

Jawad FARES^{1,2}, Nizar NATOUT³, Youssef FARES¹Fares J, Natout N, Fares Y. Neural fibrolipoma of the median nerve. *J Med Liban* 2016; 64 (1): 47-49.

ABSTRACT • We report a rare case of neural fibrolipoma (lipofibromatous hamartoma) of the median nerve of the hand. An 18-year-old male complaining of progressive and chronic macrodactyly of his thumb of the left hand, with neurological complaints, was admitted to the hospital. Magnetic resonance (MR) evaluation revealed fibro-fatty infiltration of the median nerve at the level of the carpal tunnel, and the terminal branches of the median nerve in the thumb and index. The pathological examination of excised tissue confirmed the diagnosis of neural fibrolipoma of the terminal branch of the median nerve in his thumb. Imaging is recommended; MR studies on morphological abnormalities must be done in similar cases.

Keywords: neural fibrolipoma, fibrolipomatous hamartoma, median nerve, peripheral nerve tumors, macrodactyly of thumb

INTRODUCTION

Neural fibrolipoma, also known as fibrolipomatous hamartoma of the nerve, is a benign and rare nerve sheath tumor that involves the upper and lower extremities; mainly the digits of the hand and foot. It does not become symptomatic until it has been present for many years and its symptomatology stems from compression of the nerve rather than intraneural involvement by the tumor [1].

A fibrolipomatous hamartoma is usually unilateral and has no known genetic component. Although it can present in childhood and early adulthood, it is believed to be of congenital origin [2]. While most commonly found in the median nerve, studies have reported the lesion at other sites such as the radial, ulnar, sciatic, and plantar nerves [3-5] and in the lungs as well [6]. When found in the median nerve, the lesion is usually at the level of the wrist or hand. It is pathologically composed of a fatty and fibrous tissue with intermingling nerve fibers. Magnetic resonance imaging (MRI) demonstrates fusiform nerve enlargement that is caused by fatty proliferation and thickening of nerve bundles. Nerve bundles appear as serpentine tubular structures, hypointense on both T1 and T2 weighted images [7]. Although the World Health Organization (WHO) grouped intraneural lipoma, fibrolipomatous hamartoma of the nerve, fatty infiltration, and neural fibrolipoma under lipomatosis of the nerve in 2002 [8], the clinical findings and treatments of these conditions differ. In early 2013, the new classification of tumors of soft tissue and bones was released and published

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RÉSUMÉ • Nous rapportons un cas rare de fibrolipome neural (hamartome lipofibromateux) du nerf médian au niveau de la main. Un jeune homme de 18 ans se plaignant de macrodactylie chronique et progressive de son pouce gauche, avec plainte neurologique, a été admis à l'hôpital. L'étude avec imagerie par résonance magnétique (IRM) a révélé une infiltration fibro-adipeuse du nerf médian au niveau du canal carpien et des branches terminales du nerf médian dans le pouce et l'index.

L'examen pathologique du tissu excisé a confirmé le diagnostic de fibrolipome neural de la branche terminale du nerf médian dans son pouce. L'imagerie est recommandée; des études IRM d'anomalies morphologiques doivent être faites dans des cas similaires.

under the auspices of the WHO [9]. The revised classification places neural fibrolipomas under benign nerve sheath tumors [9].

Careful preoperative planning based on clinicopathological and radiological findings is necessary for the optimal treatment of neural fibrolipomas of the median nerve [10]. Besides, surgical intervention is only necessary when the patient is symptomatic. This communication presents a case of this uncommon disease with a review of the literature.

CASE REPORT

An 18-year-old male, with right-hand dominance, presented complaining of progressive and chronic macrodactyly of his thumb of the left hand (Figure 1). The patient had noticed the painless swelling for a very long-time, and it was slowly growing in size. Lately, he noted that his left hand was becoming weaker and heavier. Neurological examination showed that the patient was suffering from numbness in his hand mostly felt in his thumb and was incapable of using his hand normally. The remainder of the physical exam was unremarkable. X-ray films were done and revealed no gross abnormality. MRI of the left hand was done as well.

Exploration of the MRI showed an enlargement of the nerve by fibro-fatty tissue (Figure 2). The pathognomonic appearance is that of low signal nerve bundles surrounded by high signal fibrolipomatous tissue on T1 weighted images (Figure 3).

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FIGURE 1. Three images showing macrodactyly of the thumb in the 18-year-old patient.



FIGURE 2. MR images of the wrist and phalanges reveal adipose tissue causing thickened nerve by surrounding fascicles both proximally (median nerve) and distally (inter-digital nerve branches), and overgrowth of the first and second digits with predominance of fat.

An open surgical treatment and biopsy was performed. It showed mature fat cells surrounding normal nerve fasciculi, with no evidence of malignancy. We started off by decompressing the carpal tunnel and debulking the fatty tissue at the levels of the carpal tunnel, thumb and index. Then, microsurgical intraneural dissection was done by performing an epineurotomy of the median nerve. The median nerve's fasciculi appeared enlarged and edematous, lying in a serpiginous manner and encased by the fibrolipomatous infiltrations. The histology later confirmed the diagnosis of fibrolipoma of the nerve with mature fat cells within the nerve epineurium.

The macrodactyly did not disappear immediately post-surgery, and the patient continued to complain from minimal numbness in the thumb but with no swelling or recurrence of the fibrolipoma. In addition, the patient was capable of showing full range motion of both of his thumb and index. His 12-month follow-up showed no clinical evidence of recurrence.

DISCUSSION

We report a rare case of neural fibrolipoma of the median nerve in the hand associated with macrodactyly of the left thumb.

Neural fibrolipoma was initially described in the English literature in 1953 [11]. The cause of this disorder is unknown, although it may be related to hypertrophy of mature fat and fibroblasts in the epineurium.

Patients typically present before 30 years of age, with a soft, slowly enlarging mass in the volar aspect of the hand, wrist, or forearm. Males and females are equally affected, and there is no familial predisposition. The upper extremity is involved in 78%-96% of cases with a marked predilection for the median nerve. Other sites include the lower extremity (4%-22% of cases), ulnar

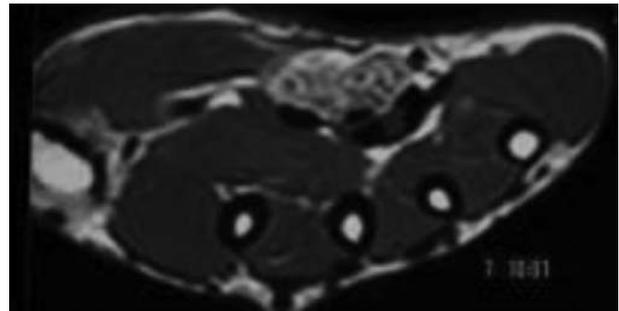


FIGURE 3. Axial T1 image of the hand reveals fatty proliferation and infiltration.

nerve, radial nerve, and brachial plexus [12,13].

Patients present with swelling, with or without accompanying pain, and neurologic symptoms including carpal tunnel syndrome. In 27%-67% of cases, neural fibrolipoma is associated with macrodactyly [14]. The severe mass effect of the fibrolipomatous hamartoma accounts for the neurological symptoms.

At gross examination, the affected nerve is diffusely enlarged. The lesion appears as a tan-yellow mass within the nerve sheath, related to infiltration of the epineurium and perineurium by fibroadipose tissue. The adipose tissue surrounds and separates the usually normal appearing nerve fascicles. The pathologic appearance of the nerve is identical, regardless of the presence or absence of macrodactyly. Patients with macrodystrophia lipomatosa have a diffuse, disproportionate increase in fibroadipose tissue in the affected digit [15].

Radiographs in patients with neural fibrolipoma without macrodactyly often appear normal or may show a focal soft-tissue mass. In patients with macrodystrophia lipomatosa, osseous and soft-tissue overgrowth are seen and often affect both the length and width of the digit. The phalanges are long and broad and often splayed at

their distal ends. The osseous overgrowth, usually more marked volarly and distally resulting in bowing, ceases at puberty but may lead to premature osteoarthritis. Increased radiolucent fat is often apparent in the soft tissues of the affected digit.

Ultrasonography of neural fibrolipoma may show alternating hyperechoic and hypoechoic bands (cable-like appearance).

The gold standard for investigation is the MRI. The MRI appearance is pathognomonic, consisting of longitudinally oriented cylindrical foci (about 3 mm in diameter) of low signal intensity surrounded by fatty signal intensity representing nerve fascicles [13,15,16].

Until recently, the WHO used to classify intraneural lipoma and neural fibrolipoma under the same 'lipomatosis of the nerve' category [8]. Now, the classification places neural fibrolipoma under a new 'nerve sheath tumor' category [9]. Intraneural lipoma and neural fibrolipoma of the nerve are both rare soft tissue tumors; however, significant differences do exist between them. Intraneural lipomas are usually encapsulated with nerve fibers running on their outer surface; thus, complete excision without damage to the adjacent nerve is possible [17]. Contrastingly, neural fibrolipoma is composed of fibrous tissues, fatty tissues, and normal nerve fibers, making complete excision without nerve damage difficult [10].

In addition to the clinicopathological difference, it would also be very important to note that intraneural lipoma and neural fibrolipoma have crucial differences in MR imaging. On T1 and T2 weighted images, the intraneural lipoma intensities are markedly high. However, on short tau inversion-recovery (STIR) images, intensities are low and equal to that of the normal lipoma. MR imaging of neural fibrolipoma has a characteristic "coaxial cable-like" appearance in cross-section or "spaghetti-like" appearance in longitudinal section [18]. Transverse sections show thin hypointense septa within the fat tissue that separate some nerve bundles [19], what we refer to as the "lotus sign". These septa may represent thickened perineurium, a characteristic pathological finding [19].

The differences in MRI would be useful in differentiating between intraneural lipoma and neural fibrolipoma, because the biopsy data is most of the times not informative.

Management of neural fibrolipoma can be conservative or surgical. Intervention is only necessary when the patient is symptomatic. Documented management range from carpal tunnel decompression to fibrofatty sheath debulking to microsurgical neurolysis and finally nerve excision. Although surgery is controversial in areas where excision is associated with a high morbidity and decompression is not possible, careful microsurgical dissection of the neural elements can achieve good prognosis.

Because MRI is often diagnostic, we recommend against doing a biopsy as it is not needed to establish a diagnosis. We must think of this pathology in front of any macrodactyly and do radiologic workup before any surgery. When MRI findings indicate a neural fibrolipoma, careful preoperative planning of reconstruction

for long-term reactivation of motor or sensory deficits should be performed.

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