



Vascular leiomyosarcoma of thigh – A rare tumour at an unusual site

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CASE REPORT

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Abstract

Leiomyosarcomas of vascular origin are rare. They originate from the smooth muscles of tunica media of major blood vessels. The majority of such tumours arising in the extremities affect the femoral vascular bundle. There is limited knowledge and experience of the clinical presentation, pathological reports and results of treatment of this type of tumour.

A case of primary leiomyosarcoma of femoral vein is being reported from a subtropical region of India that developed over the right thigh of a 35-year-old male farmer and was clinically diagnosed as benign soft tissue tumour. The diagnosis was confirmed by histopathology and immunohistochemistry.

Key Words

Femoral vein, leiomyosarcoma, tumour, vascular

Implications for Practice:

1. What is known about this subject?

Leiomyosarcomas of vascular origin comprise a seemingly rare group of tumours, with only a few hundred cases reported in literature since the initial report by Perl in 1871. To our knowledge this is the first case of primary leiomyosarcoma of femoral vein being reported from India.

2. What is the key finding in this case report?

Our case highlights several noteworthy features. Firstly, the case is unusual with respect to its anatomical location which may be misleading for the clinicians at times. Leiomyosarcomas usually arise in internal organs composed

of smooth muscles viz. uterus and gastrointestinal tract. Thigh is an unusual site for leiomyosarcoma to occur. Such tumours may pose a diagnostic challenge when they occur at unusual sites. Secondly, the clinical presentation of this tumour in this case was such that it was misinterpreted by clinicians as a benign soft tissue mass and under that impression only a simple excision of the tumour was performed.

3. What are the implications for future practice?

Our report emphasises the need for awareness about this condition as well as effective communication between clinicians and pathologists. In cases of any clinical doubt, a pre-operative fine needle aspiration cytology (FNAC) will help clinicians to ascertain the nature of the lesion and accordingly plan an appropriate treatment strategy. Since this tumour presented with an extraluminal extension, an FNAC prior to surgery would have changed the treatment protocol. Hence, the importance of FNAC in pre-operative diagnosis of any visible mass has to be stressed upon.

Background

Leiomyosarcomas are malignant tumours of smooth muscle origin and account for five to six per cent of all sarcomas.¹ They are common in the uterus and gastrointestinal tract.¹ However, a small percentage of this sarcoma subset may originate from the smooth muscle of the vessel walls and are usually of venous origin. Although vena cava has been reported in the literature as the common source for these tumours, there is limited information focussed on the non-venacaval leiomyosarcomas which arise from larger veins.² Leiomyosarcomas arising from the vessel walls are also known as vascular leiomyosarcomas and represent only two per cent of all leiomyosarcomas.^{3,4}

We present a case of primary leiomyosarcoma of the femoral vein with extraluminal extension in a 35-year-old male. The case is being presented because of its unusual clinical presentation and anatomic location. The focus of this case study was also to highlight the diagnostic errors which might occur during pre-operative evaluation of such cases.

Case details

Presentation

A 35-year-old male presented with a painless swelling on the upper part of the right thigh. The swelling had been growing steadily for the last three years. The patient did not have any systemic complaints nor did he experience any significant weight loss. Physical examination showed a firm, well-defined, non-tender, partially mobile, (7 x 4) cm mass on the upper part of the right thigh with no overlying skin changes. Clinically, it was diagnosed as intramuscular lipoma. However, Magnetic Resonance Imaging (MRI) showed the mass to be attached to the femoral vein. A simple tumour excision was done under the impression of a benign soft tissue tumour and the excised mass sent for histopathological examination.

Figure 1: Gross appearance of the tumour showing a polypoidal mass



Figure 2: Photomicrograph showing long interlacing fascicles of spindle cells (H and E, 400x)

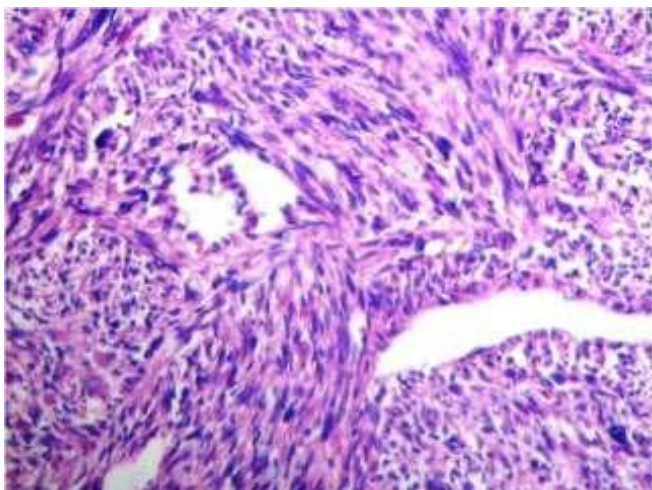


Figure 3: Photomicrograph showing tumour giant cells and mitotic figures (H and E, 400x)

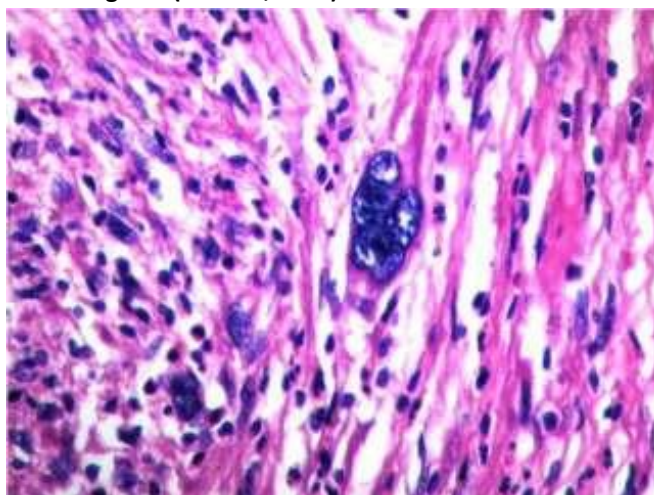


Figure 4: Photomicrograph showing haemangiopericytoma like architecture (H and E, 100x)

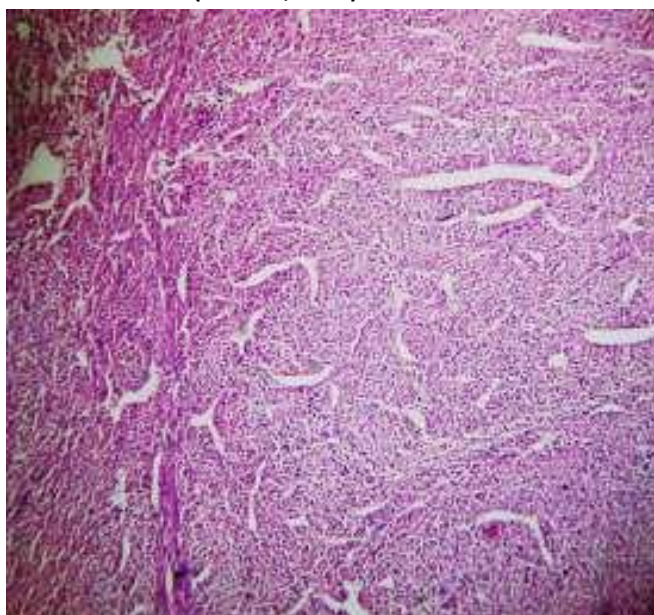
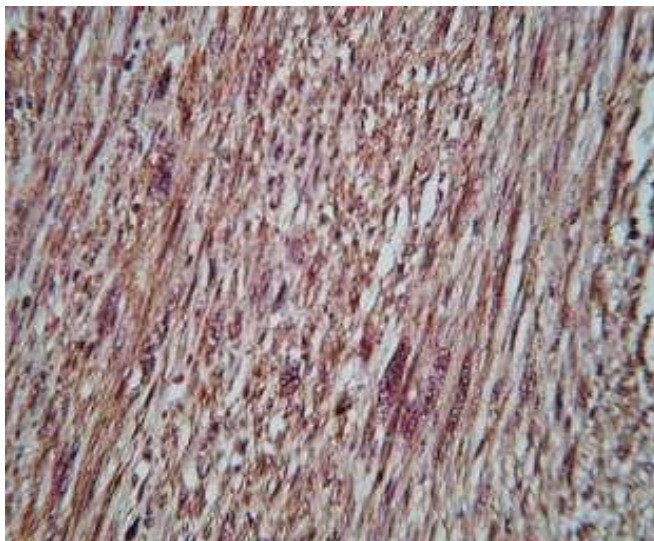


Figure 5: Photomicrograph showing smooth muscle actin positivity in the neoplastic spindle cells (400x)





Pathological features

Gross examination of the specimen revealed a globular polypoidal mass measuring (7x5x2) cm. The mass was well encapsulated, firm in consistency, brownish in colour with a lobulated outer surface (Figure 1). A cut section showed greyish white mass with whorled appearance and slit like spaces at places. No area of haemorrhage or necrosis was noted.

Microscopy showed long interlacing fascicles of spindle cells having hyperchromatic, moderately pleomorphic nuclei with rounded ends (cigar shaped) and bright eosinophilic cytoplasm (Figure 2). Huge areas of necrosis were noted. Numerous typical and atypical mitotic figures (15-20 per 10 high power fields) along with a few scattered tumour giant cells were also seen (Figure 3). The overall architecture had a haemangiopericytoma like pattern (Figure 4). The neoplastic spindle cells were positive for Smooth Muscle Actin (SMA) (Figure 5) and vimentin but were negative for cytokeratin, MyoD1 and S-100. The final diagnosis based on MRI, histomorphology and immunohistochemistry was Grade 3 leiomyosarcoma of the femoral vein.

Discussion:

Leiomyosarcomas of vascular origin comprise a seemingly rare group of tumours, illustrated by the fact that only a few hundred cases have been reported in the literature since the initial report by Perl in 1871.^{1,3} Hallock et al noted only one case in 34,000 autopsies from the University of Minnesota, Abell reported two in 14,000 autopsies at the University of Pennsylvania;¹ and Dorfman and Fisher found none in 30,000 autopsies at the Johns Hopkins Hospital.¹ Yet it should be emphasised that several features of this disease probably significantly affect its detection, diagnosis, and incidence. Hashimoto et al⁵ documented that at least one-quarter of leiomyosarcomas of peripheral soft tissue in their experience arose from or involved a vessel; while Goldblum et al⁶ observed this in at least one-third of cases. Thus, the recorded experience with vascular leiomyosarcomas is a biased one, which probably underestimates the true incidence and possibly also conveys a false impression concerning clinical behaviour. Literature regarding such presentations is rare and is mostly in the form of case reports. To our knowledge, fewer than 30 cases of leiomyosarcomas originating from the femoral vein have been reported in the English literature.² After a thorough literature search, we found that cases of venacaval leiomyosarcomas have been reported from India⁷⁻⁹ but our case happens to be the first primary leiomyosarcoma of femoral vein being reported from India.

Clinical presentation depends on whether the tumour is intraluminal or extraluminal. Intraluminal femoral vein involvement, when occlusive, can mimic the symptoms of deep vein thrombosis.² Extraluminal expansion from the media musculature into the surrounding perivascular tissue may compress the artery or nerve, usually resulting in poorly defined pain.^{2,10} Mixed forms of intra- and extraluminal development have been reported with various combined symptoms.² As shown by MRI, the tumour in our case was extraluminal and had no intraluminal component.

The diagnosis is based on radiological and pathological features; gold standard being histopathology supplemented by immunohistochemistry for smooth muscle actin (SMA). A pre-operative FNAC may, however, help in proper clinical evaluation and planning of treatment.

Differential diagnoses of a malignant soft tissue mass in the thigh include fibrosarcoma, monophasic synovial sarcoma, malignant peripheral nerve sheath tumour, spindle cell rhabdomyosarcoma, haemangiopericytoma and malignant fibrous histiocytoma.¹ Fibrosarcoma with its typical herring bone pattern is a diagnosis of exclusion and is not positive for SMA. The typical histomorphology of the present tumour along with its negativity for cytokeratin, S-100, MyoD1 and CD34 as shown by immunohistochemistry, excluded the possibility of monophasic synovial sarcoma, MPNST, spindle cell rhabdomyosarcoma and haemangiopericytoma respectively. The histology of the tumour did not support the diagnosis of malignant fibrous histiocytoma either because of the lack of a conspicuous inflammatory infiltrate.

Management of venous leiomyosarcomas of the lower limb is difficult to evaluate because the incidence is low and most reports have been single cases. Wide surgical excision seems to be the treatment of choice.¹¹ Neoadjuvant chemotherapy may be given to downsize the tumour and increase the resectability rates. Postoperative adjuvant radiation therapy is recommended.¹¹

Leiomyosarcomas of vascular origin have a relatively poorer prognosis.¹ The poorer prognosis of venous leiomyosarcomas compared to those of soft tissue could be because these tumours generally have direct access to the venous system, involving the lumina, and tend to have early blood-borne metastases.² It has been suggested that the very presence of smooth muscle differentiation in a pleomorphic spindle cell tumour is an independent indicator for poor prognosis.¹ Therefore, it is necessary that an accurate diagnosis be made and the clinicians alerted, whenever such cases are encountered. All the cases



reported by Berlin et al. had metastases, with five cases dying from metastatic disease within five years.¹⁰ However, it was not so in our case. A PET scan, done in a higher referral centre, ruled out the presence of metastasis in this case. The patient was advised post-operative radiotherapy, but was lost to follow up.

In conclusion, it is to be borne in mind that leiomyosarcoma of the femoral vein is an uncommon condition diagnosed by imaging, histopathology and immunohistochemistry. Greater awareness and knowledge about this condition can lead to prompt diagnosis and effective management of the disease.

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PEER REVIEW

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CONFLICTS OF INTEREST

The authors declare that they have no competing interests.

PATIENT CONSENT

The authors, Deb Roy A, Deka M and Dutta UC declare that:

1. They have obtained written, informed consent for the publication of the details relating to the patient(s) in this report.
2. All possible steps have been taken to safeguard the identity of the patient(s).
3. This submission is compliant with the requirements of local research ethics committees.