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## Case report: Rare occurrence of Myofibroma in thigh

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### Abstract

Myofibroma is solitary or multiple nodular tumor of the soft tissue, bone or internal organs found in all ages, however predominant in 2-3 years of age. These lesions arise from benign fibroblast and myofibroblast proliferation containing biphasic presentation of spindle shaped cells surrounding central zone of less differentiated cells focally arranged in a hemangiopericytoma like pattern. Herein reported is a rare case of 82years male presented to Ramaiah hospital with swelling in the upper part of left thigh for 2 years. MRI of the hip joint revealed to be? Nerve sheath tumor? Hematoma. Trucut biopsy showed spindle cell lesion and the true nature of the lesion was established after complete excision of the lesion and histopathological examination.

**Keywords:** Myofibroma, predominant, hematoma, fibromatosis

### Introduction

Myofibroma is nodular tumour, present in single or multiple nodes, affects the soft tissue, bone, or internal organs that affects all ages. In 1951, Williams and Schrum<sup>[1]</sup> were the first to name the lesion as congenital fibro-sarcoma, in a subsequent study of fibrous growth in children, Stout<sup>[2]</sup> revised term to congenital generalized fibromatosis, to denote multi-centric and multi-nodular benign fibroblastic process composed of spindle cells. In 1965 Kauffman and Stout<sup>[2]</sup> grouped such tumors into two, first group affecting the skin, the subcutaneous tissue, the skeleton which are good prognosis or the second group which affect the soft tissue like muscles, bones, or internal organs are the poor prognosis. Later in 1989 Smith *et al.*<sup>[3]</sup> and Daimaru *et al.*<sup>[4]</sup> redefined the solitary variant in adults as myofibroma and myofibromatosis, respectively. Finally WHO adopted term<sup>[5]</sup> to describe solitary (myofibroma) or multicentric (myofibromatosis) as benign neoplasms of contractile myeloid cells arranged around thin walled vessels, cells with characteristics intermediate between smooth muscle, fibroblasts, and undifferentiated cells.

These lesions can occur in any age group, mainly in first decade of life, some reported 90% of cases occur before the age of 2 out of which 2/3<sup>rd</sup> were reported at the time of birth<sup>[6,7]</sup>. Foss and Ellis<sup>[7]</sup>, in a study (n= 79) reported that myofibroma was predominant in mean age group of 26 and myofibromatosis predominant in age group between 22years. These lesions clinically present with a varying appearance ranging from superficial cutaneous purplish macules to freely movable subcutaneous masses to deep –seated fixed lesions. These lesions are slow growing, painless soft tissue swelling with a normal mucosa if not ulcerated and size ranges between 0.3-0.5cm<sup>[8]</sup>.

In this reporting a rare case of 82years male presented to Ramaiah hospital with swelling in the upper part of left thigh. MRI of the hip joint revealed to be nerve sheath tumor

### Case Report

82 year old male presented to our hospital with complaints of swelling in the left thigh for 2 years, which increased rapidly in size during last 5 months associated with paraesthesia of the left lower limb and dull aching pain in the left thigh. There was no evidence of claudication and acute ischemic disease. He had no co- morbidities. However a year ago, he underwent dynamic hip screw implanting for the left hip fracture. On examination (fig 1) the solitary oval swelling firm in consistency with no mobility measuring around 8x10cm near the femoral canal. Peripheral pulses well felt.

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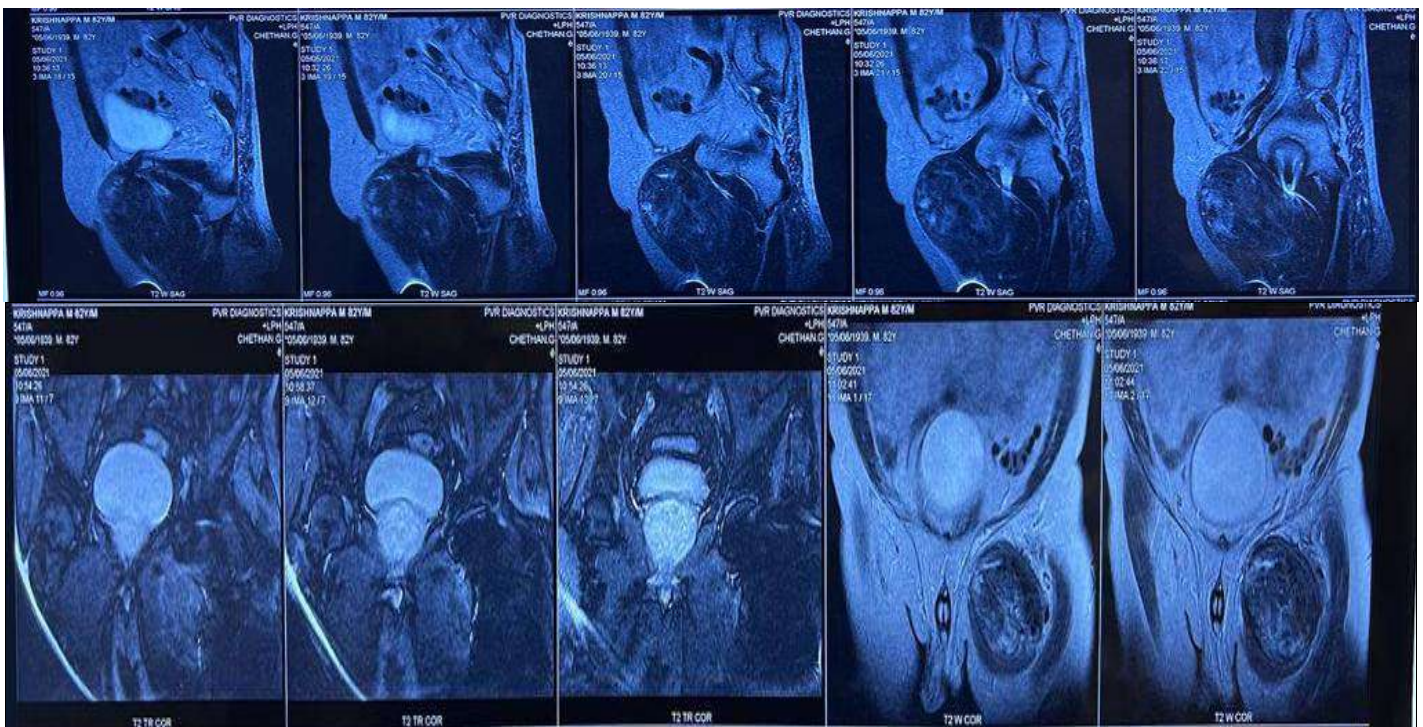


**Fig 1:** Clinical presentation – swelling in the left thigh

**Procedures and methodology**

Routine blood investigations were done. MRI of the HIP joint (fig-2) done which showed left femoral DHS screw and plate in

situ and well defined alerted signal intensity lesion in the medial aspect of left upper thigh, likely to be nerve sheath tumour ?Hematoma. TruCut biopsy revealed it to be spindle cell lesion.



**Fig 2:** A) MRI scan – showing the swelling in the left thigh, B) MRI scan – bilateral hip joint

Patient underwent wide local excision (fig 3) under spinal anaesthesia and specimen sent for HPE.

Intra-operative findings long saphenous vein was present at crossing over the swelling and femoral vessels were posteriorly.

These vessels were separated. Tumor excised (fig 4) measuring around 14 x 15cm. Intra operative and post-operative period was uneventful. Patient was discharged post-operative day3 with subcutaneous drains.





**Fig 3:** a) wide local excision- tumor identified b) Adhesions released c) tumor excised d) Skin closure and drain in situ



**Fig 4:** a) dissected specimen b) excised tumor

HPE examination showed well encapsulated neoplasm, composed of proliferation of bland spindle cells. These cells are arranged in sheets and fascicles. The nuclei are wavy, delicate and pointed ends. The stroma is densely eosinophilic, fibrotic and hyalinised. No mitotic activity or atypia or necrosis seen. Few entrapped adipose tissue present. Sections from the skin shows ulcerated areas. Impression '*benign spindle cell neoplasm*'

IHC Profile – Desmin – positive, S-100 protein: Negative, Smooth muscle Actin (SMA): Negative, Vimentin: Positive, H Caldesmon: Negative, Ki-67: <1% which favours of myofibroma.

#### Discussion and conclusion

Myofibroma (unifocal) and myofibromatosis (multifocal) are rare spindle cell neoplasm's composed of myofibroblasts. Lesions can be solitary (75%), multicentric with or without visceral involvement. These lesions can occur in any age group, most often in first decade of life; some of the authors reported

90% of case occur before the age of 2 [6, 7] and in adults most common is solitary lesions. These lesions clinically more common in head and neck followed by trunk and extremities. The most common sites are the skin, subcutaneous tissue, and skeletal muscle, with the oral lesions typically in mandible, lips, cheek and tongue [9]. Histologically lesion is mainly composed of spindle cell lesions.

In the case presented here of 82years male presented with swelling in the upper part of left thigh. MRI of the HIP joint revealed to be nerve sheath tumor? Hematoma. Trucut biopsy showed spindle cell lesion. After wide local excision of the lesion. HPE and showed benign spindle cell neoplasm and later IHC Profile – Desmin – positive, S-100 protein: Negative, Smooth muscle Actin (SMA): Negative, Vimentin: Positive, H Caldesmon: Negative, Ki-67: <1% reported as myofibroma.

Presence of the spindle cell lesion in thigh region was never reported as per the available literature.

In the case presented here, the lesion appeared clinically as a benign tumor with no clinical, radiological or histological

evidence of myofibroma and was treated by excision.

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