

Original Article

Fibrolipomatous Hamartoma of the Median Nerve: An Outcome of Surgical Management in Six Consecutive Cases

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INTRODUCTION

Lipoma is a common type of benign tumor, which can occur anywhere in the body, hence called a universal tumor. Fibrolipomatous hamartoma is one of the variants of this universal tumor, where the mature adipocytes proliferate into the peripheral nerve, resulting in a palpable mass. Other synonyms of fibrolipomatous hamartoma are intraneural fibrolipoma, fatty infiltration, fibrofatty proliferation, and lipofibroma.^[1] All these have been grouped under lipomatosis of the nerve by the World Health Organization in 2002.^[2] Lipomatosis of the nerve is a very rare condition; it can manifest as a soft, slow-growing mass in the volar aspect of the hand,

wrist, and forearm of young people.^[3] It constitutes <5% of benign tumor.^[4] Females are more affected compared to males.^[1,4] Lipomatosis of the nerve most commonly involves the median nerve^[5-7] followed by radial^[8,9] and ulnar.^[5,6,10] The affection of fibrolipomatous hamartoma toward median nerve has not been clearly explained till date.^[11] There are some reports of lipoma involving the brachial plexus,^[12,13] suprascapular,^[14] and supraclavicular

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ABSTRACT

Background: Lipoma is a nonneurogenic benign tumor. Neurolipoma and fibrolipomatous hamartoma are variants of this universal tumor. All these variants are grouped under lipomatosis of the nerve. Majority of these tumors are asymptomatic, which can be observed. Symptomatic patients require surgery, which is not standardized. As there are insufficient number of cases, no randomized controlled studies have been performed in the treatment of fibrolipomatous hamartoma. The aim of our study was to determine the pattern of presentation of fibrolipomatous hamartoma, surgical management offered, and the outcome in the form of recovery and complications. **Materials and Methods:** This retrospective descriptive study includes six patients diagnosed with fibrolipomatous hamartoma over a period of 12 years. Patient details were collected from the medical records. Patients diagnosed of fibrolipomatous hamartoma in the hand were included. Patients with other soft-tissue tumors were excluded from the study. Out of six patients, four required excision of nerve followed by reconstruction using sural nerve graft and two underwent microsurgical dissection of neural element. Patients were instructed to take care of the operated hand during the recovery phase. Institutional physiotherapy protocol was started during the 3rd postoperative week. Follow-up period was between 1 and 3 years. **Results:** All the six patients were free from symptoms postoperatively. Minimal complications were noted in two patients, which were managed conservatively. **Conclusion:** Surgical excision of fibrolipomatous hamartoma of median nerve below elbow, with nerve dissection or with nerve reconstruction using sural nerve graft, followed by proper postoperative care and physiotherapy has proven beneficial for the patients in our study.

KEYWORDS: Benign neural tumor, fibrolipomatous hamartoma, lipoma, median nerve, nerve graft, neurolipoma

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nerve.^[15] Cases with involvement of lower extremity are rare,^[3] however literatures have reported the lesion at posterior tibial^[16] and fibular nerves.^[17] Sometimes, these can be associated with bone overgrowth and macrodactyly.^[1,3]

Even today, not much randomized controlled studies have been performed in the treatment of lipomatosis of nerve,^[11] and hence it is not standardized. Few cases which are reported in literature had catastrophic motor and sensory deficits following complete nerve resection.^[18] To our knowledge, not many cases of lipomatosis of the nerve arising from the median nerve, wherein the involved nerve has been excised and reconstructed with a nerve graft, are reported. The aim of the study was to determine the pattern of presentation of fibrolipomatous hamartoma, surgical management offered, and the outcome in the form of recovery and complications.

MATERIALS AND METHODS

This retrospective descriptive study includes six patients diagnosed with fibrolipomatous hamartoma who were treated at the Department of Plastic Surgery, SDM College of Medical Sciences and Hospital, over a period of 12 years from January 2004 till December 2015.

Details such as age, sex, site and size of swelling, associated pain, numbness and tingling sensation, nerve gap following surgery, treatment offered, and complications following surgery, if any, were noted from patients' medical records [Table 1]. All the patients underwent magnetic resonance imaging (MRI) apart from routine preoperative investigations. Patients

diagnosed of fibrolipomatous hamartoma in hand were included. Patients with other soft-tissue tumors were excluded from the study. Written informed consent was obtained from all patients. Injection cefotaxime was administered intravenously as a prophylactic antibiotic. Out of the six patients, four required excision of nerve followed by reconstruction using sural nerve graft and two underwent microsurgical dissection of neural element. Patient follow-up was between 1 and 3 years. During the recovery phase, patients were instructed to take care of the operated hand, until a protective sensation was obtained. Patients were subjected to physiotherapy after 4 weeks [Table 2]. We analyzed the outcome of the procedure in the follow-up period in terms of recovery of their symptoms and complications in the form of hypoesthesia, trophic ulcer, and muscle atrophy, if any, were noted.

Surgical technique

All the cases were explored under regional anesthesia. Under pneumatic tourniquet, incision was made over the swelling. Skin flaps were raised on either side. The median nerve was identified and traced from normal segment to the lesion. Normal nerve branches were isolated. The carpal tunnel was released when required. Microdissection was performed using a microscope with $\times 5$ magnification. All neural layers were opened and Tumor excised [Figure 1]. The area where interfascicular dissection and separation of tumor from the nerve was not possible, was resected along with the nerve, and the cut ends of the nerve were reconstructed using a sural nerve graft [Figures 2 and 3]. Nerve

Table 1: Patient details

Case number	Age	Sex	Site of the swelling	Symptoms	Size of the swelling (cm)	Nerve gap after excision (cm)	Treatment offered	Complication
1	35	Male	Ulnar aspect of the left index finger	Pain	2×3	Nil	Microsurgical dissection	Hypoesthesia
2	30	Female	Left palm	Pain	3×4	2	Sural nerve cable graft from the palm to the index and middle fingers	
3	12	Male	Right palm	Pain	4×4	3.5	Sural nerve cable graft from the palm to the middle and ring fingers	
4	34	Male	Right palm and distal forearm	Pain, numbness, and tingling along the index and middle fingers	16×4	15	Sural nerve cable graft from the distal end of the forearm to the index and middle fingers	Minimal thenar muscle atrophy
5	20	Male	Radial aspect of the right index finger	Pain	2×1.5	Nil	Microsurgical dissection	
6	22	Female	Radial aspect of the left middle finger	Pain, numbness, and tingling along the middle finger	3×2	2	Sural nerve graft	

Table 2: Institutional physiotherapy protocol following nerve repair

4 th week onward (after removal of slab)
Scar mobilization and ultrasound for the scar
Active movement up to pain-free ROM of the involved joint/s
Relaxed passive movements of the involved joint/s
IG stimulation for the paralyzed muscles
To protect/take care of anesthetized part
To maintain elevation
Daily wash and cream application
Corrective splint
Full ROM active movements for the uninvolved joints
5 th week onward
Same as above
Gradual stretching (mild) of the tightened soft tissues passing over the involved joints
Gradual mobilization of the involved stiff joints
Gradual strengthening (mild) of the nonparalyzed muscles passing over the involved joints
Use of adaptive devices for daily activities of living
6 th week onward
Same as above
Gradual stretching (moderate) of the tightened soft tissues passing over the involved joints
Gradual mobilization (increase the grades) of the involved stiff joints
Gradual strengthening (moderate) of the nonparalyzed muscles passing over the involved joints
8 th week onward
Same as above
Severe stretching of the tightened soft tissues passing over the involved joints
Gradual mobilization (increase the grades)/manipulation of the involved stiff joints
Full strengthening of the nonparalyzed muscles passing over the involved joints
Once protective sensation comes back
Sensory re-education
Proprioceptive retraining
Once the signs of re-innervation are seen
Faradic type of surged current stimulation for the re-innervated muscles
Other facilitating techniques for the re-innervated muscles
Activities of daily living retraining

ROM: Range of motion

reconstruction was performed under the microscope using 9-0 nylon. The incision was closed in layers. The operated hand was immobilized in functional position.

RESULTS

In our retrospective descriptive study with a sample size of six patients, age group of the patients varied

from 12 to 35 years. Majority of the patients were males, forming 66.6% ($n = 4$) of total patients, and females formed the remaining (33.3% [$n = 2$]). Nearly 50% ($n = 3$) of the patients had swelling in the digits, 33.3% ($n = 2$) of them in the palm, and 16.6% ($n = 1$) in the proximal part of the palm extending to the distal part of the forearm. Size of the swelling varied from 2 cm × 1.5 cm to 12 cm × 4 cm. All the six patients complained of pain in the swelling. Almost 33.3% ($n = 2$) of cases who have presented with palmar swelling had numbness and tingling sensation in the median nerve territory of the digits along with pain. Nearly 33.3% ($n = 2$) of the cases were treated by excision of tumor and microsurgical dissection, which did not require nerve repair. Rest of the patients (66.6% [$n = 4$]) had a nerve gap ranging from 2 to 10 cm, which was treated by a sural nerve cable graft in three cases and a sural nerve graft in one case. Postoperatively, all the six patients were free from pain. There were no surgical complications in 66.6% ($n = 4$) of the patients. Two cases had minor complications. One patient had hypoesthesia over the ulnar aspect of the left index finger. However, protective sensation was well preserved, and the patient had no issues regarding the reduced sensation. Other patients had minimal thenar muscle atrophy with normal range of movement of the hand and were managed by physiotherapy [Table 3]. None of the four patients who underwent nerve reconstruction by sural nerve cable graft had donor-site morbidity. All the patients presented in this series had early recovery from the symptoms, and there were no postoperative problems with regard to sensation of the digits. Histological section reports confirmed the clinical diagnosis of fibrolipomatous hamartoma.

DISCUSSION

Adipose tumors arising from the peripheral nerve are uncommon.^[1,11,18,19] Intraneural lipoma, fibrolipomatous hamartoma of the nerve, fatty infiltration, and neural fibroma are grouped under one broad heading by the World Health Organization.^[1,4] Although intraneural lipoma and fibrolipomatous hamartoma of the nerve are the subsets under lipomatosis of the nerve, they do differ in their morphology and management.^[1] Among peripheral nerves, most of the lipomatosis of the nerve occurs commonly in the median nerve,^[5-7] the reason of which is unknown.^[11] The first case of intraneural lipoma was reported by Morley in 1964.^[11] “True intraneural lipoma” was the term coined by Rusko and Larson for the tumors that could be separated from the neural elements and showed the pathological characteristics of a benign lipoma without intermingling with neural elements.^[1,4]

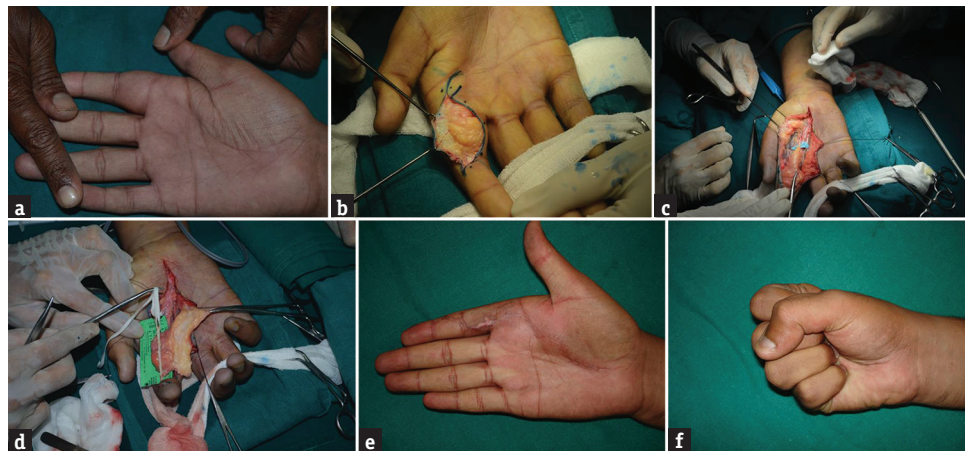


Figure 1: (a) Swelling in the right index finger (b) Elevation of skin flap (c) Tumor along the radial side of the digital nerve of the right index finger (d) Isolation of digital nerve following microdissection (e) Postoperative result (f) Good range of movement of the hand

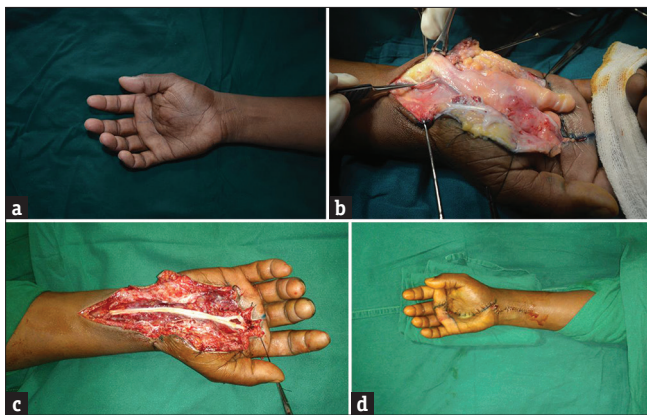


Figure 2: (a) Swelling in the right palm (b) Tumor arising from the median nerve (c) Reconstruction of the median nerve using sural nerve graft (d) Closure of the wound

Fibrolipomatous hamartoma was first described by Mason in 1953.^[1] The etiology is not clear.^[1] However, several potential etiologic factors have been described such as history of trauma, abnormal development of flexor retinaculum in children, and chronic nerve inflammation.^[1] Although it can present in childhood and early adulthood, it is believed to be of congenital origin.^[1] Although all these have been grouped under lipomatosis of the nerve, intraneural lipoma differs from fibrolipomatous hamartoma of the nerve. Intraneural lipomas commonly present as well-encapsulated tumors with normal nerve fibers running on the external surface of the tumor. Excision of these kinds of tumors in toto is possible with no damage to the adjoining nerve, whereas fibrolipomatous hamartoma of nerve is a composition of fibrous tissues, adipose tissues, and normal nerve fibers. Complete excision of tumor causing no damage to the adjoining nerve is not possible.^[1,4]

Patient's presentation depends on two different ways of adipose tissue involvement to peripheral nerve: either

Table 3: Clinical findings and outcome

Sample size: Number of patients (n=6)
Age: Varied from 12 to 35 years
Sex
Male: 66.6% (n=4)
Female: 33.3% (n=2)
Presentation:
Pain: 100% (n=6)
Numbness and tingling sensation: 33.3% (n=2)
Swelling
Digits: 50% (n=3)
Palm: 33.3% (n=2)
Proximal to palm: 16.6% (n=1)
Size
Varied from 2 cm×1.5 cm to 12 cm×4 cm
Treatment
Excision of the tumor+microdissection: 33.3% (n=2)
Excision of the tumor+sural nerve graft reconstruction: 66.6% (n=4)
Outcome
Free from pain: 100% (n=6)
Complications: 33.3% (n=2)
Hypoesthesia of the ulnar aspect of the left index finger
Minimal right thenar muscle atrophy

by direct compression by an extraneural lipoma or by a lipoma originated from the adipose cells located inside the nerve.^[4] They may complain increasing pain in the median nerve territory, tenderness, diminished sensation, or paresthesia associated with the gradually increasing mass, causing compression neuropathy. Carpal tunnel syndrome is a late complication of some lesions. There may be loss of muscle strength. Lipomatosis of the peripheral nerves poses problems in diagnosis. MRI is the gold standard in proving the diagnosis.

Treatment of lipomatosis of the nerve is controversial.^[11,18,19] Due to lack of randomized

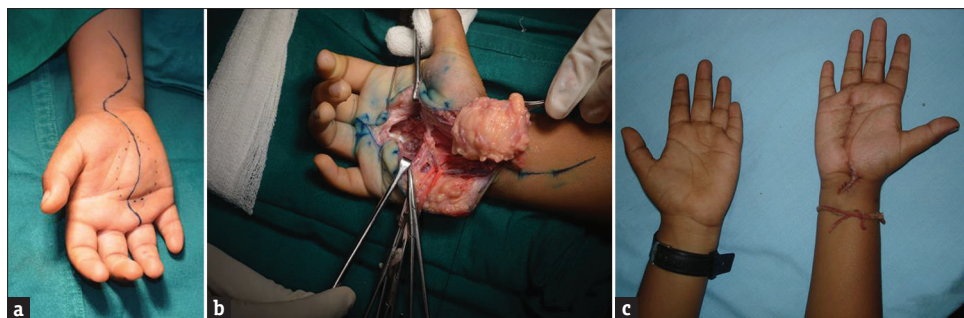


Figure 3: (a) Swelling in the right palm and marking of incision (b) Resection of tumor along with the involved nerve segment (c) Final postoperative results

controlled studies on the treatment of lipomatosis of the nerve, controversy still arises regarding the optimal approach to the problem.^[11] Both the surgeon and the patient must weigh the potential risks and benefits of surgery against those of conservative management. The conservative approach in managing fibrolipomatous hamartoma includes carpal tunnel decompression, fibro fatty sheath debulking, microsurgical dissection of the neural elements, and observation in asymptomatic patients. The definitive approach includes excision of the involved nerve with or without grafting.

However, the risk of definitive approaches is largely related to the location and size of tumor, degree of involvement, age of the patient, and surgical technique.^[5,20] Excision of median nerve at or above the elbow is associated with a high morbidity and is not recommended.^[19] Fibrolipomatous hamartoma-associated macrodactyly requires excision of the involved nerve segment and reconstruction of the nerve with or without nerve grafting. These cases may also demand amputation of the involved digit at times.^[8,21] To restore the nerve function in cases, wherein the tumor has already been insulted in major motor disturbance, conservative approaches may not help and require definitive surgical approach.

In our study, all the six patients were relieved from pain and swelling. Two patients who had hypoesthesia preoperatively were relieved from their symptoms 3–6 months after the surgery. One patient had postoperative hypoesthesia over the ulnar aspect of the left index finger. The exact reason for the complication was unknown. We think that the most probable causes for the above complication would be (a) excessive traction during surgery, leading to prolonged neuropraxia, which did not recover, and (b) vascular compromise of the nerve, provoking an intense healing response, which may, in turn, jeopardize the neurological function.

Another complication noted was minimal thenar muscle atrophy, which might have been caused due to long nerve

graft, causing delay in neurotization of the thenar muscle. However, patients had normal range of movement of hand. This was managed with physiotherapy. The limitation in our study was that we have not objectively evaluated the hand functions in the pre and postoperative period using any kind of scoring system. During the follow-up period of 1 to 3 years, there was no recurrence of the tumor. None of the four patients who underwent nerve reconstruction by a sural nerve cable graft had donor-site morbidity. This strengthens the fact of sural nerve being an expendable nerve.

CONCLUSION

Fibrolipomatous hamartoma is a benign condition, which can predominantly occur anywhere in the median nerve territory. Surgical intervention has shown to be beneficial in treating symptomatic patients. We opine from our limited case series that surgical excision of fibrolipomatous hamartoma of the median nerve below the elbow with micronerve dissection or with nerve reconstruction using sural nerve graft, followed by proper postoperative care and physiotherapy, has proven helpful for patients.

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Conflicts of interest

There are no conflicts of interest.

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