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CASE REPORT

Fibrolipomatous hamartoma of sural nerve: a new site of an unusual lesion

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Abstract

Neural fibrolipomatous hamartoma is a rare benign tumour commonly involving the median nerve. Other less frequently involved nerves include the ulnar, radial, brachial plexus, superficial peroneal nerve, inferior calcaneal nerve and median plantar nerve. Involvement of sural nerve has not been reported in the available literature so far. A three-year-old female child presented with a painless swelling over the posterolateral aspect of left leg with no associated motor or sensory deficits. Radiological investigations revealed a fat density lesion with interspersed neural element in the subcutaneous plane of the left leg. Histopathological examination of the excised specimen showed features of a fibrolipomatous hamartoma of the nerve. This report describes the occurrence of fibrolipomatous hamartoma in the sural nerve for the first time in the literature. This rare tumour should be considered in the differential diagnosis of such lesions.

Keywords: hamartoma, fibrolipomatous, sural nerve, magnetic resonance imaging

INTRODUCTION

Fibrolipomatous hamartoma (FLH) is a rare, benign tumour of peripheral nerve.[1] Although FLH was first described in 1953,[2] less than 100 cases have been documented so far in the available indexed English literature.[1,3-5] Several terms have been used to describe this condition, including intraneural hamartoma, neural fibrolipomatosis, neural fibrolipoma and perineural lipoma.[4] Although considered by some to be of congenital origin,[1] the exact aetiology of fibrolipomatous hamartoma of the nerve still remains unclear. In FLH, the epineurium and perineurium are enlarged and distorted by mature adipose tissue and fibrous tissue that infiltrates and surrounds the nerve fascicles.[5]

Most cases of fibrolipomatous hamartoma occur within the first three decades of life.[1] This tumour has particular predilection for the median nerve and its branches leading to pain and sensori-motor deficits in the affected nerve distribution.[1] Other less frequently involved sites are the radial nerve, ulnar nerve and brachial plexus.[1, 3, 6] To the best of our knowledge, no case of fibrolipomatous hamartoma involving the sural nerve has been reported so far in the existing English literature.

CASE REPORT

A three-year-old girl presented with a one year history of swelling over the back of her left leg that was insidious in onset and gradually progressive. The swelling was painless and there was no associated difficulty in walking. On examination, a 4x3 centimetres, firm, non-tender swelling was present at the postero-lateral aspect of the left leg. The overlying skin was normal. Motor and sensory examination of the left leg was unremarkable.

Magnetic resonance imaging (MRI) showed a well-defined oblong fat density lesion in the subcutaneous plane of the posterolateral aspect of the lower third of the left leg. The lesion measured 32x24 millimetres and showed interspersed linear structures suggestive of neural element. The appearance was that of a lipoma with neural element (Fig. 1a & 1b).

An excisional biopsy was performed under general anaesthesia. Per-operatively, the lesion was 3.5x2.8 centimetres, firm to hard

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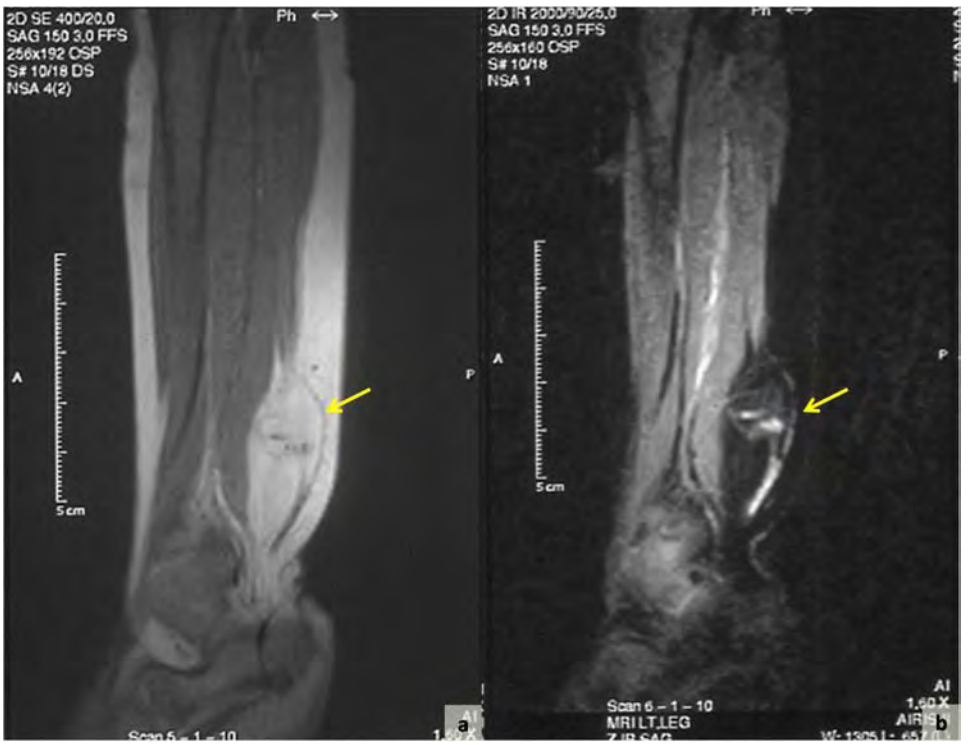

FIG. 1: Sagittal T1 (a) and T2-weighted, fat saturated (b) images show a well-defined oblong fat density lesion (arrows) with interspersed linear T2-hypointense structures (?neural elements) lying deep to the subcutaneous plane of posterolateral aspect of the left leg.

in consistency. The lesion was present in the subcutaneous plane and infiltrating the sural nerve. There was no capsule or pseudocapsule. As the sural nerve was infiltrated by the tumour, two centimetre length of the nerve was also excised. Post-operatively, patient did not report any sensory or motor deficit. The patient has been doing well in six months of follow-up.

Pathology

We received a grey-white to yellow soft tissue mass measuring 3.2x2.5x2 centimetres in size. The cut surface had a homogenous fatty appearance. The subcutaneous tissue resected along with the lesion was infiltrated by the tumour. Microscopical examination revealed an uncircumscribed lipomatous tumour surrounding the nerve fascicles (Fig. 2a). The tumour comprised of mature adipose tissue and fibrous tissue entrapping the nerve fibres and infiltrating the epineurium and perineurium of the nerve (Figs. 2b & 2c). It was accompanied by perineural fibrosis. The surrounding subcutaneous area was involved by the tumour. There was no neural hypertrophy or inflammation. A histological diagnosis of fibrolipomatous hamartoma was provided.

DISCUSSION

Fibrolipomatous hamartoma (FLH) is an unusual benign tumour composed of hypertrophied fibroadipose tissue intermixed with neural tissue. It commonly presents in childhood and early adulthood.[1] FLH affects the median nerve in majority of the cases; this predilection remains unexplained.[4,5] Other less frequently involved nerves are the ulnar nerve, radial nerve and brachial plexus in the upper extremity[3] and rarely, the superficial peroneal nerve,[7] inferior calcaneal nerve[8] and medial plantar nerve[9] in the lower extremity. Since this tumour is quite uncommon, there are only a few reported case series. The reported frequency of median nerve involvement ranges from 60 – 80% and that of ulnar nerve ranges from 10 – 40%.[3, 10] Our patient, a young female child, presented with involvement of the left sural nerve. Sural nerve involvement by FLH has not been reported earlier in the literature, to the best of our knowledge.

The majority of cases present with a longstanding painless mass. Neurological symptoms ensue after several years, mainly due to nerve compression.[5] Compression neuropathy leads to sensory and motor symptoms

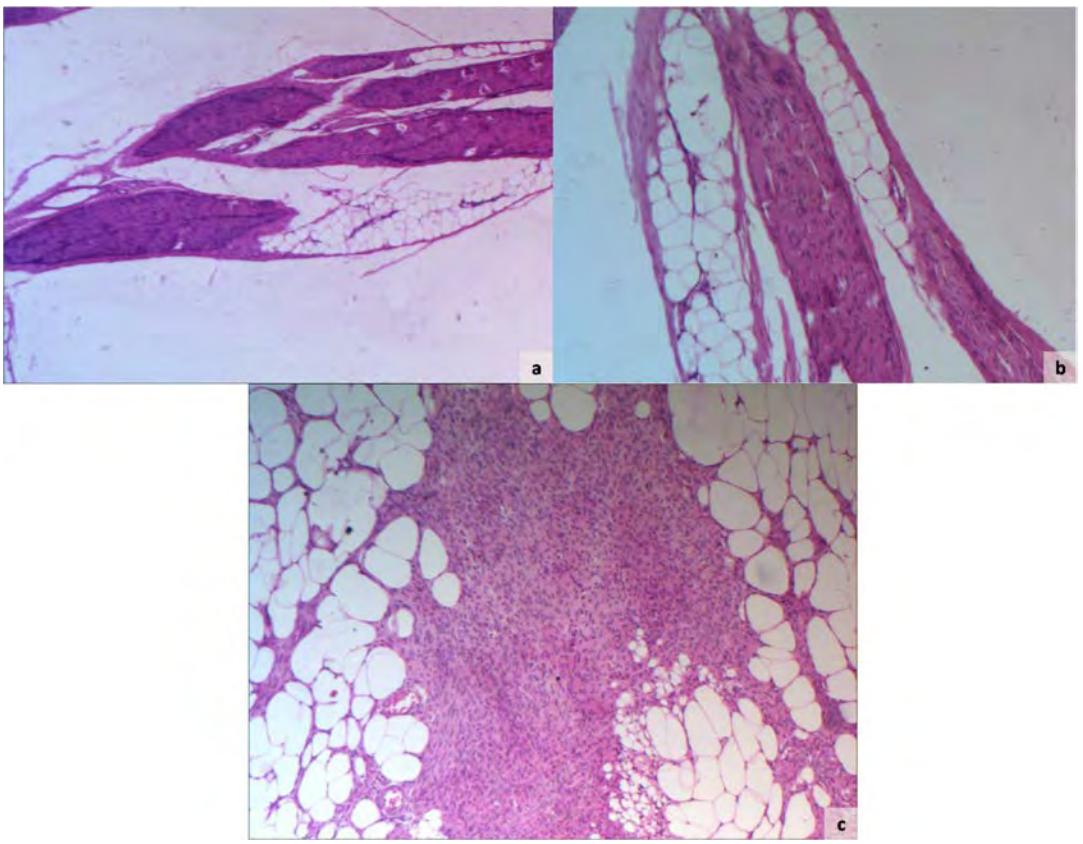

FIG. 2: Panel of photomicrographs showing a lipomatous lesion surrounding nerve bundles (a, H&E x100) with infiltration of the epineurium and perineurium (b, H&E x200). Another view shows fibrolipomatous component of the tumour (c, H&E x200).

including pain, tenderness, diminished sensation, paresthesia, and weakness.[4,5] These neurological symptoms are often slowly progressive. In our patient, the mass was asymptomatic without any neurological deficit. This presentation can also be due to the purely sensory function of the sural nerve. Therefore its involvement results in only a relatively trivial deficit, which could go unnoticed by a three- year-old child.

The appearance of FLH on magnetic resonance imaging (MRI) is now thought to be pathognomonic. The characteristic appearance of FLH on MRI is that of low-intensity nerve bundles surrounded by fibrolipomatous tissue, demonstrating high intensity on T1-weighted images and low intensity on T2-weighted images. On the axial cuts, the nerve fibres are often described as having a 'coaxial-cable appearance', whereas on coronal sectioning, they are described as resembling 'spaghetti'.[4] MRI, in our case, showed a predominantly fatty lesion on T1 and T2-weighted images with linear low-signal intensity structures resembling neural element, suggesting a diagnosis of lipoma with neural component. However, the characteristic features on MRI are reported with the lesions arising in the median nerve, which has a relatively larger cross-sectional area (5.2-9.6 mm^2).[11] It is not always possible to obtain accurate images of thin nerve fascicles, like that of sural nerve (cross-sectional area=1.10-1.78 mm^2), in the coronal or axial plane.[12,13] Consequently, it may be difficult to evaluate the typical appearance on MRI and to diagnose fibrolipomatous hamartoma in small nerves.

Confirmation of the diagnosis of FLH is by histological examination. The characteristic histopathological features are infiltration of the epineurium and perineurium by fibrofatty tissue, which separates and compresses the nerve fascicles; accompanied by extensive perineural fibrosis.[5] Our case demonstrated these findings rendering the diagnosis of fibrolipomatous hamartoma of sural nerve.

Other intraneural tumours which need to be distinguished from FLH are intraneural lipoma,

neurilemmomas and neurofibromas.[14] Intraneural lipomas are encapsulated and easily detached from the nerve fascicles, and show proliferation of mature adipocytes without intermingled nerve fibres. Neurilemmomas and neurofibromas are tumours of Schwann cells. Neurilemmomas are encapsulated tumours, extrinsic to the nerve fibres and show the characteristic alternating pattern of Antoni A and B areas. Neurofibromas are not encapsulated and small neurites are present within the tumour. Microscopical examination, however, shows interlacing bundles of spindle cells intimately associated with wire-like strands of collagen. FLH, in contrast, consists of fibrofatty tissue surrounding and infiltrating the epi- and perineurium of nerve trunk. Our case showed the characteristic intermingling of fibroadipose tissue and nerve bundles, diagnostic of FLH.

Management of fibrolipomatous hamartomas can be conservative or surgical, but no specified guidelines are currently available.[4,5] Complete excision of the tumour involving the major nerve trunk is not recommended because it may cause severe sensory or motor disturbances. Surgical intervention is done mainly for symptomatic management of this benign lesion. The main surgical options are decompression of the involved nerve or microsurgically dissecting the neural elements or excision of the nerve with or without grafting.[14] All of these treatment modalities are associated with some complications, including motor and sensory dysfunction and neurogenic pain. Decompression of the involved nerve and microsurgical dissection give better preservation of neurological function.[5] In our case, surgical debulking along with excision of the sural nerve, which has primarily sensory function, was performed.

The present case is being reported because of the rarity of the lesion and a previously unreported site of involvement. To the best of our knowledge, this is the first case report of fibrolipomatous hamartoma involving the sural nerve.

In conclusion, fibrolipomatous hamartoma of nerve is a rare benign tumour with frequent involvement of the median nerve. This report describes the involvement of the sural nerve for the first time. Radiological diagnosis may be difficult in small nerve involvement and hence, histopathological examination offers an accurate diagnosis.

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