A large vascular leiomyoma of the leg

E. Cigna^a, M. Maruccia^{a,*}, G. Malzone^a, F. Malpassini^b, G. Soda^c,
F.M. Drudi^b

^a Department of Plastic Surgery, Policlinico Umberto I, University of Rome "Sapienza", Italy

^b Department of Radiological Sciences, Policlinico Umberto I, University of Rome "Sapienza", Italy

^c Department of Anatomical Pathology, Policlinico Umberto I, University of Rome "Sapienza", Italy

KEYWORDS

Vascular leiomyoma;
Angioleiomyoma;
Subcutaneous tumor;
Benign tumor.

Abstract A 69-year-old woman with a subcutaneous, large vascular leiomyoma of the leg is presented. The patient had a painful, slow-growing, right medial malleolus mass. Clinical symptoms, US images and histopathologic features are reported. Vascular leiomyoma should be included in the differential diagnosis of painful, lower extremity subcutaneous masses also in lesions of larger dimensions.

Sommario Riportiamo il caso di una donna di 69 anni, con un grande leiomioma vascolare sottocutaneo della gamba. La paziente si presenta con una massa dolorosa e a lenta crescita a livello del malleolo mediale destro. Presentiamo le sue caratteristiche cliniche, ecografiche e anatomicopatologiche. Il leiomioma vascolare dovrebbe essere incluso nella diagnosi differenziale di una massa dolorosa, sottocutanea delle estremità inferiori anche se presenta grandi dimensioni.
© 2011 Elsevier Srl. All rights reserved.

Introduction

Vascular leiomyoma or angioleiomyoma is a rare, benign tumor originating from smooth muscle cells of the arterial or venous walls. This type of tumor can arise on any part of the body and can be found in the dermis, the subcutaneous fat or the deep fascia and it commonly occurs in the extremities, such as the lower leg (50–70%). Clinically, this tumor appears as a small (<20 mm), freely movable, subcutaneous nodule causing pain in approximately 60% of patients; it occurs more frequently in women than in men, and pregnancy may increase the severity of the pain.

Diagnosis of these lesions is frequently delayed because of their rare occurrence and due to the lack of awareness of this clinical condition. Ultrasound (US) is often used in the initial evaluation of soft tissue masses and the treatment is usually surgical excision. Due to the biological characteristics of leiomyoma the recurrence rate is low [1,2].

This report presents the case of a female patient who had an unusually large (>20 mm) angioleiomyoma of the right medial malleolus.

Case report

In March 2009, a 69-year-old woman was presented to our Department with a painful, slow-growing, right medial malleolus mass that she had had for 15 years. During these 15 years she had felt episodic pain in the affected area,

* Corresponding author.

E-mail address: marucciam@gmail.com (M. Maruccia).

Figure 1 (A) US examination showed a 22.4×13.4 mm homogeneous, hypoechoic lesion in the subcutaneous soft tissue with a small amount of posterior acoustic enhancement. (B) Power Doppler sonogram revealed diffuse arterial hypervascularity.

particularly after prolonged walking or during more intensive activity. She reported that the mass had progressively increased in size and had become large enough to painfully rub against her shoes or boots. She had no fever, lymphadenopathy, involuntary weight loss or loss of energy. There was no sign of weakness or dysesthesias in the lower extremities, and there had been no skin breakdown or skin color changes. Palpation revealed a firm, elastic, mobile mass located close to the right medial malleolus. The patient underwent US examination, which confirmed the presence of a round nodule located in the subcutaneous fat tissue between the right medial malleolus and the posterior tibial vessels. The mass presented a relatively homogeneous, hypoechoic echotexture with a small amount of posterior acoustic enhancement. No internal calcification was evident and color Doppler evaluation revealed diffuse arterial hypervascularity (Fig. 1A and B).

The patient experienced complete resolution of symptoms at inflation of a tourniquet to a value above her systolic blood pressure, thus indicating a positive ischemic response. Despite an uncertain preoperative diagnosis, the patient underwent surgical excision under local anesthesia. A vertical incision of approximately 3.5 cm was made at the posterior medial region of the right leg. In the subcutaneous tissue, there was a well-demarcated mass, not adhering to the posterior tibial vessels (Fig. 2). The mass was easily removed; it was a hard, round-shaped tumor measuring approximately 22×13 mm in diameter and not adhering to the deep or superficial tissues. The histopathological report described a well-circumscribed, non-encapsulated nodular tumor made up of smooth muscle bundles that were closely compacted and intersecting one another, surrounded by thick fibrous tissue. The smooth muscle fascicles had uniform spindle cells with eosinophilic cytoplasm and cylindrical nuclei with blunted ends. The proliferation surrounded blood vessels and slit-like vascular channels (Fig. 3A). Immunohistochemical smooth muscle actin staining evidenced the presence of muscular fibers, thus confirming the diagnosis of vascular leiomyoma (Fig. 3B).

The patient experienced immediate and complete relief of her symptoms after surgery.

Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

Discussion

Vascular leiomyomas or angioleiomyomas are benign, solitary tumors of smooth muscle cells originating from the muscular layer of the vessel walls. Stout published the first comprehensive review of this rare lesion in 1937, and it has been well described in the literature since that time [2].

The condition is more common in females than in males (female to male ratio is 2.7:1) [2]. These tumors can affect individuals of any age, but they are more common in people between 30 and 60 years of age [2]. Ninety per cent of these lesions occur in the extremities, with the head and trunk accounting for the remaining cases. The lower extremity is involved in 50–70% of the extremity lesions, which are twice as common in females. Conversely, lesions of the head, neck and upper extremities are more frequent in males. Pain is the most characteristic symptom reported in the majority of patients (60–75%) [1,2].

Figure 2 Intraoperative photograph of a well-demarcated spherical vascular leiomyoma located in the subcutaneous tissue.

Figure 3 Photomicrographs showing the histopathology of vascular leiomyoma. (A) Closely compacted, intersecting smooth muscle bundles surrounding slit-like vascular channels. No evidence of necrosis, pleomorphism, mitoses or nuclear atypia. (Hematoxylin-Eosin 100 X). (B) Positive immunohistochemical staining for smooth muscle actin (100 X).

So far, three different histological subgroups of angioleiomyomas have been identified: solid, cavernous and venous [3]. Solid tumors are found more commonly in the lower extremities [2], and pain is the most common presenting symptom. Our patient's chief complaint was therefore typical for angioleiomyoma. Vascular leiomyomas are sharply demarcated, spherical tumors that are usually up to 10 mm in diameter. The reported case shows that angioleiomyomas may be larger, also >20 mm in diameter.

Differential diagnosis for painful subcutaneous extremity lesions includes among others, glomus tumors, hemangiomas, angiolipomas, ganglions and traumatic neuromas [4]. Diagnosis depends on histopathological identification using staining techniques, such as Masson's trichrome, hematoxylin-eosin, alcian blue, van Gieson, and periodic acid-Schiff (PAS). Immunohistochemical stains for vimentin, desmin and smooth muscle actin can be of great value when available [5]. In this case, hematoxylin-eosin stain and immunohistochemical stain for smooth muscle actin were used.

There are no specific imaging techniques which can characterize vascular leiomyoma. However, US is an excellent modality for the initial evaluation of such a lesion. In this case, US was essential for identifying the distance between the mass and the posterior tibial vessels and for describing the location and size of the tumor. The homogeneity of the lesion, lack of calcification and lack of compressibility excluded a possible hemangioma, and the solid nature of the mass, as evidenced by hyperemia, basically excluded a cyst and other fluid collections. The location of the mass and the lack of compressibility were also unusual for a lipoma.

The vascular flow of these tumors shows a high resistance, thus suggesting the presence of muscular arteries. US findings suggestive of angioleiomyomas include well-defined margins, hypervascularity, and small dimensions. When a slow-growing, lower extremity mass presents these features, diagnosis of angioleiomyoma should be considered even in lesions presenting a diameter of more than 10 mm.

Hildreth's ischemic test is diagnostic for glomus tumors in the extremities, but leiomyoma can only be distinguished from glomus tumor by histological evaluation [6,7]. The test was positive in this patient for vascular leiomyoma [8]. If

the pain disappears when tourniquet is inflated above the systolic pressure, this suggests a vascular component of the lesion.

In conclusion, angioleiomyoma is a rare soft tissue tumor presenting clinical symptoms which are typical, but non-specific. Vascular leiomyoma should be included in the differential diagnosis of painful lower extremity subcutaneous masses even when the mass is larger than 20 mm in diameter. Excision of the lesion permits histopathologic diagnosis and offers complete resolution of symptoms, as shown in this case.

Conflict of interest statement

The authors have no conflict of interest to disclose.

Appendix. Supplementary data

Supplementary data related to this article can be found online at [doi:10.1016/j.jus.2011.10.007](https://doi.org/10.1016/j.jus.2011.10.007).

References

- [1] Requena L, Baran R. Digital angioleiomyoma: an uncommon neoplasm. *J Am Acad Dermatol* 1993;29:1043–4.
- [2] Hachisuga T, Hashimoto H, Enjoji M. Angioleiomyoma: a clinicopathologic reappraisal of 562 cases. *Cancer* 1984;54:126–30.
- [3] Morimoto N. Angiomyoma: a clinicopathologic study. *Med J Kagoshima Univ* 1973;24:663–83.
- [4] Arenas R. *Dermatologia. Atlas, Diagnostico Y Tratamiento*. 2nd ed. Mexico: McGraw-Hill Interamericana; 1996. 438–442.
- [5] Nicolai P, Redaelli de Zinis LO, Facchetti F, Maroldi R, Antonelli AR. Craniofacial resection for vascular leiomyoma of the nasal cavity. *Am J Otolaryngol* 1996;17:340–4.
- [6] Enzinger FM, Weiss SW. *Benign tumors of smooth muscle & glomus tumors*. St.Louis. In: Mosby CV, editor. *Soft tissue tumors*. 2nd ed., 383–401; 1988. p. 581–95.
- [7] Baran R, Requena L, Drape JL. Subungual angioleiomyoma masquerading as a glomus tumour. *Br J Dermatol* 2000;142:1239–41.
- [8] Hildreth DH. The ischemia test for glomus tumor. a new diagnostic test. *Rev Surg* 1970;27:147–8.