Original Article

Ultrasound and magnetic resonance imaging (MRI) features of angioleiomyoma

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Background: Vascular leiomyoma or angioleiomyoma is a rare benign tumor which belongs to the group of benign smooth muscle tumors. They are commonly located at the extremities, usually presented as a painful solitary lesion in the subcutaneous tissue. To date, very few reports describe its characteristics from an imaging point of view, especially their ultrasound characteristics, making it difficult to do a pre-surgical diagnosis. Our purpose is to describe the clinical, pathologic and imaging features of angioleiomyoma [ultrasound, Doppler ultrasound and magnetic resonance imaging (MRI) findings].

Methods: We retrospectively reviewed from the pathology database from Hospital Universitario Fundación Alcorcón, Madrid, Spain, the clinical histories of 139 patients who had surgical excision and histologic diagnosis of angioleiomyoma during the last 17 years, from May 31st 2006 to April 17th 2023. Of those patients, we focused on 50 who had soft tissue angioleiomyoma with imaging study [ultrasonography (US) and/or MRI] performed in our institution, making a descriptive cross-sectional study.

Results: In our series, a very characteristic ultrasonographic and US Doppler pattern was found. It consists in the presence of a well defined, homogeneous and highly vascularized soft tissue tumor, sometimes with the presence of a feeding vessel.

Conclusions: When the described US features appear in a mobile and elastic subcutaneous slow growing tumor on an extremity, the diagnosis of angioleiomyoma should be considered as highly probable.

Keywords: Angioleiomyoma; ultrasonography (US); Doppler; magnetic resonance imaging (MRI)

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Introduction

Vascular leiomyoma or extremity angioleiomyoma is a benign tumor which belongs to the pericytic (perivascular) tumor group according to the 2020 World Health Organization Classification of Soft Tissue Tumors (1). It arises from the muscularis media of small vessels.

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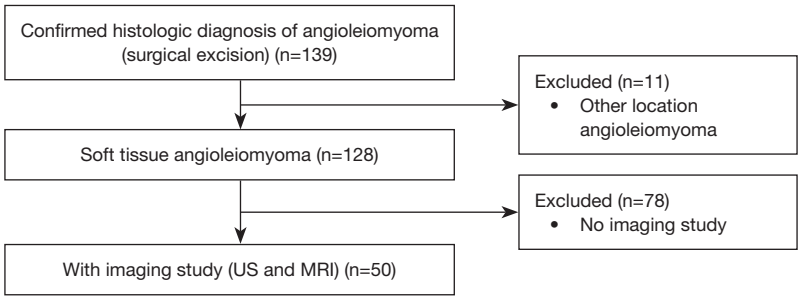

Figure 1 Cases selection flowchart. US, ultrasonography; MRI, magnetic resonance imaging.

It comprises approximately 4.4% of all benign soft tissues' neoplasms and they are commonly located at the lower extremities, usually presented as a painful solitary lesion in the subcutaneous tissue. They are most frequently found in middle-aged women. It is a variety of leiomyoma that has been already described both from the clinical and the pathological standpoint (2-5). There are three pathological subtypes: solid, cavernous and venous (6), and preference of the venous subtype for the head and neck region (5).

To date, there are very few reports describing its characteristics from an imaging point of view; and always referring to isolated cases, most of them located in the hand (5-8). The fact that it is an uncommon tumor and that its radiologic appearance has seldom been described, makes it difficult to do a pre-surgical diagnosis, considering that it can show a morphological overlap with a variety of benign and malignant soft-tissue tumors, including angiomyolipoma, myopericytoma and leiomyosarcoma.

The aim of this paper was to describe the clinical, histologic and specially imaging features of this tumor [ultrasonography (US), Doppler US and magnetic resonance imaging (MRI)] in a series of 50 soft tissue angioleiomyomas from our hospital (Hospital Universitario Fundación Alcorcón, Madrid). We present this article in accordance with the STROBE reporting checklist (available at https://qims.amegroups.com/article/view/10.21037/qims-24-602/rc).

Methods

We retrospectively reviewed from the pathology database from Hospital Universitario Fundación Alcorcón, Madrid, Spain, the clinical histories of 139 patients who had surgical excision and histologic diagnosis of angioleiomyoma during the last 17 years, from May 31st 2006 to April 17th 2023. We selected 128 of those who had soft tissue angioleiomyoma, and the 11 cases with other location tumors were excluded. Of those 128 cases, 78 were excluded because they had no imaging studies. So our study focuses on 50 patients with soft tissue angioleiomyoma with imaging studies (US and MRI) before the excision (*Figure 1*).

All of these patients were studied with the US equipment from our hospital, a Toshiba Aplio MX Ultrasound and GE Logiq E10 R3, with multifrequency US probes (7–13 MHz). B mode imaging was performed, as well as a thorough pulsed, color and power Doppler evaluation. MRI was performed in 6 of these patients with a 1.5 T imager (General Electric 1.5 T Signa Artist). Superficial and anteroposterior gradient coils were used. Several sequences were acquired: spin-echo (SE) T1 (repetition time/echo time, 400–620 ms/12–20 ms), fast spin-echo (FSE), proton density (PD) and T2 (3,000–6,000/18–120/8 echo train length) with and without fat suppression; gradient-echo (400/14/20°) and short time inversion recovery (STIR)[3,400–3,700/36–43/inversion time (TI): 150 ms]. A 256×160–256 acquisition matrix with a field of view according to the lesion size was used (8–16 cm). Four of these patients received an intravenous (IV) injection of gadolinium-diethylene triamine pentaacetate (Gd-DTPA). One patient was also studied with computed tomography (CT) and 1 with contrast-enhanced ultrasound. US, MRI and CT images were stored using a picture archiving communication system (PACS). The images were retrospectively evaluated, taking into account the clinical history. All US and MRI studies were performed and evaluated by radiologists with at least 15 years experience.

In US and US Doppler studies, we evaluated the size, morphology, echogenicity, echostructure, vascularization and presence of feeding vessels. In MRI, we evaluated size, morphology, signal intensity and contrast enhancement. Surgical excision and pathologic correlation were performed in all patients.

Table 1 Clinical aspects

Clinical aspects	Value (n=50)
Gender	
Male	22
Female	28
Age (years)	
Average	53±3
Evolution (years)	
Median, p25–p75	2, 0
Size (cm)	
Average	1±3
Location	
Hand	16
Arm	6
Leg	7
Foot	19
Head	1
Other	1
Pain	
No	19
Yes	31

Data are presented as mean ± standard deviation or number.

Table 2 Ultrasound features

Ultrasound features	N
Morphology	
Oval, well defined, smooth margins	49
Other	1
Total	50
Consistency	
Elastic	13
Soft	8
Solid	17
Total	38
Not described	12
Total	50
Echogenicity	
Hipoechogenic	41
Hiperechogenic	5
Total	46
Not described	4
Total	50
Blood flow on us doppler	
No	14
Yes	33
Total	47
Not described	3
Total	50
Feeding arterial vessel	
Yes	9
Not described	41
Total	50

The study was conducted in accordance with the Declaration of Helsinki (as revised in 2013). This study was deemed exempt by the Ethics committee's board for a formal IRB request, and in keeping with the policies for a retrospective review, informed consent was not required.

Results

Clinical aspects (Table 1)

- The mean age of our group was 53 years and the gender distribution was 28 female, 22 male.
- 26 cases were located in lower limbs (7 in legs, 19 on feet), 22 in upper limbs (6 in arms, 16 in hands), 1 in the head and 1 in the back.
- On physical examination most of them were elastic or solid and mobile tumors with slow growth (average duration of 2 years).
- 31 patients complained of pain and 19 were asymptomatic.
- The most frequent clinical diagnoses of our cases, before the imaging studies, were: lipoma, ganglion and giant cell tumors.

Imaging features

Sonography and echo-Doppler (*Table 2*)

The sonographic findings were characteristic and very similar in all cases. Their appearance was a subcutaneous

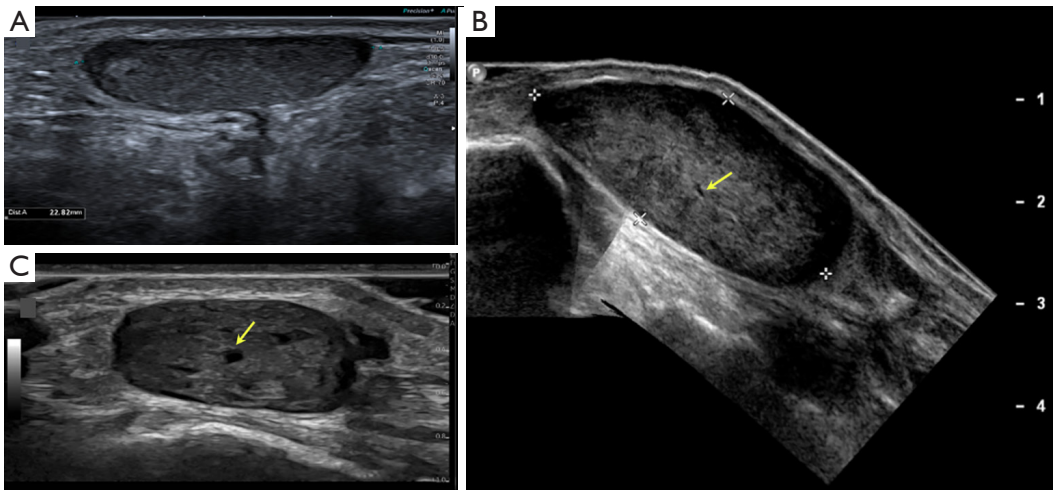

Figure 2 B mode US typical angioleiomyoma pattern in three different patients: (A) female, 30 years old, longitudinal image of the left ankle; (B) male, 42 years old, longitudinal image of the ankle; (C) female, 52 years old, angioleiomyoma of the hand. All of them are subcutaneous solid and oval tumors, with smooth or lobulated well defined margins and a homogeneous and hypoechogenic background, showing small lineal anechoic images that correspond to small vessels (yellow arrows). US, ultrasonography.

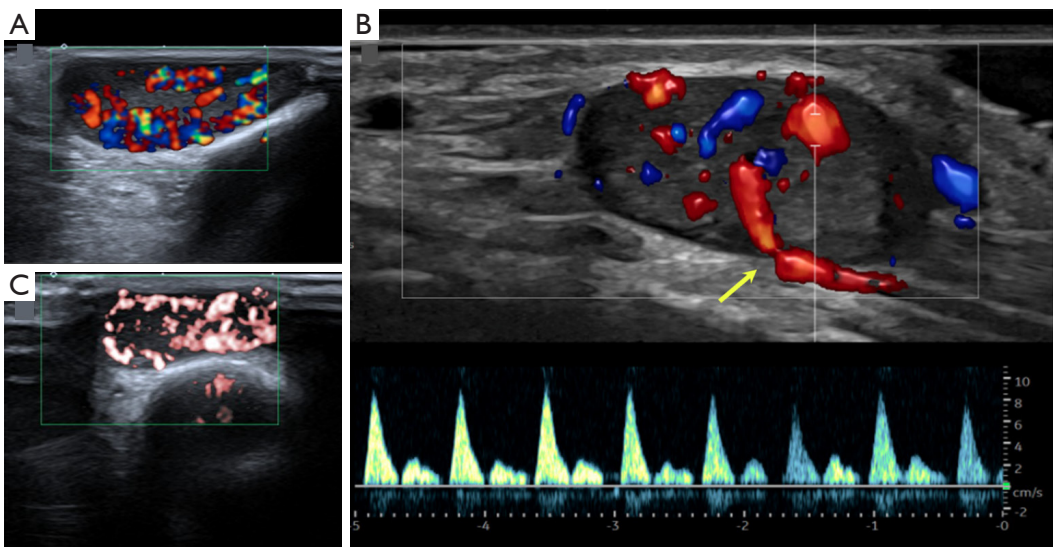

Figure 3 US Doppler typical angioleiomyoma pattern in three different patients: (A) female, 38 years old, toe angioleiomyoma; (B) male, 60 years old, hand angioleiomyoma, pulsed and color echo Doppler of the intratumoral vascular pattern with intense blood flow; (C) female, 53 years old, pulsed and color echo Doppler, longitudinal view of the hand, showing the typical vascular pattern with a feeding vessel (yellow arrow).

solid and homogeneous tumor with smooth or lobulated margin (*Figure 2*). The size ranged from 3 to 40 mm in its maximum diameter (average 11 mm).

Four patients had calcified tumors: 3 of them showed small calcifications and one was completely calcified. Small vessels with blood flow were seen in an echogenic background (*Figures 2,3*). In 9 of the cases a feeding vessel was seen (*Figure 3C*). Fourteen patients didn't show any blood flow on US Doppler; 3 of them were calcified tumors and the other 11 were small tumors, between 4 and 10 mm.

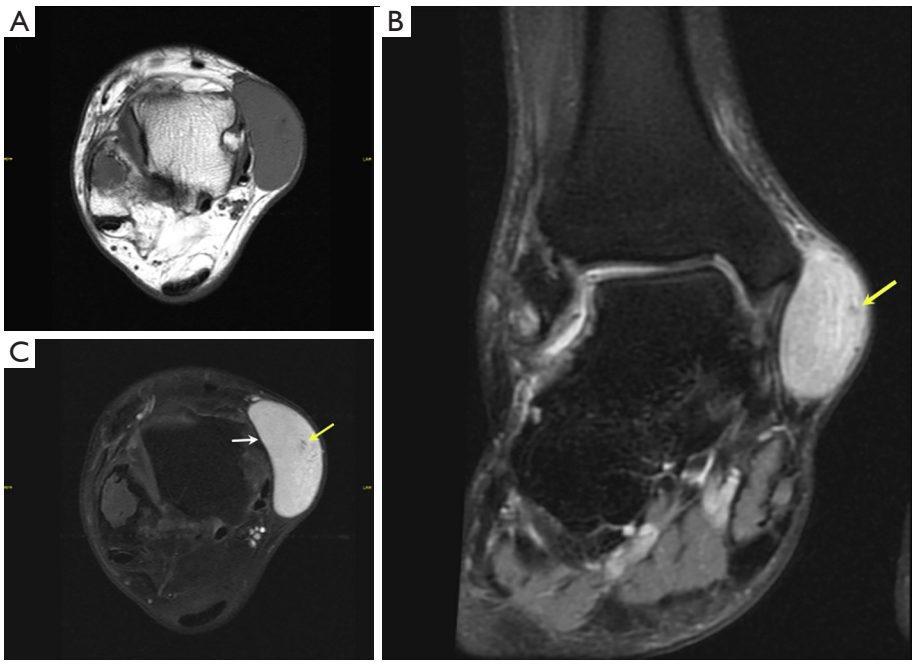

Figure 4 MRI images of a 43 years old male with a subcutaneous ankle angioleiomyoma. (A) Axial SE T1. A well defined subcutaneous tumor with a low signal intensity background. (B) Coronal FSE PD with fat suppression. High signal intensity background and thin signal voids (yellow arrow) corresponding to the dark reticular pattern. (C) Axial SE T1 fat suppression post-gadolinium. Tumor enhancement with a thin hypointense capsule (white arrow) and the characteristic dark reticular pattern (yellow arrow). MRI, magnetic resonance imaging; SE, spin-echo; FSE, fast spin-echo; PD, proton density.

MRI

MRI was performed in only 6 patients. It showed a solid well defined tumor, with an isointense to the muscle signal on SE T1 and intermediate to high signal intensity on inversion-recovery (IR), FSE, proton density-weighted sequence (DP) and T2-weighted fat suppression sequences (*Figure 4*). The images showed strong enhancement of the tumors after intravenous injection of paramagnetic contrast material, best depicted on fat suppression SE T1 (*Figure 4C*). On post gadolinium SE T1 images they also showed a thin capsule, which appears hypointense on T2-weighted images (*Figure 4C*).

Inside the mass, multiple thin hypo or iso-intense linear structures were seen on T2 and DP sequences, showing the characteristic "dark reticular sign" of angioleiomyomas.

Pathologic findings

Macroscopically the tumors were small ovoid masses with variable appearance when cut: solid whitish-yellow mass, sometimes with reddish areas; or myxoid gray or white liquid-pasty mass. In all of the cases, the histological findings were the same and very characteristic: proliferation of whirled smooth muscle tissue surrounding numerous vascular channels (*Figure 5*). They were not classified on the different types due to a lack of description in the histopathology reports.

Discussion

Angioleiomyomas are benign perivascular soft tissue solitary tumors, originating from the smooth muscle layer of blood vessels. They belong to the perivascular tumor group according to the 2020 World Health Organization Classification of Soft Tissue Tumors.

They are seen in patients aged between 30 and 60 years, with a male:female ratio of about 1:2 (1-4). The mean age and gender distribution of our group were similar to the reported cases.

In large series (3) there is a prevalence of lower limb lesions, as in our series, where 26 cases were located in lower limbs and 22 in upper limbs.

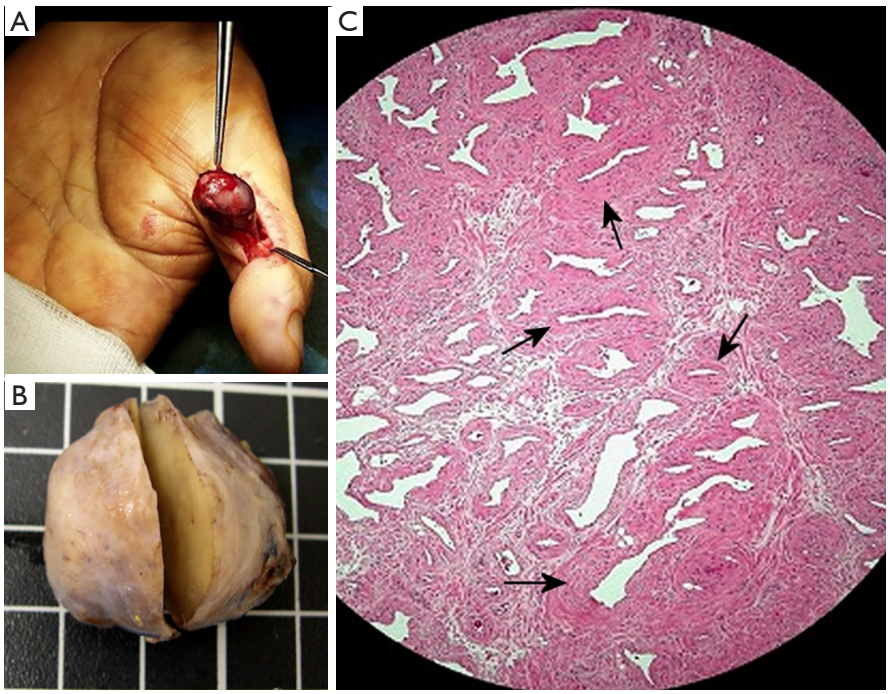

Figure 5 Macroscopical and histological characteristics of angioleiomyoma. (A) Surgical image of a hand angioleiomyoma. (B) Macroscopic image of a tumor located in the knee. (C) Histopathological features (H&E staining, ×100): smooth muscle proliferation surrounding vascular channels (arrows). H&E, hematoxylin and eosin.

The clinical symptoms and clinical course of our cases are similar to those of larger series: they are slow growing tumors, and some of the patients presented pain. The prevalence of this symptom is variable in literature, and has been related to local ischaemia (9) and to the presence of nerve fibers in the capsule or inside the tumor (1,2,9). On physical examination they are mobile and elastic or solid tumors.

Imaging features

The sonographic findings were characteristic and very similar in all cases and to large series. Their appearance was a subcutaneous solid and homogeneous tumor with smooth or lobulated margin. The size ranged from 3 to 40 mm in its maximum diameter (average 11 mm). Typically, small vessels with blood flow are seen in an echogenic background (*Figures 2,3*), and in 9 of the cases a feeding vessel was seen (*Figure 3C*). This finding has been reported in other series (10,11).

Fourteen patients did not show any blood flow on US Doppler; 3 of them were calcified tumors and the other 11 were small tumors, between 4 and 10 mm, which could explain why blood flow was not seen on US Doppler in these patients.

The presence of calcifications has been reported (3) and has been interpreted as degenerative, probably related to repetitive minor trauma. In our series, 4 patients had calcified tumors: 3 of them showed small calcifications and one was completely calcified. These findings are not common in angioleiomyomas, and could lead to a wrong diagnosis.

MRI was performed in only 6 patients and showed similar findings to those reported by other authors (6,7): a solid well defined tumor, with an isointense to the muscle signal on (SE) T1 and intermediate to high signal intensity on IR, FSE, DP and T2-weighted fat suppression sequences (*Figure 4*), with strong enhancement after IV injection of paramagnetic contrast, showing a thin capsule and multiple thin hypo or iso-intense linear structures on T2 and DP sequences, the characteristic "dark reticular sign" of angioleiomyomas (12).

Differential diagnosis

This tumor must be distinguished from the following

lesions (8): neural sheath tumor, hemangioma, inclusion epidermoid cyst and tenosynovial giant cell tumors. There is a rare subcutaneous avascular variety of leiomyoma which is more hypoechogenic than angioleiomyoma and without Doppler flow. Small nervous sheath tumors show a less conspicuous and mostly peripheral Doppler flow, and are related to a nervous structure. Angioleiomyomas may be difficult to distinguish from some subcutaneous hemangiomas which may show arterial flow and no phleboliths; but hemangiomas are usually more heterogeneous structure, with fat tissue and heterogenous intranodular blood flow. Inclusion epidermoid cysts do not have vascular structures inside and show very little or no Doppler flow at all. Angioleiomyoma must also be distinguished from protruding round tendinous sheath tumors, basically from giant cell tumor, which is related to the tendinous sheath and more lobulated, hypovascular, hypoechogenic heterogeneous background tissue. Moreover, they are also typically hypointense in GE T2-weighted images, due to the presence of hemosiderin.

Strengths and limitations

This is a retrospective, cross sectional descriptive study with a significant number of cases for an uncommon entity like angioleiomyoma. All of our patients have US studies, which help us describe with certainty the characteristic US features of this tumor, which is a notable strength of this study.

But it has some limitations. First, although it is one of the largest case series to have studied angioleiomyoma based on US findings, the number of cases is still insufficient at 50. The inclusion of cases from the anatomopathological database of the institution and the retrospective nature of the study led to some missing data and there was also some incomplete information in the anatomopathological reports.

For MRI findings we only had 6 patients, which is a small number to make a valid descriptive study. The small number of patients with MRI studies is because the US-Doppler pattern was considered enough to make a pre-excision diagnosis in most of the cases, with no need to perform an MRI.

Conclusions

Whenever a patient presents with a long duration and subcutaneous small elastic mobile nodule in the extremities, and the ultrasound features and Doppler ultrasound pattern of the tumor are those already described above, the diagnosis of angioleiomyoma should be suggested as the most probable. When performed, MRI and its characteristic findings, such as strong enhancement and the "dark reticular sign", support the diagnosis of angioleiomyoma.

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Footnote

Provenance and Peer Review: With the arrangement by the Guest Editors and the editorial office, this article has been reviewed by external peers.

Reporting Checklist: The authors have completed the STROBE reporting checklist. Available at: https://qims.amegroups.com/article/view/10.21037/qims-24-602/rc

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://qims.amegroups.com/article/view/10.21037/qims-24-602/coif). The special issue "Advances in Diagnostic Musculoskeletal Imaging and Image-guided Therapy" was commissioned by the editorial office without any funding or sponsorship. J.M.V. served as the unpaid Guest Editor of the issue. The authors have no other conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. The study was conducted in accordance with the Declaration of Helsinki (as revised in 2013). This study was deemed exempt by the ethics committee's board for a formal IRB request, and in keeping with the policies for a retrospective review, informed consent was not required.

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