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Case Report

Orthopaedics Section

Deep Soft Tissue Leiomyoma of Forearm: A Case Report and Review of Literature

BABULREDDY BOMMIREDDY[1], VIJAY GURRAM[2]

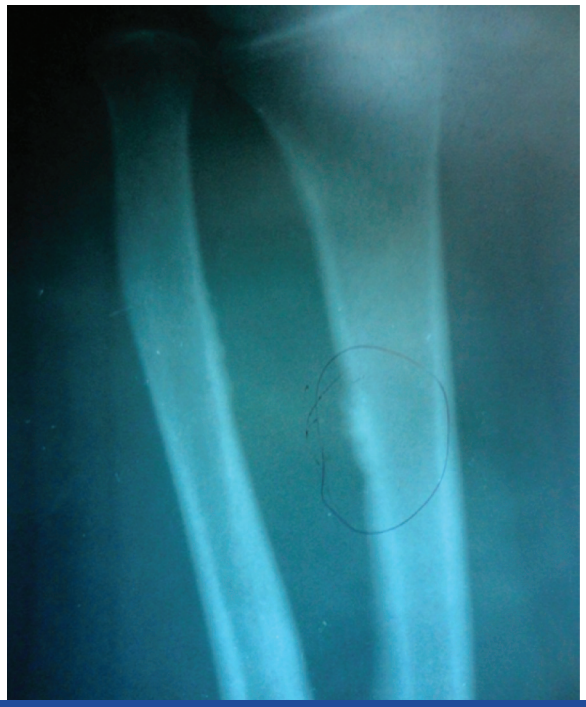
ABSTRACT

Leiomyomas are benign tumours of smooth-muscle origin representing 4.4% of all benign soft-tissue neoplasms. They are classified as cutaneous, vascular and leiomyomas of deep soft tissues. Leiomyomas rarely occur in extremities and are more common in the lower limb than in the upper extremity. Deep soft tissue leiomyomas are even rare with a very few reported cases so far in the literature. A 25-year-old female presented to us with an atraumatic slowly enlarging mass in the right forearm from 6 months with mild erosion of cortex of radius. She was otherwise healthy, MRI revealed a soft tissue lesion involving the interosseous space, isointense on T1, slightly hyperintense on T2 and hyperintense on STIR images. The tumour was excised intoto. The case is presented due to its rarity and the risk of tumor misdiagnosis. It should be considered in the differential diagnosis of any solitary painful slow growing mass of the extremities. If adequate margins are obtained recurrence of this tumour is very rare.

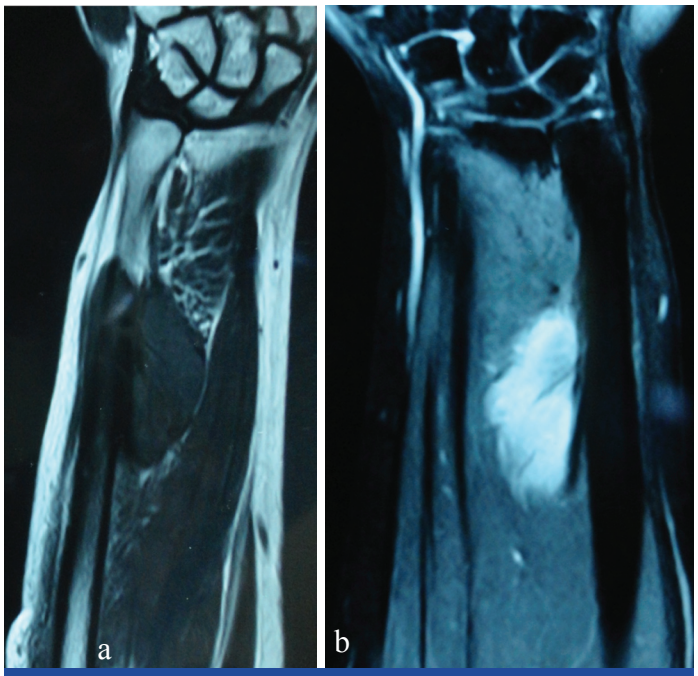
Keywords: Leiomyoma, Neoplasm, Tumour

CASE REPORT

A 25-year-old female presented to us with an atraumatic slowly enlarging mass in the right forearm from 6 months. She had occasional dull aching pain over the lesion. She was otherwise healthy. On examination there was a swelling in the distal third forearm with obscure margins, firm in consistency, non mobile and mildly tender. There was gross restriction of forearm rotation. Her haemoglobin was 12.2mg, Cell counts were normal and ESR was 12mm. Plain x-ray [Table/Fig-1] showed a soft-tissue shadow in the interosseous space causing mild erosion of the medial cortex of the radius. MRI revealed a soft tissue lesion in the distal forearm measuring 8×4×3cm involving interosseous membrane and extensor muscles of forearm. It was isointense on T1, slightly hyperintense on T2 and hyperintense on STIR images [Table/Fig-2a,b]. MRI reported a differential diagnosis of fibromatosis, fibrous histiocytoma and sarcoma. The tumour was excised completely through dorsal approach [Table/Fig-3] and sent for histopathology.



[Table/Fig-1]: Radiograph showing erosion of cortex of radius in antero-posterior view.

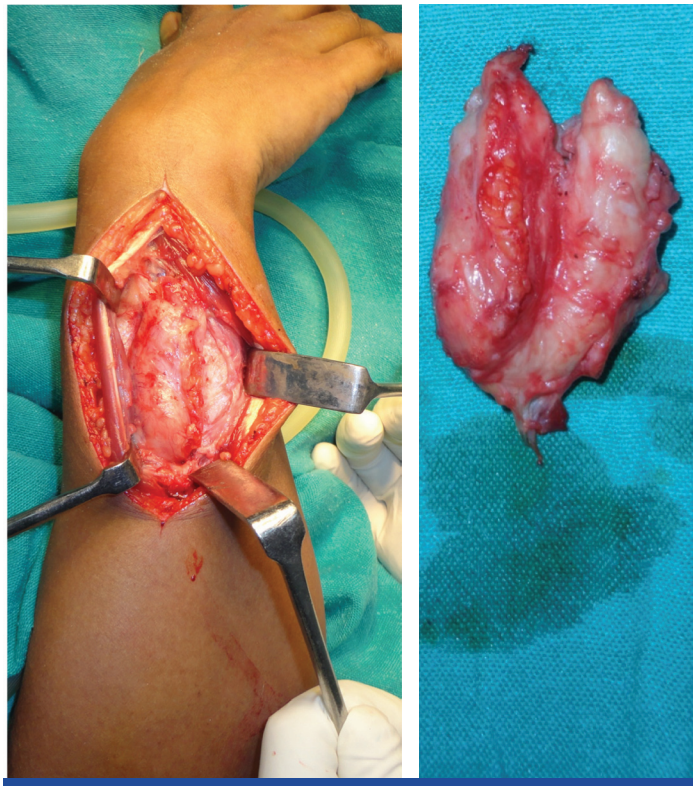


[Table/Fig-2]: MRI images showing the tumour in the interosseous space which is isointense on T1 (2a), slightly hyperintense on T2 (2b) images.

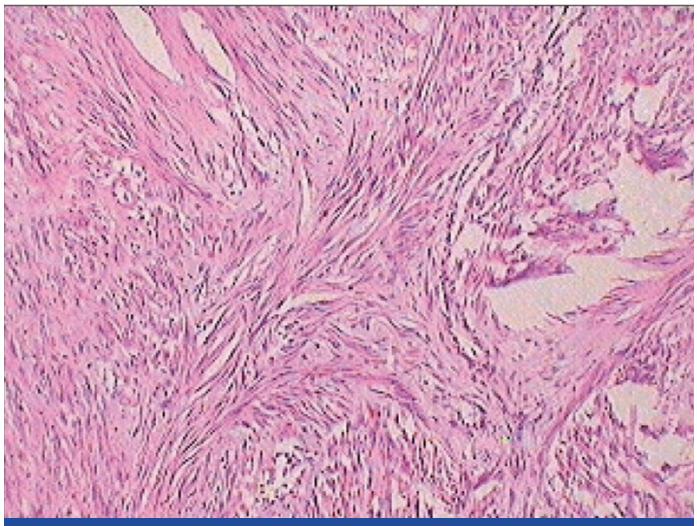
Intraoperatively it was well encapsulated, firm in consistency, involving the interosseous membrane, causing mild erosion of medial cortex of radius. Initial microscopic examination [Table/Fig-4] revealed a well circumscribed lesion showing interlacing bundles of smooth muscle cells with elongated nuclei and moderate amount of eosinophilic cytoplasm with focal areas of hyalinisation. There was no evidence of mitosis or nuclear atypia. Immunohistochemistry showed a strong reactivity against Smooth Muscle Actin (SMA) and a negative reaction against S-100 and epithelial membrane antigen (EMA) consistent with leiomyoma. She was asymptomatic at the follow-up of 2 years without any recurrence. Informed consent and ethical clearance was taken for this study.

DISCUSSION

Leiomyomas are benign tumours of smooth-muscle origin representing 4.4% of all benign soft-tissue neoplasms [1]. They



[Table/Fig-3]: Intraoperative pictures showing tumour mass.



[Table/Fig-4]: Histopathology showing interlacing bundles of smooth muscle cells with elongated nuclei and moderate amount of eosinophilic cytoplasm with focal areas of hyalinisation without nuclear atypia.

are classified as cutaneous, vascular and leiomyomas of deep soft tissues. They most commonly occur in the third and fourth decades of life [2]. These lesions are twice as common in women as in men. Leiomyomas of the uterus are the most common tumours in women [3]. Leiomyomas rarely occur in extremities and are more common in the lower limb than in the upper extremity [4]. Leiomyomas of the upper extremity are extremely rare and arise from non-striated muscles in the upper extremity, such as erector pili, sweat glands and vascular walls [5]. Deep soft tissue leiomyomas are even rare with a very few reported cases so far in the literature.

Leiomyomas, first described by Virchow in 1854, are tumours of smooth-muscle origin representing 4.4% of all benign soft-tissue neoplasms [1,6]. The hereditary form, which causes, multiple leiomyomas, was originally noted by Kloepfer et al., in 1958 [7]. They are classified as: a) Cutaneous, arising from the erector pili muscle; b) Vascular leiomyoma, arising from smooth muscle of the vein; and c) Leiomyomas of deep soft tissues [2]. Solitary cutaneous and subcutaneous leiomyomas are independent of age and that there is no predisposition by race or sex [8]. Vascular and deep soft tissue leiomyomas are more common in second and third decade of life with a female preponderance [8]. Leiomyomas of the extremity are divided into superficial and deep tumours [9]. Very few cases of leiomyoma involving the appendicular skeleton have been reported. When they occur in the extremities, they are more common in the lower limb than in the upper limb [9]. Deep soft-tissue leiomyomas can be vascular and nonvascular. Deep nonvascular soft-tissue leiomyomas are extremely rare and there are very few cases reported so far in the literature. Misumi et al., reviewed 21 cases of deep soft tissue leiomyoma from the English literature and they found to occur at almost any age, ranging from 3 to 62 years (mean, 25 years) and more frequently affected males (14 cases) than females (7 cases) [10]. Almost half of the cases were located in the extremities (10 cases), and there was only one report with multiple deep soft tissue leiomyomas [10]. Ramachandran et al., reported a case of deep soft-tissue leiomyoma of the forearm mimicking a primary bone tumour of the ulna [11]. The pathogenesis of leiomyomas remains obscure. According to Goodman et al., deep leiomyomas arise from undifferentiated mesenchymal cells or smooth-muscle rests [12]. Stout et al., suggest that these tumours may instead arise from the smooth muscle in the walls of blood vessels [8]. These tumours may not be easily recognised until become painful and it is rarely diagnosed before surgery as imaging techniques, including MRI, are not specific for diagnosis [10]. Pain in these cases may be because of contraction of smooth muscles or due to compression of cutaneous nerves because of tumour and these tumours are often found on extensor surfaces and grow slowly [8]. In addition, most patients describe the pain as “sharp” or “stabbing”, while some describe it as only localized tenderness [9]. The present report also supports this anatomic localization and characteristic of the pain. Occasionally, nerve fibres are revealed on slides of leiomyoma, histologically. Leiomyomas can usually increase in size to larger proportions, particularly in the deeper soft tissue compartments. A leiomyoma of the upper extremity sized 30×29×12cm is reported by Drew et al., [13].

Scattered calcifications have been reported in isolated deep soft-tissue leiomyomas, which may lead to their being mistaken for myositis ossificans [12]. However, in our case, there was no calcification. The imaging features are nonspecific and similar to those of many other soft-tissue neoplasms. The differential diagnosis includes lipoma, leiomyosarcoma, schwannoma or neurofibroma, haemangioma and soft-tissue giant-cell tumour of the tendon sheath [11].

Operative excision is the treatment of choice in these cases for both pathologic diagnosis and for definite treatment. Billings et al., reported neither recurrences nor metastases after excision of somatic soft tissue leiomyomas during a mean follow-up of 58.7 months [14]. However, in other studies is described that malignant formation of a finger and forearm leiomyoma respectively which required further surgical intervention [15,16]. If an adequate margin is obtained, recurrence of a leiomyoma is rare. Despite the rarity of the above phenomenon, these tumours should be approached with caution until histopathologic examination confirms the absence of nuclear atypia, necrosis and mitotic activity. There was no recurrence in this patient 2 years after the operation.

CONCLUSION

The case was presented due to its rarity and the risk of tumour misdiagnosis. Deep avascular soft tissue leiomyoma is uncommon and its appearance in forearm constitutes a rare issue. It should be considered in the differential diagnosis of any solitary painful slow growing mass of the extremities. If adequate margins are obtained recurrence of this tumour is very rare.

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PARTICULARS OF CONTRIBUTORS:
1. Department of Orthopaedics, Employees State Insurance (E.S.I) Medical College, Hyderabad, India.
2. Department of Orthopaedics, Employees State Insurance (E.S.I) Medical College, Hyderabad, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Babulreddy Bommireddy,
12-1-508/B/79,Sripuri Colony, Hyderabad, Telangana-500040, India.
E-mail:babulortho@gmail.com

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