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Case Report

# Imaging in a rare case of intramuscular angioleiomyoma around the knee joint

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## Abstract

Angioleiomyoma are rare benign tumors originating from smooth muscles of veins. They are found more commonly in extremities and are seen in subcutaneous tissue. Intramuscular angioleiomyoma is rare and can be confused with hemangioma. Though they do not have any characteristic imaging features but they should be considered in the differential diagnosis due to certain specific imaging findings on MRI which are discussed in this article.

**Key words:** Angioleiomyoma; USG, CT, MRI, hemangioma

## Introduction

Angioleiomyoma are rare benign tumors originating from smooth muscles of tunica media of veins.[1] Patients generally present with a well-defined, soft tissue swelling, however, pain with or without tenderness can be experienced in approximately 60% of cases.[2] Although these tumors can occur anywhere in the body, they are seen more often in the extremities, particularly in the in lower limbs, more than in any other part of the body. These lesions are generally located superficially in the subcutaneous fat, fascia or dermis and are easily palpable on clinical examination. Intramuscular angioleiomyoma are uncommon and have been rarely reported in the literature. We hereby present a case of intramuscular angioleiomyoma occurring in vastus medialis muscle around the knee joint. Authors need to cite references here for all information quoted. We need a revised version from the Authors, incorporating the references in the introduction section.

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## Case Report

A 36-year-old male patient presented with an insidious onset, soft tissue swelling , over the medial aspect of the right knee, which had been slowly increasing in size over the last 8 years. He also complained of chronic intermittent pain at that site during movement. There was no history of trauma. On examination, the overlying skin was normal. On palpation the lesion was firm in consistency, fixed to underlying structures and was inseparable from adjacent muscle. The lesion became prominent on contraction of the muscles. Muscle strength was normal with no sensory or motor deficit. Ultrasound revealed a well-defined heterogeneously hyperechoic oval lesion located in medial aspect of the knee, within the vastus medialis muscle. The lesion had a peripheral rim of hypo echogenicity [Figure 1A]. There was no calcification. The lesion showed



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mild central vascularity on color Doppler [Figure 1B]. A suspicion of intramuscular hemangioma was raised and patient subjected to triple-phase CT to confirm the ultrasound findings. CT showed a well-defined heterogeneous density mass with foci of interspersed fat densities measuring 3.8 × 2.4 × 2.7 cm (AP × TR × CC) in the vastus medialis muscle on medial aspect of the right knee. The lesion did not show any parenchymal enhancement in the portal phase but showed progressive parenchymal enhancement in the portal and equilibrium phase with type I time density curve [Figure 2]. In view of progressive enhancement, diagnosis of intramuscular hemangioma was considered. MRI was requisitioned by the treating surgeon for preoperative mapping. MRI revealed a well-defined round to oval lesion in vastus medialis muscle , The lesion was isointense on T1WI with interspersed hyperintensities, heterogeneously hyperintense on T2 , with a hypointense rim. The lesion was hypo- intense on STIR (short tau inversion recovery) [Figure 3A-C]. The interspersed hyperintensities on T1WI showed fat suppression on T2W FS images [Figure 3D]. Post-contrast the lesion showed avid heterogeneous enhancement [Figure 3E and F]. Apart from hemangioma, a differential diagnosis of nerve sheath tumor was suggested based on MRI appearances. The patient was taken up for planned surgery, peroperatively the lesion was excised from vastus medialis muscle and sent for histopathological examination (HPE). HPE showed a well-circumscribed lesion, arranged in fascicles. The individual cells were spindle-shaped and did not show atypia. The cells were immunopositive for SMA and desmin [Figure 4]. The tumor was diagnosed as an angioleiomyoma arising in vastus medialis muscle.

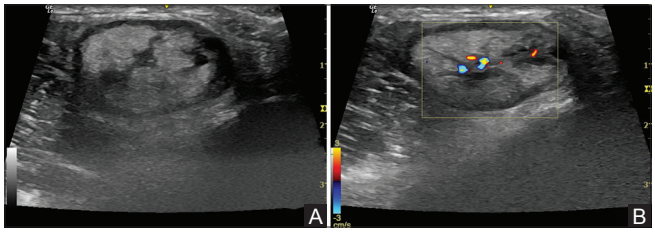


**Figure 1 (A and B):** Ultrasound showing well defined hyperechoic oval lesion with peripheral and central hypoechogenicity (A). On color Doppler, the lesion shows central vascularity (B)

## Discussion

Angioleiomyomas are rare benign tumors originating from the smooth muscles of tunica media of veins,[1] these tumors occur more commonly in middle-aged women and have a predilection for the lower extremities.[2] The lesion starts

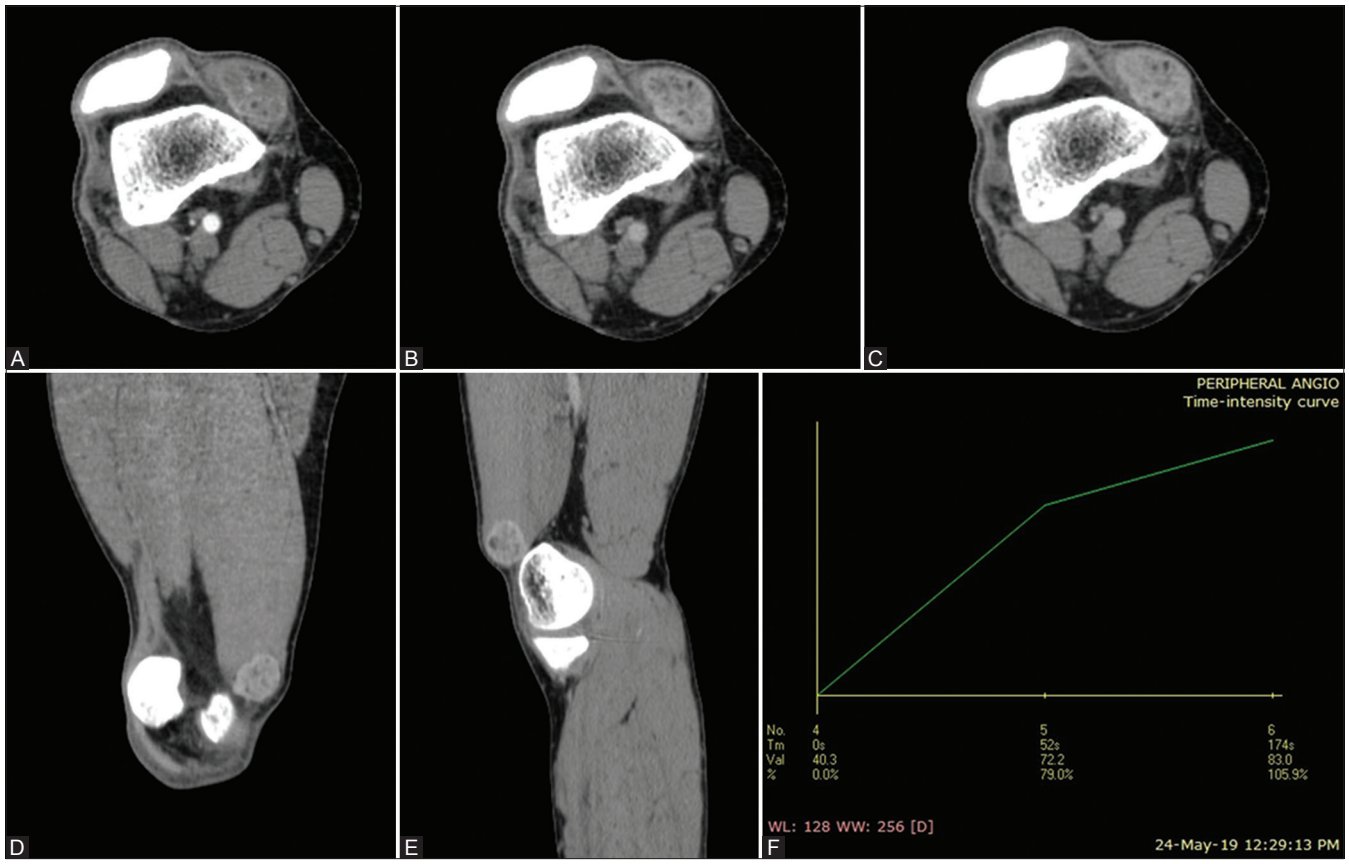


**Figure 2 (A-F):** Triple phase axial CT images showing minimal central vessel enhancement in the arterial phase (A) with increasing parenchymal enhancement in the portal (B) and equilibrium phase (C) technical correction by Authors is required. The lesion shows type I enhancement curve on time density graph (F). Coronal and Sagittal images (D and E) showing the lesion in vastus medialis muscle

off as a slow-growing nodule which is firm and mobile in subcutaneous location. Pain is a presenting feature in majority of patients as was seen in our case and has been reported in various studies. Hachisugo *et al.*[2] reported pain in 58% of cases whereas Freedman *et al.* reported pain in 62% cases.[3]

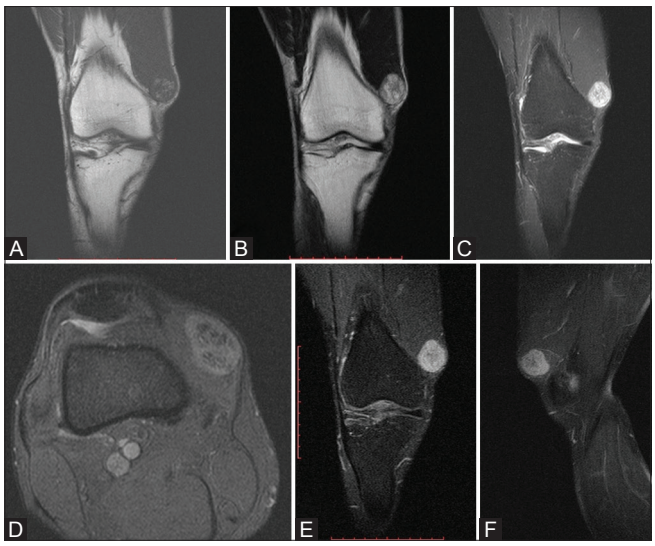


**Figure 3 (A-F):** MRI Coronal (A-C) T1W, T2W and STIR images showing well defined rounded lesion in vastus medialis muscle which is isointense on T1W with intralesional fat intensity (A), hyperintense on T2W and STIR (B and C) with T2 hypointense peripheral rim. The lesion shows fat suppression on the T2WFS image (D). Post-contrast coronal and sag (E and F) images showing avid heterogeneous enhancement

Angioleiomyomas are superficial tumors found in subcutaneous plane and are rare in muscular compartment unlike the present case which was seen in vastus medialis muscle of the thigh around the knee joint. Imaging studies are performed to narrow the differential diagnosis and exclude malignant lesions. The ultrasound features described in literature are round-to-oval lesions with well-defined margins. They are reported to be homogeneously hypoechoic and characteristically reveal moderate vascularity on color Doppler imaging.[4] As per WHO criteria, angioleiomyomas have been divided into three subtypes namely capillary (or solid), venous, and cavernous type lesions. Ultrasound features vary as per the subtypes, the capillary type lesions are more solid and echogenic than other two subtypes which are homogeneously hypoechoic. The lesion in our patient was hyperechoic with peripheral and central hypoechogenicity which favors the capillary subtype [Figure 1]. The capillary type has mild vascularity whereas venous and cavernous type have moderate-to-rich vascularity. The lesion in our case showed mild central vascularity. Calcification, which is an uncommon ultrasonographic finding in angioleiomyomas was not seen in our case.

Nevertheless, CT findings have not been described in the literature. The lesion in our case was of heterogeneous density (45-52HU) with intralesional fat densities. The lesion showed progressive enhancement after contrast injection. In the arterial phase, feeder vessels were seen centrally

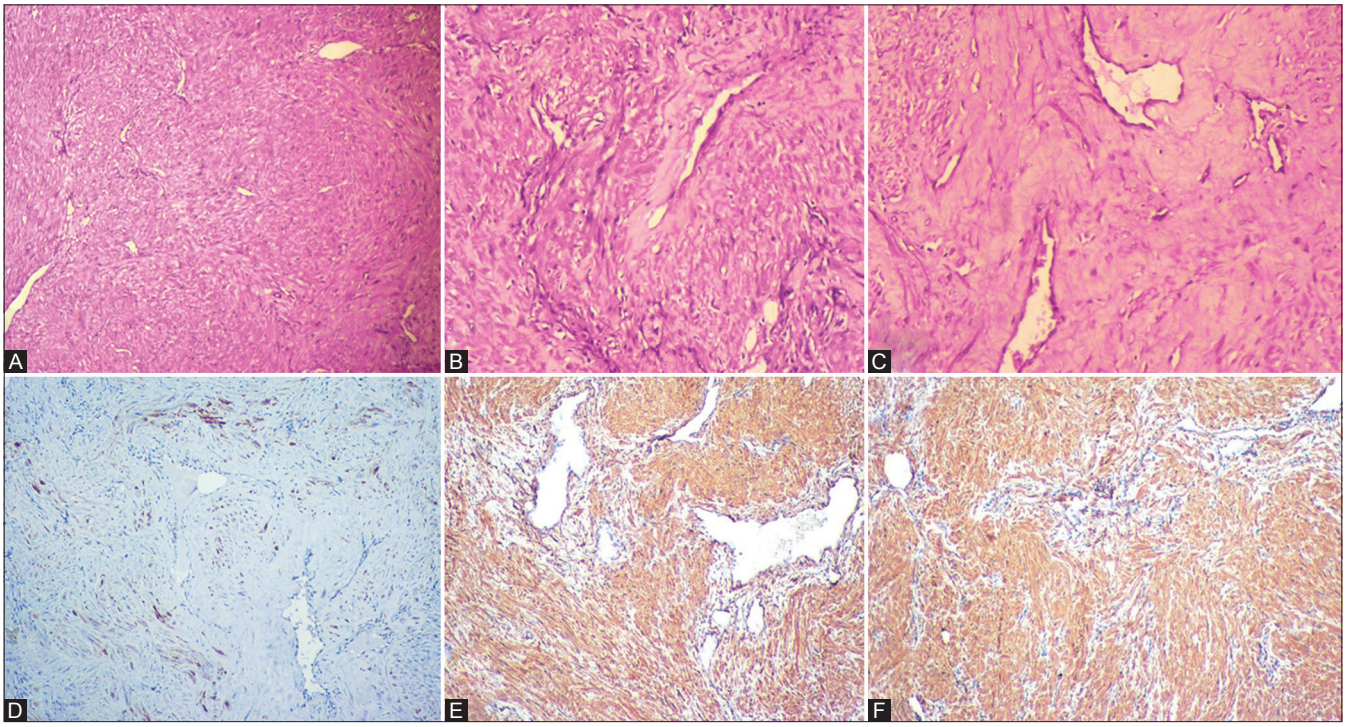


**Figure 4 (A-F):** HPE (400 × and 1000×). Photomicrograph shows a spindle cell tumor around thick-walled blood vessels (A). Spindle cells showing uniformity with no atypia or mitosis (B) and areas of hyalinization (C). IHC for CD34 was negative (D). IHC for SMA and desmin (cytoplasm) was positive (E and F)

but there was no peripheral nodular or parenchymal enhancement. In the portal and equilibrium phase, the lesion showed progressive parenchymal enhancement [Figure 2]. This progressive enhancement pattern is typically seen in haemagiomas which also typically show peripheral nodular enhancement in arterial phase which was not seen in this case. Lack of peripheral nodular enhancement can be considered to be a feature more likely to be seen in angioleiomyoma than hemangioma. Intralesional fat is also more likely to be seen in angioleiomyoma than hemangioma. However, more CT studies will be required to establish these findings as peculiar to angioleiomyomas. CT is very sensitive for detecting calcification but there was no calcification seen in our case. Hemangiomas may show presence of phleboliths within. The near similar enhancement pattern of these two lesions can be explained by the hypothesis proposed by Duhig and Ayer,[5] who state that "the proliferation of smooth muscle in a hemangioma produces an angioleiomyoma and further proliferation produces a simple leiomyoma". However, other workers have suggested that it is a hamartomatous lesion arising from the wall of the vein.[2]

MRI findings in angioleiomyoma have been described by many researchers, Gupte *et al*.[6] have described MRI findings in ten cases of angioleiomyoma which has been claimed as the largest series describing MRI findings. They found the lesions to be isointense to muscle on T1WI and heterogeneously hyperintense on T2W/STIR images. They found central fat intensity in a single case. Yoo *et al*.[1] in their MRI study of eight cases found all the lesions to be isointense on T1W and hyperintense on T2W. Our case also showed similar appearance on T1W, T2W, and STIR images except for intralesional fat which was not frequent finding in these two studies [Figure 3]. The presence of aT2W hypointense rim has been reported by Yoo *et al*. in all the eight cases they studied was observed in our case as well. They found that all the lesions showed good homogeneous enhancement except one, which showed poor enhancement. Avid heterogeneous enhancement was seen in our case. All the studies have described angioleiomyoma as subcutaneous lesions unlike our case which was intramuscular but located superficially. MRI features which should raise the suspicion of angioleiomyoma can be described as isointense lesion on T1W and hyperintense on T2/STIR with T2 hypointense rim and avid homogeneous or heterogeneous contrast enhancement. Intralesional fat densities may or may not be present.

Imaging differential diagnosis of angioleiomyoma should include giant cell tumor of the tendon sheath, nerve sheath tumors, and hemangiomas depending on the location of the lesion. Giant cell tumors of tendon sheath show low signal intensity on T1 and T2WI and show blooming on GRE images. Nerve sheath tumors are hypointense on T1 and hyperintense on T2WI and may show split fat sign, target sign or fascicular sign on MRI. Hemangiomas closely resemble angioleiomyoma on imaging in all modalities. The signal intensities are similar in both the lesions, the phlebolith if seen will favor the diagnosis of hemangioma. T2 hypointense rim as described by Yoo *et al*. and as seen in this case will favor the diagnosis of angioleiomyoma. Post-contrast the enhancement pattern in both these lesions is quite similar and may not be of much help in differentiating the two.

## Conclusion

Angioleiomyoma is a soft tissue tumor found mostly in the subcutaneous plane but can also be found in muscles located superficially. They present with local pain in majority of the cases. They have to be differentiated from other common subcutaneous swellings. Imaging can help narrow down the differential diagnosis in these cases. The diagnosis of angioleiomyoma should be suspected if the lesion is superficial and isointense on T1 and hyperintense on T2/STIR, has a T2 hypointense rim and shows avid enhancement on post-contrast imaging.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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### Conflicts of interest

There are no conflicts of interest.

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