Fibrolipomatous hamartoma of the median nerve: A cause of acute bilateral carpal tunnel syndrome in a three-year-old child: A case report and comprehensive literature review

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A three-year-old boy was investigated for inexplicable incessant crying. On examination, his left wrist was mildly swollen (three to four months) and sensitive. Exploration and carpal tunnel decompression of the left wrist with incisional biopsy was performed for the presence of a fusiform swelling intimately associated with the median nerve. Histopathology revealed the presence of enlarged nerve bundles admixed with mature fat cells and diffuse fibroblastic proliferation. Three months later, he underwent urgent contralateral carpal tunnel decompression for a similar presentation. The final diagnosis was bilateral fibrolipomatous hamartoma (FLH) of the median nerves causing acute bilateral compression neuropathy.

FLH of the median nerve is an extremely unusual cause of acute bilateral carpal tunnel syndrome in a young child presenting with 'incessant crying'. A comprehensive review of FLH including epidemiology, etiology, clinical presentation, differential diagnosis, imaging, pathology, treatment and prognosis is discussed.

Key Words: Acute bilateral carpal tunnel syndrome; Acute compression neuropathy; Fibrolipomatous hamartoma; Incessant crying; Median nerve; Pediatric hand lesions

Although carpal tunnel syndrome (CTS) is the most common entrapment neuropathy encountered in an adult surgical practice, it is relatively rare in children. In the majority of adults, CTS is believed to be idiopathic in origin. However, in children, CTS is usually secondary to an underlying pathology, such as lysosomal storage diseases, particularly mucopolysaccharidosis and mucopolipidosis. Other causes of CTS in children include genetic bony dysplasia, malformations of the anatomical structures of the carpal tunnel, or enlargement of the median nerve by intraneural lipoma, ganglionic cysts, traumatic neuromas, vascular malformations or neurofibromas. An unusual cause of childhood CTS is fibrolipomatous hamartoma (FLH), a rare benign lesion characterized histologically by the presence of mature fat cells admixed with fibroblastic proliferation within the distorted, expanded nerve bundles.

We report the first case in the published literature of acute bilateral CTS secondary to bilateral FLH of the median nerves without macrodactyly in a three-year-old boy who presented with incessant crying due to acute bilateral compression neuropathy.

CASE PRESENTATION

A three-year-old boy was brought to the emergency room by his mother due to 'incessant crying' that began earlier that morning. His

**Un hamartome fibrolipomateux du nerf médian :
une cause de syndrome du canal carpien aigu
bilatéral chez un enfant de trois ans – rapport de
cas et analyse bibliographique complète**

Un enfant de trois ans a subi des examens en raison de pleurs incessants inexplicables. À l'examen, son poignet gauche était légèrement enflé (depuis trois ou quatre mois) et sensible. Une exploration, suivie d'une décompression du tunnel carpien du poignet gauche par biopsie incisionnelle ont été effectuées pour établir la présence d'œdème fusiforme intimement associé au nerf médian. L'histopathologie a révélé une hypertrophie du faisceau nerveux mêlée à des cellules adipeuses matures et à une prolifération fibroblastique diffuse. Trois mois plus tard, l'enfant a subi une décompression controlatérale urgente du tunnel du canal carpien dans une présentation similaire. Le diagnostic final était un hamartome fibrolipomateux (HFL) bilatéral des nerfs médians responsable d'une neuropathie de compression bilatérale aiguë.

L'HFL du nerf médian est une cause extrêmement inhabituelle de syndrome du canal carpien aigu bilatéral chez un jeune enfant qui « pleurait constamment ». Une analyse complète de l'HFL est exposée, incluant l'épidémiologie, l'étiologie, la présentation clinique, le diagnostic différentiel, l'imagerie, la pathologie, le traitement et le pronostic.

distress had been progressive since he awoke and, on arrival to the emergency room, he was inconsolable. The mother was unable to identify a precipitating cause. On further questioning, the mother recalled his complaining of discomfort in the right wrist for the past three months. The boy was otherwise healthy, took no medications and his immunizations were up to date.

On physical examination, a 4 cm to 5 cm swelling of the flexor aspect of the right wrist and thenar atrophy of this hand were recognized with associated weakness in the hand (Figures 1A and 1B). Tinel's test was positive. The boy's distress escalated with active and passive movement of the wrist, which impeded the assessment of sensation and range of motion. His physical examination was otherwise unremarkable except for an incomplete syndactyly of the second and third toes.

Suspecting a neurofibroma secondary to neurofibromatosis, dermatology and ophthalmology were consulted. A complete workup for the same showed no stigmata of neurofibromatosis. Plastic surgery was subsequently consulted to perform an examination under anesthetic for exploration of the right wrist. Intraoperatively, a fusiform enlargement of the median nerve causing carpal tunnel compression was identified (Figures 2A to 2C). The surrounding tissue was otherwise normal. A biopsy of the median nerve was taken for diagnostic purposes, and a carpal tunnel release (CTR) was performed to alleviate compression symptoms.

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Figure 1) Volar aspect of the right wrist (A) shows the presence of a smooth ill-defined soft 4 cm to 5cm swelling as shown (B) associated with mild thenar atrophy

Figures 2) A to C Exploration of the right wrist under anesthetic showing the presence of a thickened swollen median nerve that has been isolated on blue vessel loops

Histopathological evaluation of this specimen showed thickened serpiginous distorted nerve bundles surrounded by collections of mature fat cells in the background of diffuse fibroblastic proliferation (Figure 3).

Postoperatively, the child's pain decreased dramatically and he was discharged. Three months later, he returned to the emergency room again with 'persistent crying'. A new 1.5 cm mass on the volar aspect of the left wrist was identified. Pain was treated with morphine and carbamazepine (Tegretol, Novartis, USA) and the child returned to the operating room for an urgent CTR and biopsy of the left wrist. The pathology report showed similar findings to the right wrist. Postoperatively, the patient once again recovered completely (Figure 4).

The final diagnosis in the present case was acute bilateral CTS resulting in acute compression neuropathy secondary to bilateral FLH of the median nerves.

DISCUSSION

FLH is a rare benign neoplasm derived from abnormal growth of fibroadipose tissue of the nerve sheath leading to fusiform enlargement of the affected nerve (1-13). The tumour originates from fatty infiltration of the nerve bundle infiltrating the epineurium and perineurium (12,14). The median nerve is affected in >80% of cases; however, several other nerves have been reported with this pathology including the brachial plexus, cranial, ulnar, radial, sciatic, peroneal, plantar and digital nerves (4). These nerves may be individually affected (isolated median/ulnar nerve), or as multiple nerves (15) with or without a constellation of features such as reported: bilateral median nerve, right ulnar nerve with macrocheiria and late-onset nerve entrapment syndrome in a 46-year-old man (1).

Several synonyms have been used to describe FLHs including lipofibromatous hamartoma, lipofibroma, fibrofatty proliferation of nerve, intraneural lipoma, neurofibroma, fatty infiltration of nerve,

Figure 3) Photomicrograph of hematoxylin and eosin-stained slides at medium power demonstrates the thickened serpentine enlarged nerve fascicles (#) admixed with collections of mature fat cells (^) in the background of diffuse fibroblastic proliferation (+)

Figure 4) Postoperative examination of the hands following bilateral carpal tunnel release demonstrating full recovery

fibrolipoma of nerve and nerve lipoma (5,6,12,16-20). Other synonyms described for this lesion include neurolipomatosis, neural fibrolipoma and perineural lipoma (21). The proper terminology for this lesion has been inconsistent in the literature and it is currently recommended that the most accurate term for this lesion is 'fibrolipoma' to reflect the hamartomatous nature of this lesion (6,22).

Epidemiology

FLH was first described by the orthopedic surgeon Mason in the mid-20th century (23). As discussed by Patil and Nagle (24), the term 'lipofibromatous hamartoma' is attributed to Johnson and Bonfiglio (25). FLH most commonly occurs in the young population, specifically in the first three decades of life (3). They are more common in Caucasians (26). This lesion has been described in infants, children, young adults and elderly patients to present either asymptotically or due to a compressive neuropathy (4). Often a mass has been present for years before the development of symptoms (27). A slight predominance of males has been reported (28). FLHs are three times more common in the upper limb than the lower limb (29). FLH associated with macrodactyly more commonly occur in women and upper extremities compared with men and lower extremities (30). FLH of the median nerve remains a rare tumour in the pediatric (<18 years of age) population. Table 1 lists 14 published cases found in a PubMed/MEDLINE search (1986 to

TABLE 1

PubMed/MEDLINE literature (English) review (1986 to 2012) fibrolipomatous hamartoma of the median nerve in the pediatric (<18 years of age) population

Case	Author (reference), year	Age, years	Sex	Clinical presentation	Laterality	Macroductyly	Treatment
–*	Senger et al (–), 2014	3	Male	Incessant crying	Bilateral	None	CTR + incisional biopsy
1	Okubo et al (17), 2012	17	Male	Numbness, mass	Right	None	CTR + biopsy
2	Okubo et al (17), 2012	15	Male	Numbness, mass	Right	None	CTR + biopsy
3	Ulrich et al (31), 2009	8	Female	Pain, numbness, motor/sensory defect	Right	Index	CTR, epineurolysis with partial excision of fibrofatty tissue
4	Sirinoglo et al (32), 2009	8	Male	Painful mass	Right	None	CTR, excision of mass, neurolysis + tissue diagnosis
5	De Smet (33), 2008	5	Female	Macroductyly with paresthesia	Right	3rd and 4th digits	Stripping of digital nerves
6	Nardella et al (34), 2007	15	Female	Dysesthesia, CTS	Right	None	Observation
7	Bains et al (35), 2006	9	Female	CTS: pain, paresthesia	Right	None	CTR, neurolysis, limited excision
8	Jain et al (30), 2006	14	Male	Painless mass	Left	None	Observation with follow-up
9	Hsu et al (10), 2004	16	Male	Painless mass	Right	None	CTR, epineurectomy, microsurgical intraneural dissection, nerve graft
10	Brodwater et al (36), 2000	3	Male	Macroductyly	Left	Long finger	CTR, neurolysis, excision of excess fatty tissue
11	Camilleri and Milner (22), 1998	3	Male	Painless mass, paresthesia	Left	None	CTR with excision of mass
12	Walker et al (37), 1991	3	Male	Painless mass	Left	None	CTR + incisional biopsy
13	Langa et al (38), 1986	14	Female	Painless mass, numbness	Left	None	CTR, partial debulking
14	Sondergaard and Mikkelsen (28), 1984	4	Male	Painless mass	Left	None	Biopsy

*Present case. CTR Carpal tunnel release; CTS Carpal tunnel syndrome

2012) restricted to the English-language literature (10,17,22,28,30,31–38). Among these publications, no reports of bilateral median nerve involvement were identified. Therefore, the present indexed case is the first report of bilateral involvement of the median nerves by FLH in a child. This case is also unique due to its clinical presentation of ‘incessant crying’ due to acute compressive neuropathy.

Etiology

The cause of this abnormal growth of the nerve remains unknown (39). It is proposed that the histogenesis is due to hypertrophy of mature fat cells and fibroblasts of the epineurium (8). The cause of these transformations, however, remains elusive. Some believe this neoplasm is congenital/developmental in origin, whereas others maintain it is an acquired/reactive process due to factors such as nerve irritation or trauma (1,4,5,10,24,40). Those who support the congenital theory, especially in relation to FLH of the median nerve in the carpal tunnel, propose that this patient population is born with an abnormally developed flexor retinaculum or an abnormally developed transverse carpal ligament (26,28,41). It is possible that the true etiology is a combination of both, with an underlying congenital predisposition coupled with trauma or irritation that incites the formation of this neoplasm. Microtrauma due to repetitive pressure on the nerve from the carpal ligament or from an abnormally developing flexor retinaculum may initiate a series of events that culminates in the progressive development of this tumour (42).

Nerve territory-oriented macroductyly is a common concurrent finding, with 20% to 66% of FLH being associated with such macroductyly (1,30,31). No hereditary or chromosomal abnormality is reported in these cases. The relationship between these two entities remains poorly understood. Some authors hypothesize that they are both derived from an end-organ genetic abnormality in which an unidentified trophic factor is responsible for the development of these two pathologies. Other authors believe the nerve dysfunction of FLH causes the development of the progressive type of macroductyly: macrodystrophia lipomatosa progressiva with overgrowth of fatty tissue in the affected limb, in contrast to the static macroductyly congenita, wherein the enlargement is present at birth and does not grow

disproportionately with growth. The precise cause and relationship between FLH and macroductyly, however, remains elusive (31,33).

Clinical presentation

FLH can present with signs and symptoms of painful nerve compressive neuropathy including sensory and/or motor deficits in the nerve distribution. Pain, tenderness, decreased sensation, paresthesia and/or mass effect are common presenting complaints (3,4,43). Signs and symptoms are usually longstanding, often existing for years before seeking medical attention for a painless mass (2,29,40). FLH most often occurs unilaterally (18). These lesions become symptomatic once their size exceeds the passageways through which it passes (40). Because the median nerve is commonly affected, the mass is most often on the volar aspect of the wrist or distal forearm (26). Many clinical characteristics overlap CTS, with symptoms of numbness, paresthesia and weakness within the median nerve distribution. Although distal median nerve compression is the most common site of FLH, proximal median nerve compression of the forearm due to coexisting osteochondroma has also been reported (44). Cubital tunnel syndrome presents with similar symptoms within the distribution of the ulnar nerve (6).

On physical examination, a soft fusiform mass is often visible and located on palpation. However, there are no physical findings that are pathognomonic for distinguishing FLH from other hand or wrist pathologies.

FLH is associated with macroductyly in the affected nerve territory in one-third of patients, which is termed ‘macrodystrophia lipomatosa’, first described by Feriz in 1925 (45). These patients have not only an increased amount of adipose tissue within the affected nerve, but also an overgrowth of the mesenchymal elements of the bone, surrounding soft tissue and skin of the affected digit. The phalanges are most often affected; however, macroductyly of the metacarpals has also been reported in an older patient with severe disease (26). When associated with macroductyly, these neoplasms are most commonly identified in the upper extremities of females (30). Macroductyly may affect both the radial and ulnar sides of the digit equally or asymmetrically, and is unilateral in 90% of cases (26). FLH has also been reported to

be associated with macrodactyly and port-wine stains (46). In patients presenting with hypertrophy of the hand or macrodactyly, particularly if the patient has signs or symptoms of nerve entrapment, an FLH should be considered in the diagnostic profile (1).

Isolated case reports have documented the association of FLH with Klippel-Trenaunay syndrome, a rare congenital disorder characterized by the triad of cutaneous capillary malformations, varicose veins, and hypertrophy of bone and soft tissue (47). Less commonly associated findings with FLH include ectopic calcification of soft tissues, fatty muscle infiltration, bony exostoses, subcutaneous encapsulated lipomas and vascular tumours (7,48).

Differential diagnosis

When diagnosing FLH, several differential diagnoses have been described including ganglionic cysts, hereditary hypertrophic neuritis of Dejerine-Sottas syndrome, vascular malformations, traumatic neuromas, schwannomas, neurofibromas and intraneural lipomas (4,6,21,34,37).

FLHs can be distinguished from true lipomas because the latter are sharply demarcated, well-encapsulated masses that normally occur on the surface arising in the perineurium or epineurium and contain no neural elements (5,49). Such non-neural sheath lesions, also including hemangiomas and ganglions, are amenable to complete excision if accurately identified (39).

In the presence of lipomatous macrodystrophy, the clinician should be suspicious of an associated hemangioma, lymphangioma, arteriovenous malformation or neurofibromatosis (7). FLH can be distinguished from neurofibromas based on their histological characteristics because NFs are nonencapsulated and intermingle with nerve fascicles (26).

Imaging

Standard x-rays are not sensitive for the detection of FLH. They may be normal or show soft-tissue thickening, which is nondiagnostic (7). They are, however, useful in evaluating patients presenting with macrodactyly for musculoskeletal evaluation (30).

On ultrasound, FLH appears as a hyperechoic mass containing coaxial hypoechoic bands (7). The hypoechoic bands are the neural bundles and the hyperechoic regions are the fatty involvement (28). Colour Doppler demonstrates no vascularization (7).

Magnetic resonance imaging characteristics are pathognomonic for FLH and are the diagnostic key for preoperative evaluation. Findings are, however, insufficient to determine the extent of fascicular involvement (29). The mean size of these neoplasms is typically reported to be 3 mm in diameter (30). They have a high fat-intensity signal on both T1- and T2-weighted images with saturation on fat suppression images (7). T1-weighted imaging shows a coaxial cable-like image of high-signal adipose tissue interspersed with the low-signal nerve tissue (29).

Coronal plane images demonstrate 'serpiginous structures', or thickened nerve fascicles that are evenly dispersed throughout the nerve sheath, surrounded by fat, which has high intensity on T1-weighted images and low intensity on fat-suppressed T2-weighted images (40,50,51). Distribution of fat between the fascicles is asymmetric (26). These images, with the appearance of 'spaghetti', are pathognomonic for this neoplasm (4,12,15,16,52).

Axial cuts show a coaxial cable appearance from uniformly distributed nerve bundles within the nerve sheath that are separated by mature fat within the interfascicular connective tissue (4). Transverse sections may show the 'lotus sign' that occurs due to the thin hypointense septa within the fat tissue that separates the fat bundles. This is consistent with the characteristic pathological findings seen in FLH of the thickened perineurium due to perineural fibrosis with septation of nerve fascicles (2,17). Therefore, visualization of the coronal and axial characteristics obviates the need for a biopsy to make the diagnosis of FLH (10,30,42).

Electromyography and nerve conduction studies may additionally aid in the diagnosis and confirm compressive neuropathy (26).

Pathology

On gross examination, these fusiform tumours appear as an irregular palpable yellow/tan-coloured mass within the peripheral nerves due to proliferation of mature adipocytes and associated connective tissue overgrowth (2,6,15,42). Involved nerves may be elongated and enlarged (up to 17 cm and 30 mm, respectively). Along their course, affected peripheral nerves do not adhere to the surrounding tissue (26).

Histopathological examination demonstrates a fibrofatty infiltration of the epineurium causing compression of the nerve fascicle and subsequent delayed neural atrophy. On further examination, the overgrown epineurium, perineurium and endoneurium are disorganized; however, the surrounding tissue demonstrates no involvement or inflammation. Epineural and perineural fibrosis with collagen deposition is usually observed. The affected nerve may also show features including perineural septation of the nerve fascicles and formation of microfascicles (1,2,26,53). Unlike lipomas, FLH demonstrates an infiltrative pattern with an asymmetric distribution of fat between fascicles (26,38). On immunohistochemical examination, FLH stains positive for CD34, S100 and vimentin, and is negative for epithelial membrane antigen, desmin and glial fibrillary acidic protein antibodies (54).

Treatment

In the context of a benign tumour, treatment of FLH is difficult to determine because the lesion cannot be truly completely excised without sacrificing the involved nerve because the proliferating fibrofatty tissue is intermingled with the nerve fascicles, thus necessitating an aggressive procedure that is associated with a risk for significant neurological deficit. Additionally, the rarity of this lesion precludes randomized controlled studies for identifying optimal treatment with best practice protocols. Published reports range from recommendations of observation with no action or a simple biopsy, to microsurgical intraneural neurolysis and/or CTR. Treatment options are, thus, determined on a case-by-case basis, with invasive surgical procedures being generally reserved for patients with symptomatic neurological deficits.

Conservative treatment is the primary gold standard whenever possible (29,31). This approach is three-pronged and includes observation for asymptomatic patients, decompression with or without biopsy, and debulking of the fibrofatty sheath with or without biopsy (17). Some authors argue that young adults with FLH should be managed by observation alone because these lesions may be self-limiting. Others have reported a natural decrease in the size of the tumour over time if left untreated (54). Because there is no known risk for developing malignancy, any treatment undertaken is for comfort or cosmetic reasons and, therefore, no interventional treatment is required for asymptomatic or minimally uncomfortable lesions.

In patients presenting with compressive neuropathy of the median nerve, CTR with a cutaneous nerve biopsy is often the recommended treatment of choice (4,55). This can be performed as an open procedure or by microsurgical or endoscopic techniques with or without epineurotomy or intraneural neurolysis (30). Decompression alleviates symptoms of pain and paresthesia and reduces the risk of permanent motor and sensory sequelae (26). It has also been reported that CTR with release of the fascia for FLH of the median nerve may not only alleviate the pain but also decrease the size of the mass and demonstrate improvement in the strength of opponens pollicis (9,29). Prophylactic decompression may also be considered as an option for patients with large asymptomatic FLH (26). Decompression and debulking of the fibrofatty sheath is associated with more positive results (42). Because FLH presents as a deep-seated mass, surgical exploration with an open incisional biopsy is often the recommended procedure of choice for confirmation of the diagnosis, especially in the absence of corroborative imaging characteristics.

Radical surgery involving interfascicular microdissection is a more aggressive option and is rarely indicated except as an option for patients with progressive, persistent and disabling nerve compromise despite previous CTR (29). Using microsurgical techniques, the bulk

of the tumour may be resected without damage to the nerve if the tumor is small. Interfascicular dissection may additionally cause a disruption in the segmental vascular supply, which can cause ischemia and its resulting complications (26,29). A large tumour increases the risk of neurological deficit due to vascular compromise and the fibrotic healing response that occurs will further interrupt nerve function (19,22,24,27,29,32). The specific indications for this technique, however, remain controversial, with some authors further reporting that such attempts may downgrade nerve function, thereby further compromising outcome (27).

Due to the relatively high risk of nerve damage, tumour excision is generally reserved for cases presenting with progressive and disabling nerve compromise despite CTR (6). Other authors maintain vehemently that complete excision of this fibrofatty growth is contraindicated because it may cause intolerable sensory or motor disturbances and, therefore, should never be offered as a treatment option for FLH (18,26,43). However, others have suggested that it may be indicated in children younger than two years of age due to their ability to re-educate sensibility (24). This surgery is at a high risk for complications because the intraneural elements are intimately associated with the nerve tissue. The majority of authors do not support this treatment modality due to this associated high morbidity. Additionally, in cases in which the FLH extends and involves the brachial plexus, identification of resection margins is often difficult, if not impossible (26). The postoperative fibrous healing response of this aggressive intervention may jeopardize the long-term function of the nerve because fibrosis associated with healing can interfere with nerve transmission (30). In this context, permanent devastating loss of motor and/or sensory innervation is a well-accepted long-term complication of this procedure (29). While motor disruptions can be treated with opponens-plasty, sensory loss has no satisfactory solution (26,27). These patients are also at risk for postoperative neurogenic pain (26). Thus, surgical resection of FLH with or without nerve grafting is not routinely recommended (24).

Several treatment options are available for patients presenting with macrodactyly associated with FLH. The main principle guiding such treatments is cosmetic improvement with preservation of neurological function. These include various options such as staged multiple debulking procedures, epiphysiodesis and osteotomies (36). In the presence of macrodactyly, excision of the affected nerve segment with or without nerve grafting has also been reported (17). In selected cases, tendon transfers and excision of osseous overgrowth have been described (26). Amputation is another option available for this population, particularly in older patients (23).

Prognosis

Published reports regarding the outcome of surgical treatment for FLH of the median nerve has been mixed, with some authors reporting positive outcomes with full motor and sensory function, whereas others have a high complication rate with severe postexcision loss of neurological function (4). Although very little is known about the predictive prognostic factors of the success of a single surgical intervention for these lesions, in the majority of cases of FLH of the median nerve, surgery is limited to simple decompression with an incisional biopsy for diagnostic purposes.

CONCLUSION

Due to the rarity of FLH, the precise epidemiology, etiology and best management strategies continue to be poorly understood. The index case is a unique presentation of FLH of the median nerve causing 'incessant crying' secondary to acute compressive neuropathic pain presenting bilaterally that, to the best of our knowledge, has not been previously reported. In this context, a high index of clinical suspicion for FLH is imperative in patients presenting not only with painless masses as often described, but additionally with acute compression neuropathy. Pathognomonic magnetic resonance imaging findings are confirmatory of the diagnosis and, once established, a conservative

approach to management should be chosen whenever possible because a poorly planned surgical treatment is a significantly greater hazard than the natural benign indolent course of this lesion. Nevertheless, optimal treatment is guided by the severity of symptoms on a case-by-case basis. In this context, therefore, increased clinical awareness of this condition by clinicians, radiologists and pathologists is vital for best practice management.

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ETHICS APPROVAL: Because all relevant patient material in this case report was completely de-identified, it is considered exempt from formal ethics approval from the institutional review board.

REFERENCES

1. Meyer BU, Roricht S, Schmitt R. Bilateral fibrolipomatous hamartoma of the median nerve with macrocheiria and late-onset nerve entrapment syndrome. *Muscle Nerve* 1998;21:656-8.
2. Sheela Devi CS, Suchitha S, Sunila R, Manjunath GV. Lipofibromatous hamartoma of median nerve. *Indian J Pathol Microbiol* 2010;53:177-8.
3. Kim YJ, Kim SY, Kang SJ, Kim GM. Neural fibrolipoma. *J Am Acad Dermatol* 2005;53:528-9.
4. Hankins CL. Carpal tunnel syndrome caused by a fibrolipomatous hamartoma of the median nerve treated by endoscopic release of the carpal tunnel. *J Plast Hand Surg* 2012;46:124-7.
5. Guthikonda M, Rengachary SS, Balko MG, van Loveren H. Lipomatous hamartoma of the median nerve: Case report with magnetic resonance image correlation. *Neurosurgery* 1994;35:127-31.
6. Louaste J, Zejjari H, Chkoura M, Houmadi A, Rachid K. Carpal tunnel syndrome due to fibrolipomatous hamartoma of the median nerve. *Hand (NY)* 2011;6:76-9.
7. Nouria K, Belhiba H, Baccar S, et al. Fibrolipoma of the median nerve. *Joint Bone Spine* 2007;74:98-9.
8. Silverman TA, Enzinger FM. Fibrolipomatous hamartoma of nerve. *Am J Surg Pathol* 1985;9:7-14.
9. Warhold LG, Worcester MA, Urban MA, Bora FW, Brooks JS, Peters SB. Lipofibromatous hamartomas of the median nerve. *J Hand Surg* 1993;18A:1032-7.
10. Hsu CC, Chen DJ, Chen HC. Lipofibromatous hamartoma of the median nerve with long-term follow-up. *Chang Gung Med J* 2005;28:111-6.
11. Liaw K, Kan JH. Lipofibromatous hamartoma. *Pediatr Radiol* 2008;38:1032.
12. Nanno M, Sawaizumi T, Takai S. A case of fibrolipomatous hamartoma of the digital nerve without macrodactyly. *J Nippon Med Sch* 2011;78:388-92.
13. Kronberger P, Rainer C, Hittmair A, Anderl H. Lipofibromatous hamartoma (neural fibrolipoma) of a flexor nerve of the index finger. *Scan J Plast Reconstr Hand Surg* 1998;32:237-9.
14. Sarawagi R, Anderson GA, Cherian RS. Fibrolipomatous hamartoma of the median nerve presenting with carpal tunnel syndrome. *Neurol India* 2009;57:361-2.
15. Toms AP, Anastakis D, Bleakney RR, Marshall TJ. Lipofibromatous hamartoma of the upper extremity: A review of the radiologic findings for 15 patients. *Am J Roentgenol* 2006;186:806-11.
16. Taddie KL, Fallat LM. Lipofibromatous hamartoma of nerve. *J Foot Ankle Surg* 2007;46:116-9.
17. Okubo T, Saito T, Mitomi H, et al. Intraneural lipomatous tumor of the median nerve: Three case reports with a review of literature. *Int J Surg Case Rep* 2012;3:407-11.
18. Ha JF, Teh BM, Abeyuriya DT, Luo DY. Fibrolipomatous hamartoma of the median nerve in the elbow: A case report. *Ochsner J* 2012;12:152-4.
19. Houpt P, van Leeuwen S, van den Bergen HA. Intraneural lipofibroma of the median nerve. *J Hand Surg Am* 1989;14:706-9.
20. Rusko RA, Larsen RD. Intraneural lipoma of the median nerve – case report and literature review. *J Hand Surg Am* 1981;6:388-91.

21. Kameh DS, Perez-Berenguer JL, Pearl GS. Lipofibromatous hamartoma and related peripheral nerve lesions. *South Med J* 2000;93:800-2.
22. Camilleri IG, Milner RH. Intraneural lipofibroma of the median nerve. *J Hand Surg Br* 1998;23:120-2.
23. Bisceglia M, Vigilante E, Ben-Dor D. Neural lipofibromatous hamartoma. A report of two cases and review of the literature. *Adv Anat Pathol* 2007;14:46-52.
24. Patil VS, Nagle S. Lipofibromatous hamartoma of the median nerve: A case report and review of the literature. *Indian J Plast Surg* 2009;42:122-6.
25. Johnson RJ, Bonfiglio M. Lipofibromatous hamartoma of the median nerve. *J Bone Joint Surg Am* 1969;51:984-90.
26. Razzaghi A, Anastakis DJ. Lipofibromatous hamartoma: Review of early diagnosis and treatment. *J Can Chir* 2005;48:94-9.
27. Yamauchi T, Yoshii T, Sempuku T. Whether to excise a lipofibroma of the median nerve. *Scand J Plast Reconstr Surg Hand Surg* 2009;43:346-9.
28. Sondergaard G, Mikkelsen S. Fibrolipomatous hamartoma of the median nerve. *J Hand Surg Br* 1987;12:224-5.
29. Robinson AJ, Basheer MH, Herbert K. An unusual cause of carpal tunnel syndrome. *J Plast Reconstr Aesthet Surg* 2010;63:e788-91.
30. Jain TP, Srivastava DN, Mittal R, Gamanagatti S. Fibrolipomatous hamartoma of the median nerve. *Australas Radiol* 2007;51:B98-100.
31. Ulrich D, Ulrich F, Schroeder M, Pallua N. Lipofibromatous hamartoma of the median nerve in patients with macrodactyly: Diagnosis and treatment of a rare disease causing carpal tunnel syndrome. *Arch Orthop Trauma Surg* 2009;129:1219-24.
32. Sirinoglu H, Sonmez A, Sav A, Numanoglu A. Lipofibromatous hamartoma of the median nerve. *Ann Plast Surg* 2010;65:174-6.
33. De Smet L. Lipofibromatous hamartoma of the median nerve and macrodactyly. *Genet Couns* 2008;19:341-2.
34. Nardella D, Sohawon S, Carlier A. Lipofibromatous hamartoma of the median nerve. Three case reports. *J Plast Reconstr Aesthet Surg* 2009;62:e314-7.
35. Bains R, Kotwal A, Saeed W. Recurrent carpal tunnel syndrome in a child due to fibrolipomatous hamartoma of the median nerve successfully treated by limited excision and decompression. *Plast Reconstr Aesthet Surg* 2006;59:1394-7.
36. Brodwater BK, Major NM, Goldner RD, Layfield LJ. Macrodystrophia lipomatosa with associated fibrolipomatous hamartoma of the median nerve. *Pediatr Surg Int* 2000;16:216-8.
37. Walker CW, Adams BD, Barnes CL, Roloson GJ, FitzRandolph RL. Case report 667. Fibrolipomatous hamartoma of the median nerve. *Skeletal Radiol* 1991;20:237-9.
38. Langa V, Posner MA, Steiner GE. Lipofibroma of the median nerve: A report of two cases. *J Hand Surg* 1987;12:221-3.
39. Afshar A. Carpal tunnel syndrome due to lipofibromatous hamartoma of the median nerve. *Arch Iran Med* 2010;13:45-7.
40. Nilsson J, Sandberg K, Dahlin LB, et al. Fibrolipomatous hamartoma in the median nerve in the arm – an unusual location but with MR imaging characteristics: A case report. *J Brachial Plex Peripher Nerve Inj* 2010;5:1.
41. Hauck RM, Syracuse NY, Banducci DR, Hershey PA. The natural history of a lipofibromatous hamartoma of the palm: A case report. *J Hand Surg Am* 1993;18:1029-31.
42. Al-Jabri T, Garg S, Mani GV. Lipofibromatous hamartoma of the median nerve. *J Orthop Surg Res* 2010;5:71.
43. Gupta A, Geetha V, Monappa V, Bhat SS. Multiple neural fibrolipomas with macrodactyly. *Indian J Dermatol* 2011;56:766-7.
44. Murphy SM, Browne K, Tuite DJ, O'Shaughnessy M. Dual pathology proximal median nerve compression of the forearm. *J Plast Reconstr Aesthet Surg* 2013;66:1792-4.
45. Feriz H. Macrodystrophia lipomatosa progressiva. *Virchows Arch* 1925;260:308-68.
46. Ban M, Kamiya H, Sato M, Kitajima Y. Lipofibromatous hamartoma of the median nerve associated with macrodactyly and port-wine stains. *Pediatr Dermatol* 1998;15:378-80.
47. Matsubara M, Tanikawa H, Mogami Y, Shibata S, Uchiyama S, Kato H. Carpal tunnel syndrome due to fibrolipomatous hamartoma of the median nerve in Klippel-Tréaunay syndrome. *J Bone Joint Surg Am* 2009;91:1223-7.
48. Al-Qattan MM. Lipofibromatous hamartoma of the median nerve and its associated conditions. *J Hand Surg (Br Eur Vol)* 2001;26B:368-72.
49. Gennaro S, Merciadri P, Secci F. Intraneural lipoma of the median nerve mimicking carpal tunnel syndrome. *Acta Neurochir* 2012;154:1299-301.
50. Marom EM, Helms CA. Fibrolipomatous hamartoma: Pathognomonic on MR imaging. *Skeletal Radiol* 1999;28:260-4.
51. De Maeseneer M, Jaovisidha S, Lenchik L, et al. Fibrolipomatous hamartoma: MR imaging findings. *Skeletal Radiol* 1997;26:155-60.
52. Sagtas E, Orguc S, Elcin F, Memis A, Menzilcioglu S. Quiz case of the month. *Eur Radiol* 2000;10:547-8.
53. Boren WL, Henry RE Jr, Wintch K. MR diagnosis of fibrolipomatous hamartoma of nerve: Association with nerve territory-oriented macrodactyly (macrodystrophia lipomatosa). *Skeletal Radiol* 1995;24:296-7.
54. Jung SN, Yim Y, Kwon H. Symmetric lipofibromatous hamartoma affecting digital nerves. *Yonsei Med J* 2005;46:169-72.
55. Cavallaro MC, Taylor JA, Gorman JD, Haghighi P, Resnick D. Imaging findings in a patient with fibrolipomatous hamartoma of the median nerve. *Am J Roentgenol* 1993;161:837-8.