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Case Report

Neurofibrolipoma of the dorsum of hand: a case report

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ABSTRACT

Neurofibrolipoma, also called fibrolipomatous hamartoma, is a rare benign lesion which commonly involves the upper limb and its nerves with the median nerve being frequently involved. It also affects other nerves such as the ulnar, radial and brachial plexus.

Keywords: Benign, Hamartoma, Rare, Surgical excision

INTRODUCTION

Neurofibrolipoma, also called fibrolipomatous hamartoma, is a rare benign lesion which commonly involves the upper limb and its nerves with the median nerve being frequently involved. It also affects other nerves such as the ulnar, radial and brachial plexus.[1-3] Clinically, it presents as a soft, slow-growing, fusiform swelling usually accompanied by pain, redness and decreased sensation. Here, we report a case of a child presenting with a neurofibrolipoma of the dorsum of left hand which was surgically excised without post-operative deficits.

CASE REPORT

A 2-year-old male child was brought by his mother to the department of plastic surgery with swelling over the dorsum of left hand for the past 1.5 years. The child was born to parents of non-consanguineous marriage by normal full term vaginal delivery. The mother noticed a small swelling on the dorsum of left hand from the age of 6 months and has gradually progressed to attain the present size. There is no history of pain, bleeding or ulceration of the swelling. On examination, the boy was active with stable vitals and a 6×5×3 cm swelling over the dorsum of the left hand also involving the volar aspect of the ulnar border (Figure 1 and 2).

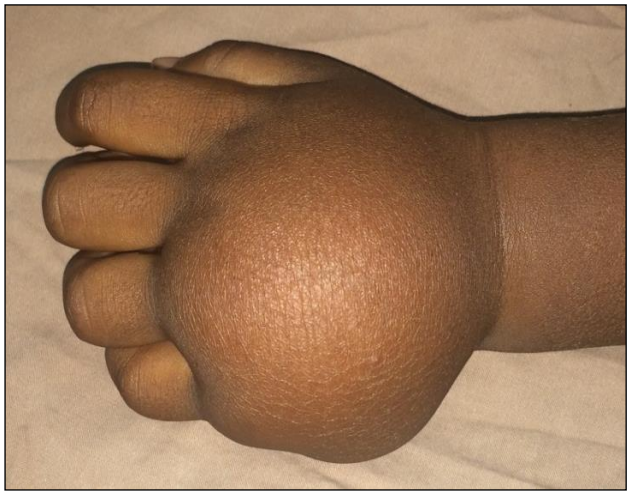

Figure 1: Clinical picture showing swelling on the dorsum of left hand.

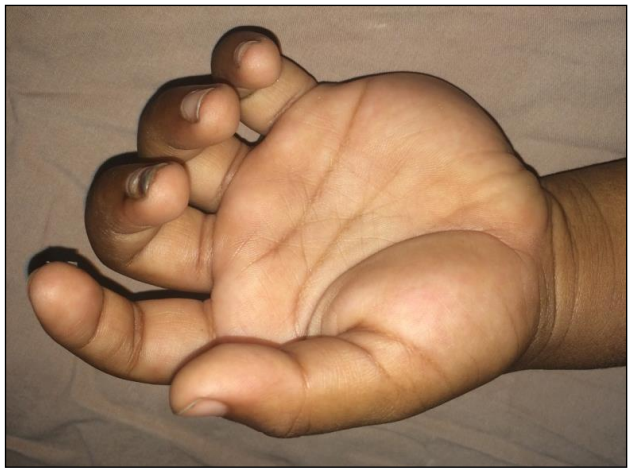

Figure 2: Clinical picture showing swelling involving the volar and ulnar aspects of left hand.

The swelling was non-tender and soft to firm in consistency. It was mobile with the overlying skin being free. Finger extensors were normal with no distal neurovascular deficit. A clinical diagnosis of lipoma was made. MRI showed a completely fat containing lesion in the medial aspect of the hand involving the volar and dorsal aspects and infiltrating the hypothenar muscles. It is completely encasing the extensor tendons of the 3rd, 4th and 5th fingers with partial involvement of the dorsal interossei muscle of 2nd, 3rd and 4th space on T1 weighted images, features suggestive of diffusely infiltrating benign lipomatosis of the left hand (Figure 3a and 3b).

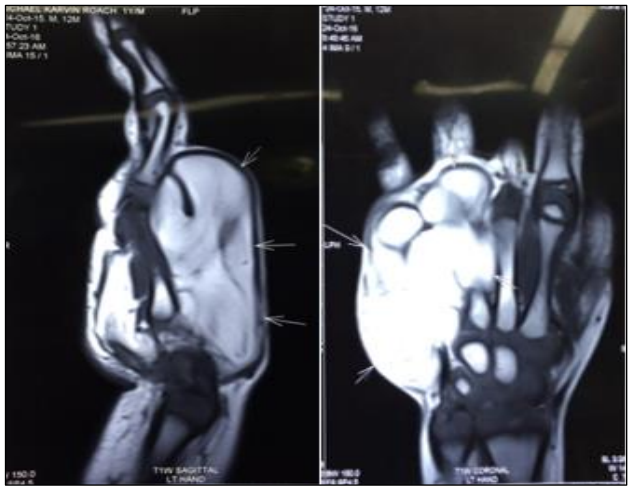

Figure 3a and 3b: MRI showing fat containing.

We proceeded for surgical excision. Under general anaesthesia and tourniquet control, a lazy S incision was made over the swelling and deepened in layers (Figure 4). The lesion was dissected off the extensor tendons taking precaution to preserve them and excised in toto (Figure 5). The lesion had a fibrous component to it. The specimen was sent for histopathological examination (Figure 6). After securing haemostasis following tourniquet removal, the excess skin was excised and then closed with 3-0 nylon over tube drains. A compression was applied. Post-operative period was uneventful with patient discharged on the 2nd post-operative day (Figure 7) and suture removal done on the 7th post-operative day. Patient was reviewed after 1 month and the scar was soft and supple (Figure 8).

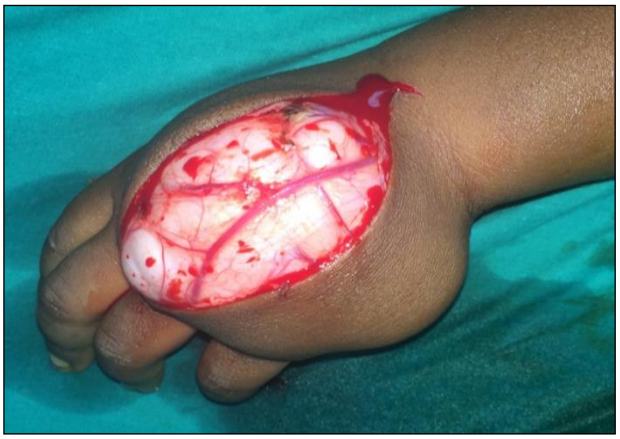

Figure 4: Lazy S incision made on the dorsum of left hand.

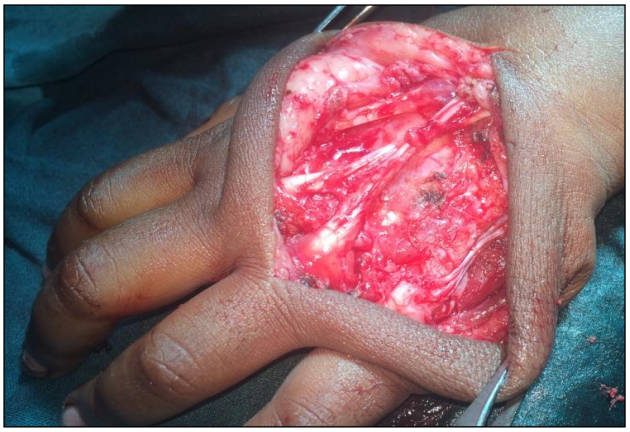

Figure 5: After complete excision of the lesion showing intact extensor tendons.

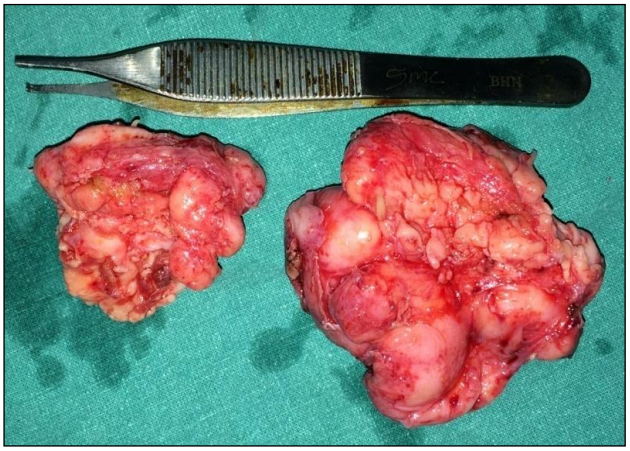

Figure 6: Specimen after excision.

Figure 7: Clinical picture on the 2nd post-operative day.

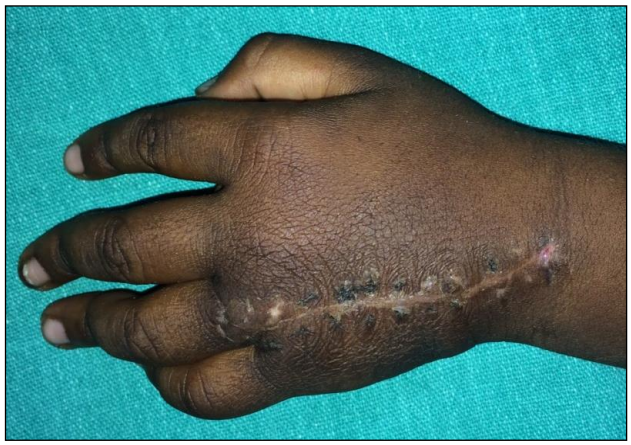

Figure 8: 1-month post-operative follow-up picture.

Histopathology showed a lesion composed of lobules of mature adipose tissue with fibrous septae and scattered areas showing short spindle cells in sheets and fascicles with few congested blood vessels, features suggestive of neurofibrolipoma (Figure 9 and 10).

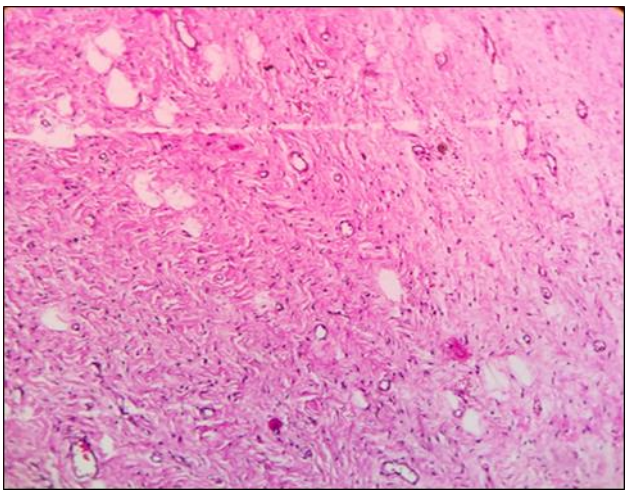

Figure 9: Microscopy showing predominantly spindle cells with wavy nuclei and scattered adipocytes 10x.

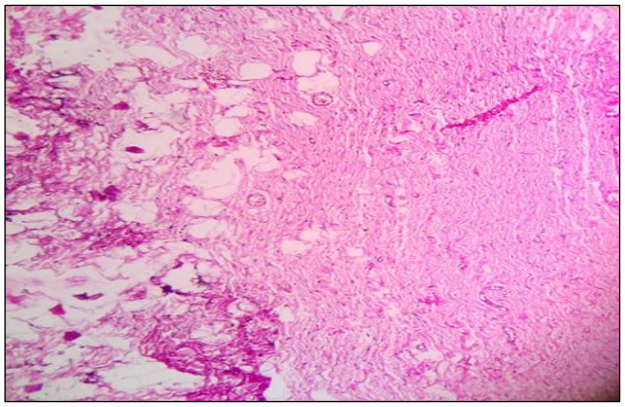

Figure 10: Microscopy showing mature adipocytes with scattered neural cells and fibrous plaques 40x.

DISCUSSION

Neurofibrolipoma is a proliferation of mature adipocytes within and around the peripheral nerves. When neurofibrolipoma is associated with overgrowth of bone and macrodactyly, it is known as macrodystrophia lipomatosa. It was first described by Feriz and Barsky and can be complicated by hamartomas in the palm, dorsum and forearm.[1] Median nerve is the most commonly affected nerve with the other affected nerves being ulnar, radial.[2,3] Few isolated cases and cranial nerve involvement have also been reported.[4]

There are many differential diagnoses of macrodactyly, both acquired and congenital, namely dactylitis, infarction, Still's disease, osteoid osteoma, lymhangioma and hemangioma.[5]

Major clinical features of neural fibrolipomas include gigantism of a digit, hands, feet or whole extremity since birth.[6] There are mostly always seen during the first three decades of life. History of pain, tenderness, decreased sensation, paraesthesia with enlarging mass causing compression neuropathy may be present. Carpal tunnel syndrome is a late complication seen in some cases. Radiological imaging findings include enlargement, lipomatous infiltration and coaxial cable appearance of peripheral nerves.[4]

On gross examination, neurofibrolipomas present as a soft, gray-yellow, fusiform, mass that has diffusely infiltrated parts of a large nerve and its branches, varying from small simple lesion to extensive complex lipomatosis involving all the nerves of the upper extremity.[3] Microscopy reveals mature adipose and fibrous tissue infiltrating epineural and perineural compartments. The involved nerves may show pseudo-onion bulb and metaplastic new bone formation.[7]

The pathologic differential diagnosis include neuromas, lipomatous neurofibromas and diffuse lipomatosis. Morton's neuroma reveals degenerative changes, edema and fibrosis of nerve. Lipomatous neurofibromas are

smaller dermal lesions which shows interlacing fascicles or whorls of uniform, delicate spindle cells regularly interspersed with adipose tissue on microscopic examination.[8] In diffuse lipomatosis, there is diffuse overgrowth of mature adipose tissue affecting large portions of extremity or trunk with no involvement of nerves.[9]

There is no standard therapy for neurofibrolipoma. Complete excision of fibrofatty tissue is avoided as it may cause sensory or motor disturbances.[9]

CONCLUSION

Neurofibrolipoma is a rare lesion with a multitude of clinical presentations ranging from a small lesion to a large mass involving the entire extremity with or without macrodactyly. Hence, a thorough knowledge of the characteristic histological and radiological features is crucial for accurate diagnosis and treatment.

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