

Clinical Quiz

Angioleiomyoma: an unusual cause of thigh pain

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Case

We report the case of 25 year old woman with a 7 year history of an increasingly painful thigh lump. Pain was triggered after gentle touch of the lateral side of her right thigh across iliotibial band and felt like an electric shock through her femur. On physical examination the thigh was found normal in shape compared with the other side. Overlying skin had normal appearance and mobility. No inguinal lymphadenopathy was found.

Radiographs of the involved area were non-significant, apart from the one taken 9 months prior to consultation, where a soft tissue lesion with peripheral and central calcification, without osseous continuity was evident (Figure 1A). This was confirmed on computed tomography scan (Figure 1B). Ultrasound scan of the mass revealed the lack of continuity with neighbouring structures and a homogenous, decreased echogenic texture.

An MRI scan of the right hip followed; the lump demonstrated intermediate signal intensity on T1 and high signal intensity on T2, with marked enhancement after iv administration of Gadolinium (Figure 1C). A three phase Tc99 bone scan and a Selective Digital Angiography of the right external inguinal artery followed. On the bone scan the first two phases were negative, while the third showed slight enhancement in the soft tissue of the right thigh, without indications of high vascular infiltration or bone involvement. On digital angiography, neither evidence of arterial involvement was found, nor an outflow vein.

Excisional biopsy was performed under general anaesthesia due to the benign features of the lesion, resulting

from the imaging workup. The mass was widely excised with the superficial vastus lateralis fascia attached. Histological examination revealed irregular calcification, fascicular cell arrangement, with fusiform nuclei and thin nucleolus, without atypia, but with degeneration. No mitoses were present. The neoplasm included several vessels, most with well-marked walls and was excised with clear margins (Figure 2A).

On immunohistochemical evaluation, SMA and caldesmon were positive, few cells were S-100 positive, while EMA, CD68, CD34, CD99, bcl2 and ki-67 index were negative (Figure 2B). So the immuno-morphological diagnosis was angioleiomyoma with degenerative changes. The patient was followed regularly at the outpatient clinic, with the last visit being 4 years after the excision. No local recurrence or distant metastases were noticed.

Commentary

Angioleiomyoma is a relatively uncommon benign subcutaneous soft tissue mass that most often occurs in the extremities. The term is angioleiomyoma, synonym with vascular leiomyoma and angiomyoma which was first described by Stout in 1937¹. It is a benign solitary soft tissue smooth muscle tumour, originating from the tunica media of vessels. Being a smooth muscle tumour, it can be found anywhere in the body. It frequently can also occur in extra-skeletal sites, visceral as the ovaries, uterus, bladder, lung, and the gastrointestinal tract². Approximately 50-70% of all the angioleiomyoma are found in the lower limbs.

Angioleiomyoma is a benign soft tissue tumour that arises from the smooth muscle layer of blood vessels. It can usually be found in the skin or subcutaneous tissue. Exceptional presentations include viscera and bone. The aetiopathology of this lesion is yet unknown and frequently it is asymptomatic. Possible causative factors include local traumatic events, infections, hormonal imbalance and vascular malformations³. Pain is not pathognomonic of an angioleiomyoma nevertheless approximately 60% of patients are symptomatic. Pain severity can vary

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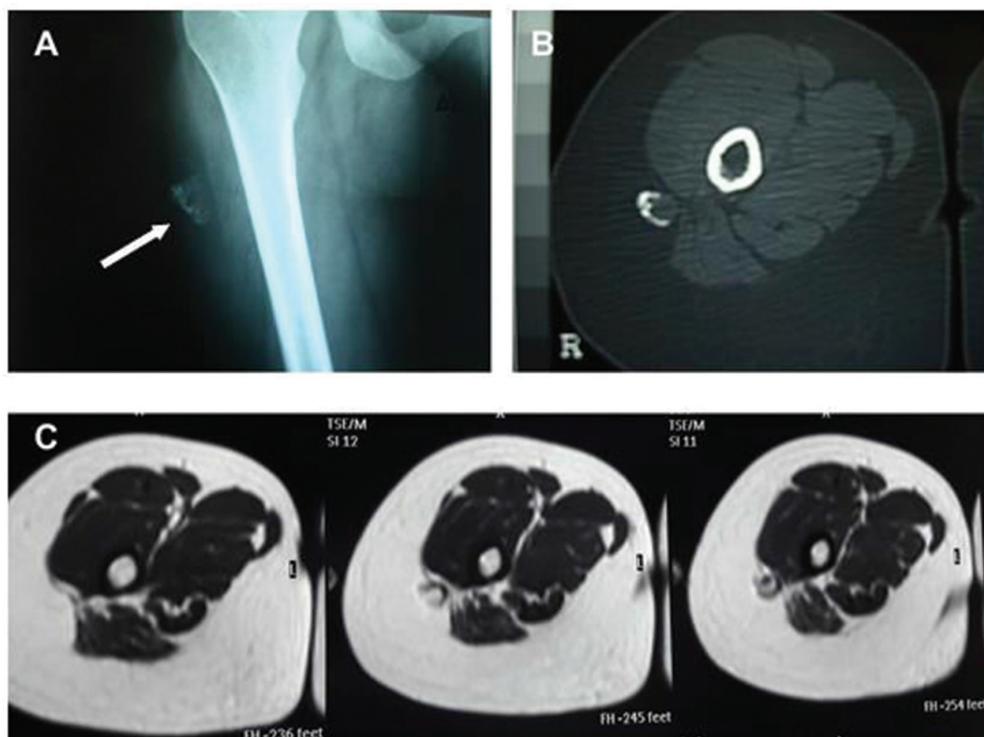


Figure 1. A. Peripheral and central calcification (arrow) lesion, located in the soft tissues, without osseous continuity. B. Computed tomography scan, according to which, the mass was situated in the adipose tissue, without edema or infiltration of regional musculature. C. MRI scan of right upper thigh. T1 sequence transverse section, lump with intermediate signal in adipose tissue.

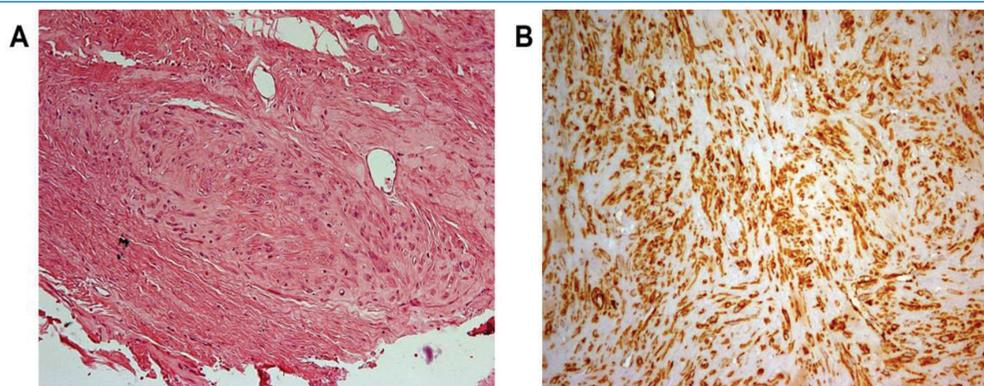


Figure 2. A. Neoplasm with marginally bundled configuration. Neoplastic cells with ovoid and spindle - shaped anisochromatic nuclei. The neoplasm encloses several vessels. B. Positive SMA (smooth muscle actin) immunohistochemic stain.

depending on ambient temperature or compression of local neurovascular structures⁴. The cardinal clinical sign of an angioleiomyoma is paroxysmal pain, initiated by the slightest touch or exposure to environmental stimuli. A histological explanation could be given by the presence of nerves within the capsule and interstitial of angioleiomyoma. Pain in solid type occurs at the time of vessel contraction in the tumour,

which may cause ischemia. Pain is triggered by pressure, cold and hormonal changes⁵.

In this paper we will consider the typical presentation, which is a solitary, painful subcutaneous nodule, with characteristic sharp pain. The overall incidence of angioleiomyoma is hard to be estimated, since most benign lesions remain undetected. Main reasons for excision are either pain or a palpable mass.

Usual localizations for subcutaneous angioleiomyoma are the extremities [89,7%], the head and neck [8,5%] and the trunk [2,5%]. It commonly occurs in the 40-60 age group with patients having a mean age of 52 years, with female predominance (male to female ratio varying between 1: 1,7 and 1: 2,2).

The diagnostic workup comprises of ultrasound scan and magnetic resonance imaging. Ultrasound scan is often used as initial evaluation of soft tissue nodules, because it describes their localization, size and relation to surrounding structures. Characteristic features of angioleiomyomas are the lack of continuity with neighbouring structures, the relatively homogenous - decreased echoic texture with small amounts of post acoustic enhancement and the increased vascularity on color Doppler. On MRI the appearance seems to be characteristic irrespective of the tissue the lesion is embedded. On T1 sequence we have a well defined area with decreased signal intensity. On T2 sequence we have marked hyperintensity. When situated in bone, the lesion has sclerotic margins while in soft tissue it is encapsulated. Excisional biopsy is usually curative. Differential diagnosis includes lipoma, fibroma, ganglionic cyst, glomus tumour, neurofibroma, neurinoma and leiomyosarcoma.

Macroscopically it presents as an approximately well encapsulated solitary module grey/white in color or brown. Microscopically 3 types can be distinguished: 1) The solid type, consisting of dense smooth muscle bundles, compacted and intersecting one another surrounding small slit like vascular channels. Male to female ratio is 1:3. It occurs in 66% of cases of angioleiomyoma. It is usually painful and mean time to excision from diagnosis is 4 years. It is typically located in the lower extremities of females. 2) The cavernous type containing dilated vascular channels with smaller amounts of smooth muscle. The muscle walls of vessels are difficult to distinguish from intravascular smooth muscle bundles. Male to female ratio is 4:1 and occurs in 11% of cases. Mid time to excision from diagnosis is 5 years and it typically occurs in the upper extremities of men. 3) The vein type, where venous

type channels with thick muscle walls and without compacted muscle bundles are found. The vascular walls are easily distinguished from intervacular smooth muscle bundles. Mid time to excision from diagnosis is 7 years. It is commonly painless and situated in the lower extremities.

Histological signs of the lesion being benign are the fact that it is an encapsulated module that it rarely invades surrounding structures and that few cases of recurrence have been reported. Histological differential includes angiomyolipoma, leiomyosarcoma and pillar leiomyoma, which tends to present with multiple lesions. On immunohistochemical analysis angioleiomyomas test positive for muscle specific actin, desmin, collagen type IV and smooth muscle actin and are negative for HMB45. As previously mentioned the capsule commonly contains nerves, which are positive for PGP 95 and S-100.

Excisional biopsy will suffice in most cases. Symptomatic lesions are more likely to be operated⁴. In the rare case of recurrence, a wide local re-excision should be performed, which is common in cases of low grade malignancy⁵.

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Questions

1. Which is the main differential diagnosis for angioleiomyoma?

- A. Fibroma
- B. Glomus tumor
- C. Angiomyolipoma
- D. Lipoma

Critique

The exact diagnosis is set after excisional biopsy. Groups of mature adipose cells were demonstrated in

certain percentage in angioleiomyoma, differing from angiomyolipoma. The differences between those two entities are clinical, histopathological, involving the adipose tissue content (2,5% to 3% for angioleiomyoma and 20% to 30% for angiomyolipoma) and immunohistological – angiomyolipoma is positive for HMB 45. Furthermore, cutaneous epithelioid angioleiomyoma could represent the benign counterpart to the epithelioid leiomyosarcoma of the skin, or could be considered a variant of leiomyoma. The correct answer is C

2. What is the indicated treatment method for symptomatic musculoskeletal angioleiomyoma?

- A. Observation
- B. Simple excision
- C. Wide excision
- D. Wide excision and external beam irradiation

Critique

Angioleiomyomas usually present in the extremities as a solitary painful subcutaneous nodule. The main reasons for excision are either pain or casuistic considerations. Simple excision is adequate due to the benign nature of the lesion, lack of continuity with neighbouring structures, small rate of recurrence and malignant transformation. Wide excision with or without external beam irradiation are reserved for malignant soft tissue tumors.

The correct answer is B