowish fusiform shaped mass involving digital branch of median nerve at left second finger. Another intraoperative photograph **(D)** shows that the continuity of the digital nerve (arrow) was preserved after debulking of the fibrolipomatous hamartoma of the nerve. Photomicrograph **(E)** shows adipose and fibrous tissue surrounds and infiltrates the nerve trunk, causing enlargement of the nerve (Hematoxylin & Eosin staining, $\times 40$).

DISCUSSION

Fibrolipomatous hamartoma of the nerve is a rare and benign lesion, which accounts for less than 1% of the benign soft tissue tumor (4). It has also been reported as lipofibromatous hamartoma, neural fibrolipoma, fibrofatty nerve enlargement and neurolipoma (1, 2). The World Health Organization tumor classification describes this tumor as lipomatosis of the nerve; although it does not do justice to the fibrous element within the lesion (5). Terzis et al. (6) classified benign fatty tumors of the peripheral nerves into three types; intraneural lipomas, FLH, and macrodystrophia lipomatosa. Intraneural lipomas are well capsulated benign tumors that arise from adipose cells within the nerve (7). FLH is characterized by diffuse enlargement of the nerve, caused by an overgrowth and fibrofatty tissue proliferation within the epineurium, perineurium, and endoneurium (1, 7). In patients with FLH, 27-66% may have been associated with macrodactyly, which is referred to as macrodystrophia lipomato-

sa (1, 2). Macrodystrophia lipomatosa is characterized by diffuse enlargement of the digits, caused by fatty infiltration and hypertrophy of all components of the digit, including the skin, bone, and nerves (7). However, it still has been unclear of the causal relationship between nerve enlargement and macrodactyly (1, 8). In our two cases, both lesions were fibrofatty enlargement of the nerve, not associated with macrodactyly.

Patients typically presented with longstanding painless mass in the third or fourth decade of life, and often were first noted during childhood (1, 5). Eventually, patients may have paraesthesia, motor deficit, and pain caused by nerve compression in the distribution of the affected nerve (1). Although the etiopathogenesis is unclear, some consider FLH a congenital tumor (1); whereas, others believe it to be a hamartoma incited to grow by nerve irritation or inflammation, infrequently associated with prior trauma (3). FLH most commonly involves the median nerve and its digital branches followed by the ulnar nerve, radial nerve, brachial plexus, nerves of the lower extremity and cranial nerves (1, 5, 9). His-

topathologically, epineural sheath is expanded with various proportions of mature adipose tissue and fibrous tissue, and nerve bundle is surrounded and separated by these fibrofatty tissues (1).

Characteristic MR imaging features of FLH has been described in several reports. MR imaging demonstrates serpiginous low intensity structures on all sequences, which represents thickened nerve fascicles surrounded by or embedded in excessive fatty tissue, which appears as high signal intensity on T1-weighted images and low signal intensity on short T1 inversion recovery and fat-suppressed T2-weighted images (2, 3). These findings resemble a coaxial cable on axial images, and the nerve bundles could be shown a spaghetti-like appearance on coronal images (3). Our two cases showed coaxial-cable-like appearance on axial images and spaghetti-like appearance on coronal images of an MR imaging. Differential diagnosis of FLH from other nerve sheath tumor is generally not difficult. Intraneural lipoma is primary considerable tumor of differentiation with FLH, and is a focal fatty mass, separated from the individual nerve bundles in comparison with the even fatty distribution characteristic on FLH on MR imaging (3). Other considerable tumors, including peripheral nerve sheath tumor (schwannoma and neurofibroma), intraneural hemangioma, and hereditary hypertrophic interstitial neuritis of Dejerine-Sottas disease, show high signal intensity on T2-weighted images, which is usually not shown in FLH (3). Peripheral nerve sheath tumors are frequently characterized by a split-fat sign and a target appearance on T2-weighted images, which are not seen in FLH (10). Intraneural hemangioma has a markedly increased signal intensity on T2-weighted images, whereas FLH have a low intensity on T2-weighted images (3).

The treatment of FLH has not been established and still controversial because the tumor cannot be completely excised, without sacrificing the involved nerves (7). Several reports have shown severe motor and sensory deficits after a surgical excision of the mass, which is currently not recommended (1, 3). However, surgical decompression and debulking are recommended for patients with motor or sensory deficits and restricted digit range of motion (7). In a previous study, Gundes et al. (11) described that microsurgical tumor debulking is associated with good results and preservation of the neural function. Our two patients underwent surgical tumor debulking and achieved good recovery with preservation of the neural function.

We report two cases of FLH with characteristic MR imaging findings, surgical findings and pathologic findings. With this knowledge of characteristic findings, accurate diagnosis could be achieved without unnecessary biopsy or surgical excision.

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정중신경의 손가락 분지에 발생한 부분거대증을 동반하지 않은 섬유지방종성 과오종: 자기공명영상 소견을 포함하는 2증례¹

최우선¹ · 정지영¹ · 송인섭¹ · 이재성² · 이태진³ · 정윤양³

섬유지방종성 과오종은 매우 드문 양성 종양으로 손의 정중신경을 흔히 침범하며, 섬유지방 조직이 신경을 감싸며 신경 바깥막과 신경주위막을 침윤하는 병리소견을 보인다. 우리는 병리진단으로 밝혀진 정중신경의 손가락 분지에 발생한 부분거대증을 동반하지 않은 섬유지방종성 과오종의 두 증례를 경험하여 이에 대한 자기공명영상의 소견과 수술소견 그리고 병리학적 소견을 보고하고자 한다. 두 증례의 자기공명영상에서 비대된 정중신경의 손가락분지를 둘러싸고 있는 지방 신호강도의 경계가 분명한 종괴가 보였으며, 특징적으로 축면영상에서 동축 케이블 소견과 관상영상에서 스파게티 소견이 보였다.

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