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Lipofibromatous Hamartoma of the Finger presenting as Congenital Macrodactyly – A Rare Case Report

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Abstract
Hamartomas are benign malformations composed of tissue elements normally found at that site, but which are growing in a disorganized fashion. Lipofibromatous harmartomas (LFH) are rare lesions that require early diagnosis and treatment. LFH is characterised by fibrofatty infiltration around the nerve fascicles which differentiates them from other intraneural tumours. They usually present as macrodactyly or later as a mass lesion or symptoms due to compressive neuropathy. MRI is an important diagnostic tool confirmed with a tissue biopsy. Treatment for macrodactyly is generally debulking, joint reconstruction, amputation or prophylactic decompression of affected nerves at all points of possible compression.

Key Words: *Benign, Lipofibromatous, Hamartoma, Rare, Macrodactyly, Surgery*

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I. Introduction

Lipofibromatous hamartomas (LFH) was first reported by Mason in 1953 and then by several others.[1-4] LFH are rare benign tumors of childhood, usually occurs in the first 2 years of life presenting as congenital macrodactyly.[5] Enzinger in 1965 and later Larralde de Luna et al. in 1990 described the term 'lipofibromatous hamartoma of infancy' with lesion in precalcaneal region with similar morphology.[5,6] Synonyms for the LFH include fatty infiltration, intraneural lipofibroma, fibrofatty proliferation, lipofibroma and hamartoma, but LFH is currently the most accepted name.[3,7-11] The symptoms are generally due to compressive neuropathy and hence surgical decompression is required most of the time.

II. Case Report

21 year old female presented to us with a swelling of the left index finger since birth. The swelling gradually increased in size and was not painful till 1 month ago. There was no history of co-morbidities. On examination, a swelling measuring 5 x 1.5cm over the radial aspect of left index finger extending from the DIP joint till base of proximal phalanx was present. The swelling was not warm or tender with variable consistency and not fixed to underlying tendon or bone. **(Fig. 1)** The skin was free with no restriction in movement of the finger and no distal neurovascular deficit. A clinical diagnosis of a congenital soft tissue lesion was made. MRI showed a well-defined soft tissue lesion in the radial aspect of left index finger with fat signal intensity in the form of bright T1 and T2 weighted images suggestive of macrodystrophica lipomatosa. **(Fig. 2)** We planned for exploration and excision of the lesion under axillary block, tourniquet control and loupe magnification. The finger was explored through curvilinear incision over the swelling along the neutral line. **(Fig. 3)** A 10 x 2 x 1cm lipomatous growth with fibrofatty tissue left index finger was identified with both the flexor tendons and the extensor tendons intact with no bony deformity. The nail plate complex was normal and the digital

neurovascular bundle was intact. The fibrofatty tissue was excised safeguarding the neurovascular bundle. **(Fig. 4)** Haemostasis was secured and excess skin was trimmed and closed with 4-0 nylon sutures. **(Fig. 5)** Finger bandage was applied. Microscopy showed a lesion composed of lobules of mature adipose tissue and fibrous tissue interspersed with collections of hypertrophied nerve bundles and few capillary sized blood vessels, features consistent with lipofibromatous hamartoma.

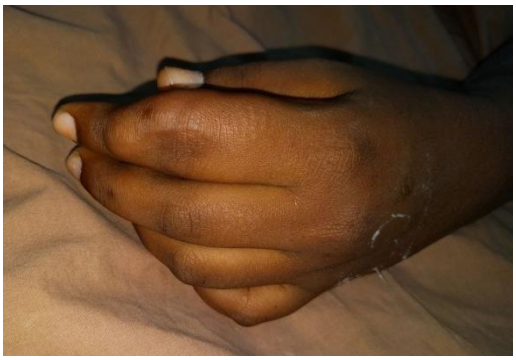

Fig. 1 – Clinical photograph showing the lesion

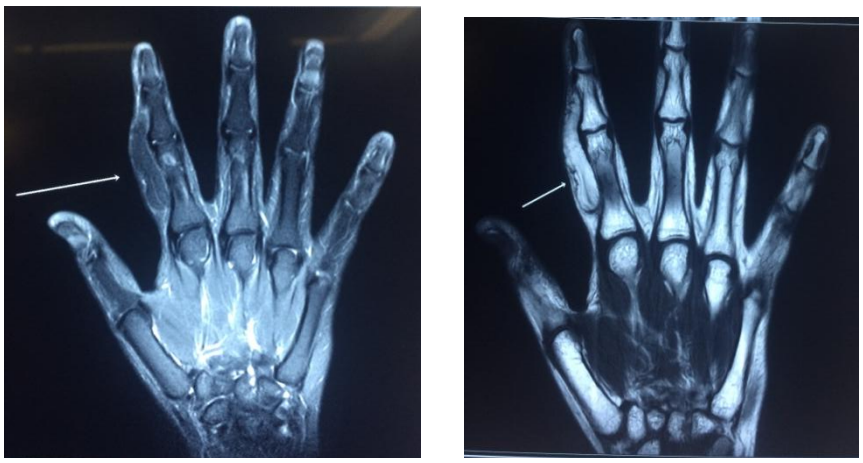

Fig. 2 – MRI findings of the lesion in T1 and T2 weighted images

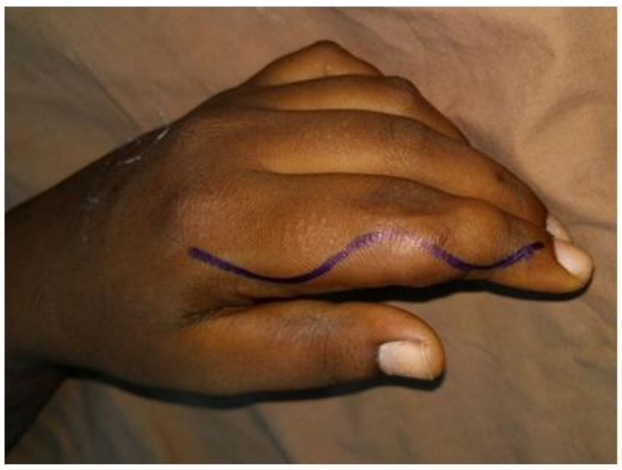

Fig. 3 – Photograph showing the incision marking

Fig. 4 – Intra-operative photographs showing the lesion being excised

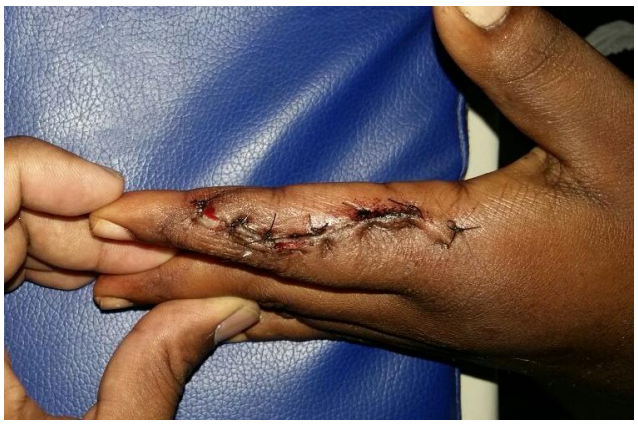

Fig. 5 – Immediate post-operative picture showing the suture line

III. Discussion

Lipofibromatous hamartoma (LFH) is a rare benign intraneural tumour, affecting the peripheral nerves, most commonly in the upper extremity. LFH is associated with macrodactyly in more than one-third of cases presenting in early childhood or later in adolescence.[12,13] Symptoms consistent with compressive neuropathy of affected peripheral nerves can also occur later in life. Many etiologic factors have been proposed for LFH which includes abnormal development of flexor retinaculum in children,-antecedent trauma-and chronic nerve irritation.[8,9,14] Several authors believe that LFH is a congenital condition.[3,12] It most commonly manifests among caucasian population and usually presents before the third decade.[15,16] It occurs sporadically and has no association with neurofibromatosis.[17] LFH commonly results in digital enlargement due to an increase in perineural soft tissue and skin. When accompanied by true macrodactyly, the tumour is referred to as macrodystrophia lipomatosa. More than one-third of LFHs are associated with unilateral macrodactyly.[12,13] Females are more commonly affected when macrodactyly is present.[12,13] The distal peripheral nerves are most often affected in LFH; cranial nerve involvement is rare.[18] The macrodactyly which is associated with LFH can be present at birth or can be diagnosed during early childhood.[19] Macrodactyly can be symmetric involving the radial or ulnar sides of the same digit equally or asymmetric. Although the phalanges are usually affected, the metacarpals are rarely involved. According to Dell, the involvement of metacarpals commonly occurs in older patients and is a measure of the severity of the disorder.[19] A detailed history and physical examination are vital in making the diagnosis of LFH. Plain radiographs are valuable for assessing changes in the underlying skeleton when macrodactyly is present. MRI plays a major role in confirming the diagnosis of LFH and also provides a detailed assessment of nerve involvement preoperatively. A characteristic feature on MRI is the appearance of serpentiform nerve fascicles surrounded by fibrofatty tissue within the expanded nerve sheath.[20] Distribution of fat between fascicles is asymmetric. On coronal images, the nerve has a spaghetti-like appearance that is pathognomonic for LFH.[21,22] Gross pathological examination shows that it consists of lobulated, soft, grey-yellow, sausage-shaped masses within the epineural sheath. Nerves may be markedly increased in length and diameter in the involved area and the affected peripheral nerves do not adhere to surrounding tissue.[14,21] In few cases, hypertrophy of surrounding skin and soft tissue is associated with the enlargement of the bone (macrodystrophia lipomatosa).[14] Microscopic examination shows infiltration of the epineurium by fibrofatty tissue, which separates and compresses the nerve fascicles causing atrophy of the neural elements, often accompanied by

extensive perineural fibrosis and septation.[12,24,25,30] If LFH is associated with gross macrodactyly in an adult, the preferred procedure is either debulking or amputation of the finger. Amputation of the finger is less frequently indicated in childhood.[19] In cases of LFH without macrodactyly, a conservative management that includes decompression of all compromised peripheral nerves to reduce pain and paresthesia and to decrease permanent motor and sensory sequelae.[8,11] Some patients require other reconstructive procedures like joint debridement, excision of osseous overgrowth and tendon transfers aimed at improving hand function.

IV. Conclusion

LFH are benign tumours occurring in children involving the peripheral nerves. LFH differs from other intraneural tumours owing to its characteristic fibrofatty infiltration around the nerve fascicles. LFH requires early prophylactic decompression at points of potential compression in order to avoid permanent nerve damage. Patients with significant macrodactyly may benefit from debulking, joint reconstruction or amputation. Patients who present late with advanced disease may benefit from reconstructive procedures aimed at improving hand function. Radical excision of these tumours is not recommended.

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