CASE REPORT

Angiomyoma presenting as a painful subcutaneous mass: a diagnostic challenge

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SUMMARY

Angiomyoma of the extremity is a notoriously elusive preoperative diagnosis, as the list of differentials for its described classic clinical features of a painful mobile subcutaneous mass is quite vast. Imaging features described for angiomyomas are far from being specific. On ultrasound, angiomyomas are mostly described as a well-defined solid mass lesion showing robust internal vascularity. On T2-weighted MRI they have been described as homogenous to heterogeneously hyperintense relative to skeletal muscle. We report a pathologically proven angiomyoma around the knee joint in a middle aged man, describe its clinical and imaging features, and outlay an approach in diagnosing this rare entity as a differential for painful subcutaneous mass lesions.

BACKGROUND

An angiomyoma is a rare benign smooth muscle tumour originating from the tunica media of the vessel walls.^{1 2} As the histological origin suggests, it can occur in any part of the body with the favoured site being the extremities, particularly lower limbs in females and upper limbs in males. Pain associated with a slowly growing subcutaneous mass is the most common presentation of angiomyomas.¹⁻³

Angiomyomas have rarely ever been correctly diagnosed clinically or radiologically, first because of their relative rarity of occurrence and, more importantly, due to the paucity of relevant radiological literature. The purpose of this case report is to highlight the imaging findings of angiomyoma and to sensitise clinicians, particularly radiologists, to consider angiomyoma as an important differential in the approach to a painful subcutaneous soft tissue mass.

CASE PRESENTATION

A 36-year-old man presented with a slow-growing, mildly painful mass around the left knee joint for the past 2 years. There was no history of trauma or difficulty in walking. On examination, a 3×2 cm soft, fluctuant mass was seen on the medial side of the knee along the patellar tendon. The mass lesion was mobile from the underlying bone and tendon with 'slip sign' positive. There was no ulceration or discolouration of the overlying skin and the skin could be easily pinched from the lesion. Temperature and touch sensation of the overlying skin was normal. Thus, a clinical diagnosis of ganglion cyst was made and the patient was referred for an ultrasound examination.

INVESTIGATIONS

Ultrasound revealed a well-defined, oval, homogeneously hypoechoic solid mass lesion in the subcutaneous tissue of the medial aspect of knee joint, showing rich internal vascularity on colour Doppler. Subsequently, MRI of the knee was performed revealing a well-defined, smooth and marginated, oval subcutaneous soft tissue mass measuring 3×2.5 cm on the medial aspect of the knee anterior to the patellar tendon. The lesion was hypointense to muscle on T1-weighted images and hyperintense on T2-weighted and showed thin interlacing internal septa with a thin peripheral hypointense rim on T2-weighted images (figure 1A, B). There was no evidence of any haemorrhage or calcification seen. Postcontrast, there was intense homogenous enhancement and a small vessel was seen leading up to and ending near the superior border of the lesion (figure 2A, B). There was no evidence of any definite nerve leading into the mass lesion. Owing to the vivid enhancement of the mass lesion we could not entirely rule out a malignant possibility, so the mass lesion was categorised as an indeterminate mass and biopsy was suggested.

Routine laboratory parameters (haemogram, blood counts, renal function tests and serum electrolytes) were unremarkable.

DIFFERENTIAL DIAGNOSIS

The differentials for a painful subcutaneous soft tissue mass of an extremity is quite vast and includes benign conditions such as neuroma, eccrine spiradenoma, angioliipoma, glomus tumours and malignant conditions such as synovial sarcoma.

TREATMENT

After proper preoperative evaluation the patient underwent excisional biopsy, whereas the mass was easily resected from surrounding subcutaneous tissue.

OUTCOME AND FOLLOW-UP

The postoperative specimen consisted of a single nodular, glistening grey white tissue piece measuring 3×2.5×1 cm. The cut section showed a well-circumscribed grey brown to grey white area measuring 2×1.5 cm, surrounded by yellow fatty areas. Histopathology revealed an encapsulated, well-circumscribed tumour composed of ectatic thick-walled vessels with circumferentially arranged smooth muscle cells, splaying of peripheral muscle fibres and areas of myxoid changes; features consistent with angiomyoma (figure 3A, B). Postsurgery, the patient had an uneventful recovery with regular dressing and antibiotics.

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Figure 1 MRI. T1-weighted sagittal images (A) revealing a well-defined hypointense subcutaneous mass lesion (arrow) anterior to the patellar tendon. On T2-weighted sagittal images (B) the lesion is heterogeneously hyperintense with a thin hypointense capsule (white arrow). Note is made of thin interlacing septae within the mass (black arrow).

DISCUSSION

Angiomyoma, also known as vascular leiomyoma or angioleiomyoma, is a benign tumour of smooth muscle origin arising from the muscular wall of veins and comprising less than 5% of all benign smooth muscle tumours.^{1–4} Peak incidence is seen in the fourth to sixth decades with the site of predilection being the subcutaneous tissue in the lower limbs, followed by upper limbs. Rare sites such as the labia majora, nipple, hard palate, pinna and sellar region have also reportedly been affected.^{1 3 5 6} The lower extremity has been described as a favoured location for women whereas the upper limbs have been the favoured site for men.^{3 7} This is in contrast to our present case, where the angiomyoma was found in the lower extremity in a middle aged man. Pain is a consistent finding, seen in up to 60–75% of cases, especially in lower limb locations.^{7 8}

The pathological entity of angiomyoma got credence after the comprehensive review by Sturdy in 1937, whereas he postulated the painful subcutaneous condition previously classified as tubercula dolorosa was nothing but a smooth muscle neoplasm arising from the muscular walls of the vein.¹

There have been a few case reports describing the clinical and imaging features of angiomyoma of the extremities, but the point in common in all these studies has been the inability to diagnose the condition preoperatively.^{3 7 8} Angiomyoma usually presents as a painful, mobile, solitary subcutaneous lesion of the extremity, with peak incidence in the fourth to sixth decades.³

On ultrasound, the tumour presents as an oval, smoothly margined, homogeneously hypoechoic mass with diffusely increased internal vascularity. On MRI, the lesion has been described as isointense to skeletal muscle on T1-weighted images and homogenous to heterogeneously hyperintense on T2-weighted sequences. The characteristic features described for angiomyomas include: a hypointense peripheral capsule with linear or branching internal hyperintensities on T2-weighted images and postcontrast diffuse homogenous enhancement, with one study describing a vessel leading up to the lesion in a majority of cases, as also seen in our case.^{3 8 9}

The clinicoradiological differentials for angiomyoma include ganglion cyst, traumatic neuroma, schwannoma, eccrine spiradenoma, angiolipoma, fibroma and synovial sarcoma.^{3 8 9–11}

Ganglion cysts can be easily differentiated from angiomyomas due to their typical fluid signal characteristics, appearing anechoic on ultrasound, and their lack of post-contrast enhancement on MRI.

Neurogenic tumours mimic angiomyomas very closely, presenting as well-defined hypoechoic, oval nodules with internal vascularity; however, they do not show a leading vessel, rather they show connection to a nerve.¹²

Eccrine spiradenomas are rare benign dermal tumours of the sweat gland, appearing heterogeneously hypoechoic on ultrasound with increased internal vascularity on Doppler. On MRI, these lesions are hypointense on T1 images with homogenous postcontrast enhancement, but they have intermediate to low

Figure 2 Postcontrast fat suppressed T1-weighted axial (A) and sagittal images (B) reveal diffuse homogenous enhancement of the mass lesion (black arrow) with a vessel converging on the superior pole of the mass lesion (white arrow).

Figure 3 Gross (A). Cut section showing a circumscribed glistening white nodule with specks of haemorrhage. (B) Microscopy showing a well-circumscribed, encapsulated (arrow) tumour composed of variably sized thick-walled blood vessels encircled by intersecting bundles of smooth muscle cells (arrowhead).

signal on T2 images, thereby differentiating them from angiomyomas.¹⁰

Angiolipomas, which present as painful subcutaneous nodules, are a variant of lipomas. They have a fatty as well as vascular component, with their fatty content rendering them hyperintense on T1-weighted as well as T2-weighted images with signal suppression on short τ inversion recovery (STIR) images helping differentiate them from angiomyomas.¹¹

Fibromas characteristically show hypointense signals irrespective of the pulse sequence employed, thus clinching the diagnosis.³

Synovial sarcomas are slow-growing malignant mesenchymal neoplasms that, unlike other soft tissue sarcomas, are painful. On MRI, large synovial sarcomas have a classic 'Triple sign' suggestive of heterogenous internal architecture but small lesions show signal intensity very similar to that of angiomyomas, appearing hypointense on T1-weighted images and homogeneously hyperintense on T2-weighted images with diffuse post-contrast enhancement.^{13 14} However, synovial sarcomas show an earlier age of incidence, being found in adolescents and young adults, and they lack the T2-weighted well-defined hypointense peripheral rim found in angiomyomas.

Learning points

- ▶ Subcutaneous angiomyoma of the extremity is a rare painful benign condition for which a high suspicion index is warranted as it has a vast differential list including malignant entities such as synovial sarcoma.
- ▶ Radiological features such as a thin peripheral hypointense capsule and internal lacy appearance on T2-weighted MRI, with a leading vessel and diffuse homogenous contrast enhancement, can help diagnose this condition with greater confidence preoperatively.
- ▶ Simple excision is usually curative, with no known recurrence or malignant potential reported.

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