Vascular leiomyoma presenting as medial joint line pain of the knee

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KEYWORDS

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Abstract Vascular leiomyoma or angioleiomyoma is a rare benign solitary smooth muscle tumor that occurs mostly in the extremities. Most of these tumors are composed of venous vessels, but in some reports small arteries have been detected in the tumors.

We present a rare case of a 60-year-old man with a subcutaneous vascular leiomyoma of the right knee. Clinical findings, ultrasound (US), magnetic resonance (MR), and histopathologic features are reported, and the literature is reviewed. The knowledge of specific imaging findings allows to include vascular leiomyoma in the differential diagnosis of lower extremity subcutaneous masses and to achieve an early accurate diagnosis.

Sommario Il leiomioma vascolare — o angioleiomioma — è un raro tumore benigno, solitario che si localizza soprattutto a livello delle estremità. Gran parte di questi tumori sono composti di vasi venosi, ma in alcuni reprot sono state trovate delle piccole arterie.

Presentiamo qui il caso di un uomo di 60 anni con un leiomioma vascolare sottocutaneo del ginocchio. Rivisitiamo qui la presentazione clinica, l'iconografia, le caratteristiche istopatologiche e la letteratura pertinente. La conoscenza delle caratteristiche specifiche di imaging permette di includere i leiomiomi vascolari nella diagnosi differenziale delle masse sottocutanee dolenti delle estremità e di ottenere una diagnosi precoce e mirata.

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Introduction

Vascular leiomyomas are rare small benign lesions originating from arterial smooth muscle cells or the venous tunica media with a predilection for the lower extremities

in middle-aged females [1,2]. Diagnosis is frequently delayed because of their rare occurrence.

Therefore, knowledge of the typical presentation can expedite diagnosis and lead to surgical resection which is the only way to relieve symptoms. We present the ultrasound (US) and magnetic resonance (MR) findings in a case

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of a 60-year-old man with a subcutaneous vascular leiomyoma of the right knee.

Case report

A 60-year-old healthy man presented with chronic pain in the right knee associated with swelling. The pain had been sporadic and vague at the onset, 1 year before presentation. He denied any trauma. The symptoms were generally mitigated by local rest; however he could not achieve full extension in the knee. In the last few months, the pain had become more intense and frequent. He described a burning, disabling pain over the medial face of the knee where he noted a small blue mark, associated with increasing swelling. The palpation of this well-localized area caused severe pain.

On examination the right knee revealed a loss of passive full extension (-10°) and a swelling of the whole knee. A small spot of bluish discoloration of the skin just distal to the medial joint line was visible; palpation caused pain and discomfort. The lesion was stiff–elastic and movable from the surrounding tissue. There was slight muscle atrophy in the affected thigh, but typical signs of meniscus lesions were not detected.

Plain orthogonal radiographs of the knee showed no bone abnormalities. US localized the lesion in the subcutaneous soft-tissue; it appeared oval-shaped and well-margined, mildly hypoechoic (Fig. 1), with peripheral and diffuse vascularization.

This lesion, measuring 35 mm in diameter, was clearly clivated from the adjacent tendons but strictly connected to a vessel. The patient subsequently underwent MR of the knee which confirmed the presence of the lesion. It appeared hyperintense in T2-weighted images and hypointense in T1 (Fig. 2) and showed marked gadolinium enhancement. The patient therefore underwent surgery.

Definitive treatment involved complete excision of the bluish-tan firm nodule. Histopathologic examination showed a benign lesion consisting of muscle tissue (confirmed by smooth muscle actin staining) penetrated by

Fig. 1 US: the lesion, localized in the subcutaneous soft-tissue, appears oval-shaped, well-margined, mildly hypoechoic.

Fig. 2 MR: the lesion appears hyperintense in T2-weighted images.

multiple small vessels, consistent with the diagnosis of vascular leiomyoma. Postoperatively, the patient had immediate and complete relief of symptoms.

One year after the operation no recurrence of the tumor has been detected.

The patient provided informed consent to the publication of this case description for the benefit of the scientific community.

Discussion

Vascular leiomyomas or angioleiomyomas are benign solitary tumors of smooth muscle origin that arise from the smooth muscle of blood vessels. Stout [1] published the first comprehensive review of this rare lesion in 1937, and it has been well characterized in the literature since that time. Although predominantly found in the lower extremities, several angioleiomyomas around the knee have also been reported. Hachisuga et al. [2] described the clinico-pathologic data of 562 cases of angioleiomyomas. The peak incidence is between the fourth and the sixth decade of life.

Angioleiomyomas commonly arise in the deep layers of the dermis or in the subcutaneous tissue. Histologically, these lesions are composed of varying amounts of smooth muscle bundles surrounding vascular channels contained within a thin fibrous capsule. Morimoto [3] described three subtypes: solid or capillary, cavernous, and venous. Each histological subtype has different features related to the location of the lesion and the gender of the patient. The solid form (most common) is three times as common in females and typically involves the lower extremities. In contrast, the cavernous subtype is four times as common in males and accounts for a majority of head and upper extremity lesions.

Ninety percent of these lesions occur in the extremities, with the head and trunk accounting for the remaining cases. The lower extremity is involved in 75% of the extremity lesions and is twice as common in females [4]. In contrast, head and upper extremity lesions are more frequent in males. Pain and/or tenderness are the most characteristic complaint in 60–75% of patients [3,4]. The solid histological subtype, mainly in the lower extremities, typically presents as a painful subcutaneous mass. Well-localized pain, exquisite tenderness, and temperature sensitivity are not uncommon findings. These are also the classic triad of symptoms for glomus tumors, and these two lesions can only be distinguished by histological evaluation. Along with glomus tumors, the differential diagnosis for small painful extremity lesions include hemangiomas, angiolipomas, ganglions, neurilemmomas, traumatic neuromas, and eccrine spiradenoma. There are no specific imaging techniques. However, as presented in our case, at US the lesion usually appears hypoechoic, clivated from the tendons, but MR can better delineate the lesion and define its relationship to the adjacent structures allowing a better pre-excisional planning [5]. Hwang et al. [6] found that the smooth muscle and numerous vessels within vascular leiomyomas corresponded to hyperintense areas on T2-weighted MR images. In addition, the fibrous capsule was identified as a well-defined peripheral hypointense area on T2-weighted MR.

Marked gadolinium enhancement, as in our case, is suggestive of the vascular origin of the lesion. The gross appearance of vascular leiomyomas is sharply demarcated

spherical tumors in approximately 80% of cases commonly less than 2 cm in size.

Sonographers should be familiar with the US features of this lesion. By including vascular leiomyomas in the differential diagnosis of painful lower extremity subcutaneous masses, an early accurate diagnosis is possible. Excision of the well-demarcated soft-tissue lesion enables a histopathologic diagnosis and offers complete resolution of symptoms as shown by the present case.

Conflict of interest statement

The authors have no conflict of interest.

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