

Magnetic resonance imaging features of vascular leiomyoma of the ankle

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ABSTRACT

Vascular leiomyoma is a benign soft tissue tumour with a predilection for middle-aged women. It is most often seen in the extremities, particularly in the lower leg. The typical lesion is a small, slow-growing subcutaneous nodule. These tumours are often unexpected or preoperatively confused with other soft tissue tumours including low-grade sarcomas, leading to wide surgical excision. This may partly be due to the relatively few studies delineating the characteristic imaging features of this entity. Here, the imaging findings of a case of vascular leiomyoma in the ankle are presented. Literature review of the magnetic resonance imaging findings of published reports and series of vascular leiomyomas of the extremities is also performed.

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Case report

A 47-year-old previously healthy Hong Kong Chinese man presented in January 2012 with a 2-year history of a slow-growing painless mass over the right medial malleolus. Physical examination showed a soft, well-marginated, non-tender mass measuring 2 cm in diameter over the right medial malleolus. The patient was referred for ultrasound and subsequently magnetic resonance imaging (MRI; Figs a to i). The lesion was excised. Macroscopically, it was a disc-shaped mass with smooth outer surface. Cut section showed a mass with a thin capsule and homogeneous, greyish-to-whitish material without necrosis. Microscopy showed proliferation of smooth muscle cells associated with thick-walled blood vessels without evidence of malignancy. The histopathological diagnosis was vascular leiomyoma (Figs j and k).

Discussion

Vascular leiomyoma, angiomyoma or angioleiomyoma, is a rare benign smooth muscle tumour that originates in the tunica media of veins and arteries. It can be located in the skin, subcutaneous fat, or superficial fasciae of the extremities. It has a predilection for middle-aged women. It can occur anywhere in the body, but is most often seen in the extremities, particularly in the lower leg.¹

The most frequent clinical presentation is a mass that enlarges slowly over several years. The

size usually ranges from subcentimetre to a few centimetres in diameter, but occasionally may grow larger. They are usually oval or round in shape, and can be located in the skin, subcutaneous fat, or the superficial fasciae of the extremities.

Pain, with or without tenderness, has been reported in about 60% of patients, and is thought to be caused by the active contraction of smooth muscles resulting in local ischaemia, and is also suggested to be mediated by intratumoural nerve fibres.² Treatment usually consists of marginal excision.²

Angioleiomyomas are rarely diagnosed preoperatively. In a series of 10 cases by Gupta et al¹ in 2008, the preoperative or pre-biopsy imaging diagnoses included sarcoma not otherwise specified, schwannoma, myositis ossificans, synovial sarcoma, and fibroma. This may be partly due to the relatively few studies delineating the characteristic imaging features of this entity. The preoperative differentiation of angioleiomyoma from other soft tissue tumours is of clinical importance, especially sarcomas, since angioleiomyomas are benign and can be treated with simple excision. Literature review of the MRI findings of currently published reports and series of vascular leiomyomas of the extremities is presented below.

Literature review

Materials, methods, and patient demographics

A PubMed search of the English literature

腳踝血管性平滑肌瘤的磁力共振影像特徵

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血管性平滑肌瘤是一種良性軟組織瘤，患者多為中年女性，通常見於四肢，尤其下肢。典型的血管性平滑肌瘤是一顆生長緩慢的小結節。手術之前一般很難預料到這種瘤，也容易和其他軟組織瘤混淆，例如一些低度惡性肌瘤，因而引致廣泛切除的手術方法。部分原因可能是現時文獻有較少關於血管性平滑肌瘤特有的影像診斷特色。本文報告一宗位於腳踝的血管性平滑肌瘤的病例，並回顧相關文獻。

was performed, using the key words “vascular leiomyoma”, “angioleiomyoma”, and “angiomyoma”. From 1998 to 2011, 36 cases of biopsy-proven vascular leiomyomas in the extremities of adults with detailed descriptions of T1-weighted images (T1WI) and T2-weighted images (T2WI) were found. Articles without detailed descriptions or figures of T1WI and T2WI were excluded. Not all studies in the literature may have been included in this review because of unavailability in PubMed or in English language. After including our case, this review has 37 cases. The mean age of the patients

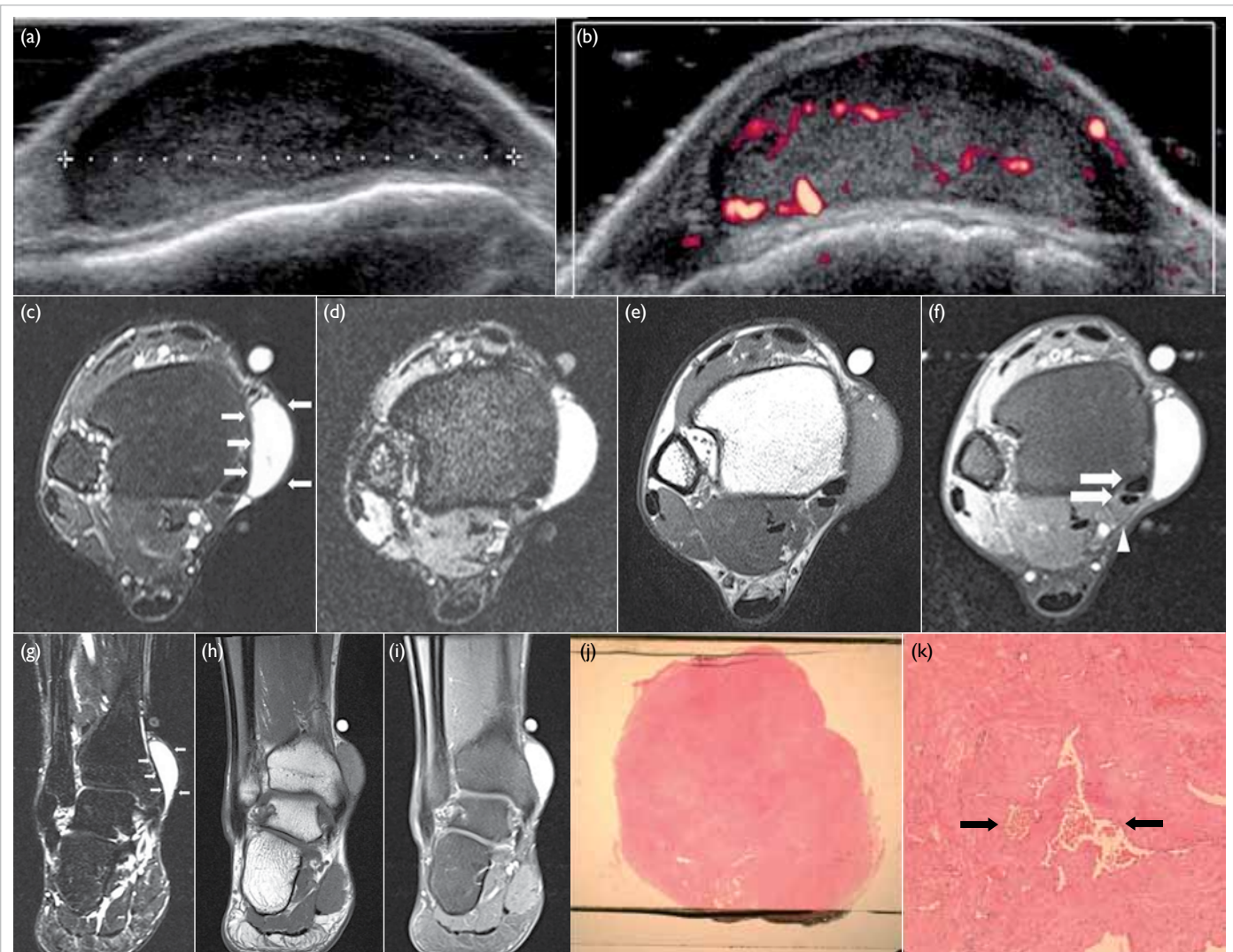


FIG. A 47-year-old man with a 2-year history of a slow-growing, painless, soft ankle mass

(a) Grey-scale ultrasound and (b) power Doppler study demonstrate a well-circumscribed, mildly compressible, mildly echogenic, subcutaneous lesion adjacent to the right medial malleolus with intralesional slow-flow vessels. No feeding artery, dilated draining vein, or phlebolith is found. Overall features are suggestive of a mildly vascular solid mass. (c, g) It is well-circumscribed and markedly hyperintense with a peripheral hypointense rim (arrows) on axial and coronal T2-weighted images. (d) No susceptibility artefacts are observed in the lesion on gradient echo sequence image, suggesting the absence of intralesional haemorrhage. (e, h) It is slightly hyperintense to muscle on T1-weighted images, with (f, i) homogeneous enhancement upon intravenous administration of gadolinium. The underlying bone, subjacent flexor retinaculum (arrowhead) and medial ankle tendons (arrows) were intact. (j, k) Microscopic pictures of the tumour section include central blood vessels (arrows) surrounded by peripheral smooth muscle. The histopathological diagnosis is vascular leiomyoma (H&E, original magnification: [j] $\times 5$ and [k] $\times 100$)

was 51 years (range, 20-72 years). There were 16 male and 17 female patients; the gender of the remaining four patients was not stated.

Results

Among the 26 lesions with documented sizes, the mean size of the lesions was 3.2 cm (range, 0.4-12 cm). Overall, 40.5% (15/37) of the lesions were in the upper limb and 59.5% (22/37) were in the lower limb. All of them were located in the subcutaneous layer, were well-defined, and round, oval or disc-shaped. On T1WI, 91.9% (34/37) of the tumours showed isointense-to-slightly high signal intensity, 5.4% were heterogeneous, and 2.7% showed low signal intensity. On T2WI, all the cases demonstrated high signal intensity. Signal voids were seen in 10.8% (4/37) of the tumours, either on T1WI or T2WI. Among the 33 cases in which contrast was administered, only two (6.1%) cases showed no or poor enhancement, 93.9% (31/33) showed enhancement, 42.4% (14/33) were homogeneous, and 39.4% (13/33) were described as showing heterogeneous enhancement. One case showed peripheral enhancement, one showed central enhancement. One case showed rapid enhancement and one case demonstrated slow enhancement. Among the cases in which the presence or absence of peripheral hypointense rim was recorded, a hypointense rim was found on T2WI in 85.2% of cases (23/27; Table¹⁻¹⁴).

Vascular leiomyomas often show similar signal intensity to that of muscle on T1WI. A T2WI is expected to demonstrate mixed areas that are hyper- and isointense to muscle. A well-defined peripheral T2-hypointense rim may be seen, representing the fibrous capsule. It has been reported that T2-hyperintense areas correlated with strong contrast enhancement, whereas isointense areas did not show enhancement after intravenous administration of contrast material.³ It was suggested that the smooth muscle and numerous vessels corresponded to the hyperintense areas, and the fibrous tissue appeared isointense on T2WI. Tortuous vascular structures with signal void may also be seen.

Imaging differentials

The differentials of a well-defined, enhancing, subcutaneous nodule or mass with T2-hyperintense signals include synovial sarcoma, other low-grade soft tissue sarcomas, haemangioma, neurogenic tumour, and nodular fasciitis.

Low-grade sarcomas such as synovial sarcoma and low-grade myxofibrosarcoma may be slow growing and appear well-circumscribed on MRI, giving the misleading impression that the lesion is well-localised. Haemorrhage may be present in synovial sarcomas, which may be seen as fluid-fluid levels, T2 hypointensity, or "triple signal intensity", namely areas of hyperintensity, isointensity and

TABLE. Magnetic resonance imaging findings in 37 patients with vascular leiomyomas in extremities¹⁻¹⁴

Variable	Data
Age in years (among 33 patients with documented age)	
Mean	51
Range	20-72
Sex (among 33 patients with documented sex)	
Male	16 (48.5%)
Female	17 (51.5%)
Location (n=37)	
Upper limb	15 (40.5%)
Lower limb	22 (59.5%)
Size in cm (among 26 lesions with documented sizes)	
Mean	3.2
Range	0.4-12
Shape	
Well-defined; and were round, oval or disc-shaped	All
T1-weighted signal intensity to muscle (n=37)	
Isointense-to-slightly high signal intensity	34 (91.9%)
Heterogeneous	2 (5.4%)
Slightly low	1 (2.7%)
T2-weighted signal intensity to muscle	
High signal intensity	100%
Signal void on either T1- or T2-weighted images (n=37)	4 (10.8%)
Enhancement (n=33)	
No or poor enhancement	2 (6.1%)
Enhancement present	31 (93.9%)
Homogeneous enhancement	14 (42.4%)
Heterogeneous enhancement	13 (39.4%)
Rapid enhancement	1 (3.0%)
Slow enhancement	1 (3.0%)
Enhancing, not otherwise specified	2 (6.1%)
Peripheral T2-hypointense rim (among cases where its presence/absence was noted) [n=27]	
Hypointense rim present	23 (85.2%)

hypointensity relative to fat, due to presence of cystic, solid and fibrous elements with haemorrhage. It is unknown whether the absence of haemorrhage, a more homogeneous appearance, and the presence of a peripheral hypointense rim are reliable distinguishing features favouring angioleiomyoma over otherwise benign-appearing, soft tissue sarcomas; this may be a potential knowledge gap that future prospective comparison studies may serve to fill.

Haemangiomas may show homogeneous signals if these are small, making it challenging to differentiate from angioleiomyomas. Phleboliths

can be sought for on plain radiographs. Fatty and serpentine vascular elements may be identified in haemangiomas, which are pathognomonic. The classical 'target' sign, 'split-fat' sign, and fusiform tumour shape demonstrated in neurogenic tumours are not found in angioleiomyomas. Although nodular fasciitis demonstrates similar shape and size as angioleiomyomas, linear extension along the fascia, surrounding oedema, low T1 signal, heterogeneous T2 signal, and non-homogeneous enhancement are features that differ from characteristic imaging features of angioleiomyoma.¹⁵

On microscopic examination, the presence of tortuous vascular channels surrounded by smooth muscle bundles and areas of myxoid change may be seen. This explains the heterogeneity of signal intensity in the tumour on T2WI. Magnetic resonance imaging–histopathological correlation published by Hwang et al² stated that the smooth muscle and numerous vessels within each type of vascular leiomyoma corresponded with the hyperintense areas on T2WI, and the tough fibrous tissue appeared isointense on T2WI. In addition, a well-defined peripheral hypointense area on T2WI correlated with the fibrous capsule, and the interlacing isointense areas within the tumour correlated with the various quantity of connective tissue and intravascular thrombus.³

Conclusions

Vascular leiomyoma should be considered a possible diagnosis when a well-demarcated oval or round subcutaneous mass with T1-isointense-to-slightly high signal, T2-high signal intensity, hypointense rim, and intense enhancement is seen in the soft tissue of the extremities. It is unknown whether the absence of haemorrhage, a more homogeneous appearance, and the presence of a peripheral hypointense rim are reliable distinguishing features favouring angioleiomyoma over otherwise benign-appearing soft tissue sarcomas; this may be a potential knowledge gap that future prospective comparison studies may serve to fill.

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