

Intravascular Myopericytoma in the Heel: Case Report and Literature Review

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Abstract: Intravascular myopericytoma (IVMP), regarded as a variant of myopericytoma, is a rare tumor. Very few cases have been described, none in the foot.

The first case of IVMP located in the heel of the foot is described in this article. A literature review is reported of all cases of IVMP published in the English literature.

A 48-year-old man possessed an IVMP on the heel of the right foot. The physical examination and histopathological and ultrasound studies are described. The literature review yielded 5 cases of IVMP, 2 of which were in the thigh and 1 each in the oral mucosa, the periorbital region, and the leg.

The possibility that these lesions may be malignant suggests that the histopathological study of vascular tumors should include immunohistochemical tests.

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Abbreviations: H&E = hematoxylin and eosin, IVMP = intravascular myopericytoma, SMA = smooth muscle actin.

INTRODUCTION

Myopericytoma, first described by Granter et al in 1998,¹ is a rare tumor typically located in the dermis and superficial soft tissues of the lower limbs, but it may also occur in other locations such as the upper limbs, head, and neck.² It commonly affects men of between 30 and 40 years of age, although cases have also been reported in children. The tumor manifests itself in the form of single lesion, although multiple forms have been described.³ The tumors are usually benign, but they may recur, and malignant cases have been described.⁴ It shares morphological characteristics with angioliomyoma, angioliopoma, myofibroma, and glomus tumor, but is assumed to represent a specific perivascular myoid cell (myopericyte) neoplasm.¹ The term myopericyte was coined by Dictor et al⁵ to

describe what they considered a transitional cell between pericytes and smooth muscle cells of blood vessels. In 1996, Requena et al⁶ suggested that some adult cutaneous myofibromas were actually benign perivascular neoplasms composed of cells with morphological, immunohistochemical, and ultrastructural characteristics of immature pericytes or myopericytes. Later, in 1998, Granter et al¹ used the terms “myopericytoma” or “perivascular myoma” to refer to these neoplasms. The term “myopericytoma” was officially recognized by the World Health Organization in 2002.⁷

Intravascular myopericytoma (IVMP) is an extremely rare tumor. Only a very small number of cases have been reported, the first description being that of McMenamin and Calonje in 2002,⁸ as a “long-standing myopericytoma that was entirely located within the lumen of a subcutaneous vein.”

Here we present a case of IVMP located in the heel of the right foot and report a literature review of all cases of IVMP described in the English literature.

Case Report

A 48-year-old man was treated by 3 different professionals over the preceding 15 months period for a papillomatous or warty lesion on the outer plantar side of the heel of the right foot, with topical application of acid (salicylic and nitric acids), cantharidin, and liquid nitrogen. There was nothing of relevance in his medical history and no record of previous trauma to the limb. Physical examination showed a minor, dry, discreetly crusted, epidermal lesion with no clinical signs of papilloma. However, palpation revealed a consistent, movable mass that was painful on compression and lateral pinching (Figure 1).

Static biomechanical examination of the feet showed pathological pronation of the subtalar joint. This was confirmed by the dynamic examination, together with displacement and torsion of the plantar heel fat.

Ultrasound showed a well-defined lenticular, homogeneous, hypoechoic nodule of lobulate appearance, about 12 × 12 × 12 mm, at a depth of about 7 mm, at a superficial level in the fat pad (Figure 2). The patient had previously received conservative treatments (orthotics and physical therapy) that did little to alleviate the pain so the possibility of surgery was raised.

Surgery, under local anesthetic block and tourniquet control, consisted an incision made on the lateral zone of the heel, in the dorsal skin (Figure 3), and with a careful dissection of soft tissue, through which the tumor was extracted in its entirety and the wound was closed in a single plane. This surgical approach allowed the patient to resume restricted ambulation for 48 hours and almost normal ambulation thereafter. The pain disappeared completely soon thereafter, and at 12 months postsurgery, there was no recurrence of the lesion.

The resected specimen measured 1.5 × 0.4 cm. Macroscopically, its appearance showed an expansion of the dermis

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FIGURE 1. Appearance of the lesion. Exploration and pinching the tumor.

at the expense of a whitish–pink tissue. Histopathology showed a lesion in the subcutaneous tissue, which had a venous-type vascular structure, was dilated, and was partially occupied by a proliferation of spindle-shaped cells exhibiting a “hemangiopericytomatous” focal pattern. The presence of vessels of flattened endothelium surrounded by a concentric arrangement of rounded or slightly spindle-shaped cells was evident. No atypia was observed, and neither mitosis nor necrosis. Immunohistochemistry showed these cells to express smooth muscle actin (SMA) and caldesmon, but not CD34 antibody, desmin, factor VIII, epithelial membrane antigen, or cytokeratins (Figure 4). The resection margins were not compromised by the lesion and the histopathological diagnosis is “biopsy of 1 right heel with IVMP.”

MATERIAL AND METHODS

We performed a literature search in PubMed/Medline covering January 2002 to October 2014, using the key term “intravascular myopericytoma.” This yielded 11 results, of which only 5 corresponded strictly to IVMP. We excluded the term “perivascular myopericytoma” and 2 cases were specifically excluded: one of intramural myopericytoma⁹ and the other of digital artery IVMP.¹⁰ The reason for exclusions was that they did not conform to the histopathological and immunohistochemical characteristics described for IVMP.

This research followed the principles and standards of the Declaration of Helsinki of 1975, last revised in Seoul in 2008.

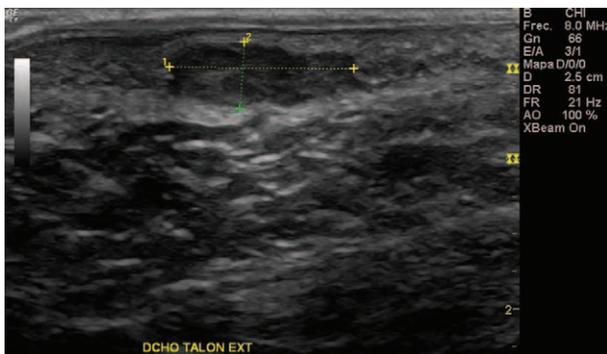


FIGURE 2. Ultrasound showing a lenticular, homogeneous, and hypoechoic nodule.



FIGURE 3. Appearance of the resected tumor.

They have respected the principles of confidentiality and patient autonomy.

In this case, ethical approval by an ethics committee was not necessary because it is a necessary surgical treatment for the patient. He was not included in any control group and no experimental treatment was performed. The patient was informed of his injury, diagnosis, and treatment. Informed consent was given.

RESULTS

We found only 5 cases of IVMP; 2 were located in the thigh, and 1 each in the oral mucosa, the leg, and the periorbital region. The present case is therefore the first reported case of IVMP in the foot. The evolution of the literature cases ranged from 5 to 15 years. The age of the patients (3 men and 2 women) ranged from 45 to 79 years, with a mean of 59.8 years (Table 1).

DISCUSSION

There are some clinical differences between IVMP and other myopericytomas. Although myopericytoma, in general, manifests as lesions that may or may not be painful, all of the IVMP cases reviewed presented with painful symptoms. According to McMenamin and Calonje,⁸ this is probably caused by the thrombus associated with the lesion.

Some authors have argued for the trauma/microtrauma etiology of some tumors including myopericytoma.² Regarding IVMP, Xia et al¹² report a case in which the patient had suffered

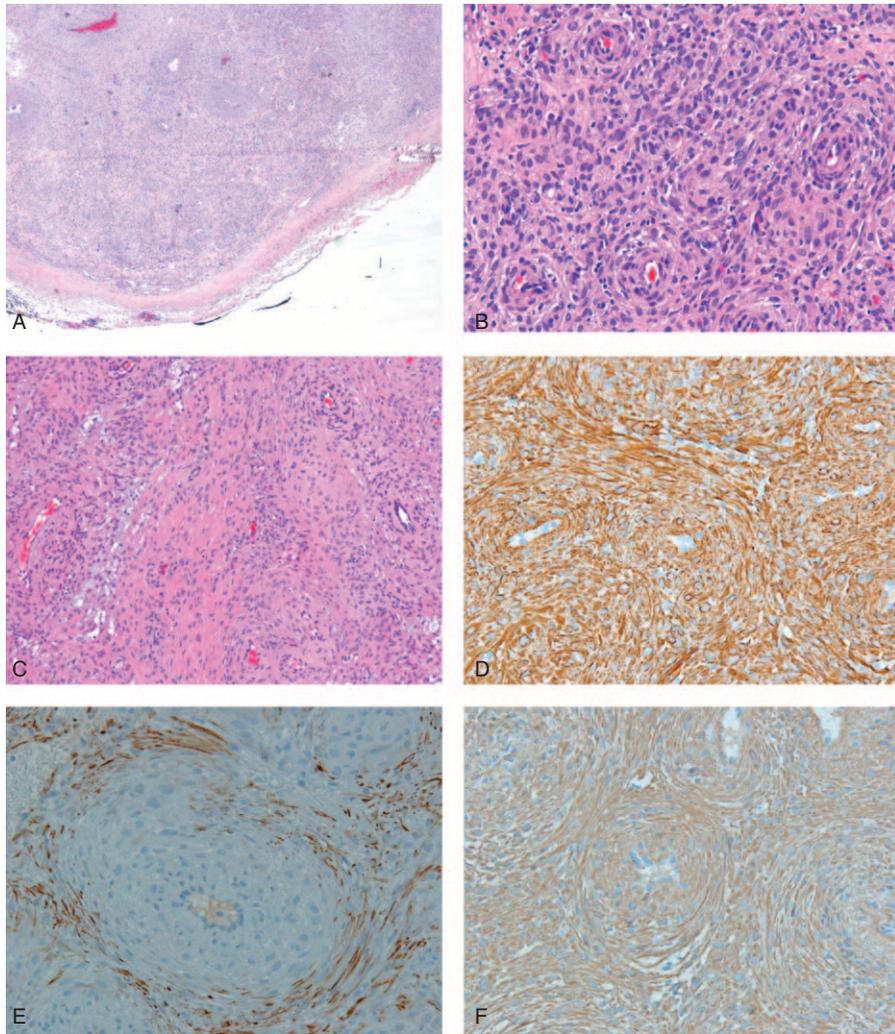


FIGURE 4. Overview of the intravascular lesion. (A) Hematoxylin and eosin (H&E) $\times 5$. Proliferation of spindle-shaped myopericytic cells with a characteristic concentric distribution—(B) H&E $\times 20$; (C) H&E $\times 10$. The myopericytic cells express caldesmon (D), are negative for desmin (D, E), and express smooth muscle actin (F).

a blow 5 years before with a sharp object in the area where the tumor later appeared. McMenamín and Calonje⁸ refer to a subcutaneous nodule that evolved slowly over the course of the 10 years before the onset of IVMP. However, Ide et al¹¹ refer to a patient who had undergone surgery for a fatty tumor in the same region 9 years before, but they did not believe that this had

any direct relationship with the IVMP. Ko et al¹⁴ describe a patient who, for 15 years, had a slowly growing mass in the same area in which the IVMP appeared. In the present case, the torque exerted and the consequent shearing of the soft tissues caused by the pronated subtalar joint could have been 1 of the causes of the pain and perhaps of the trauma/microtrauma

TABLE 1. Cases of IVMP

Authors/Year	Age/Sex	Etiology/Hypothesis	Evolution, y	Site
McMenamin and Calonje (2002) ⁸	54/Male	Previous tumor	10	Thigh
Ide et al (2007) ¹¹	45/Female	Unknown	9	Oral mucosa
Xia et al (2009) ¹²	54/Male	Trauma	5	Leg
Park et al (2010) ¹³	79/Female	Unknown	5	Infraorbital area
Ko et al (2011) ¹⁴	67/Male	Previous tumor	15	Thigh

IVMP = intravascular myopericytoma.

etiology of the tumor itself, as is the case with other similar lesions.

Myopericytomas affecting viscera have been described that seem to be related to the Epstein–Barr virus.^{15,16} In the present case, the history of the wart or plantar papilloma for which the patient was treated, although it was not confirmed by anything except the clinical aspect, could mean that either the infection itself or the aggressive treatment with acids or caustics may have influenced the origin and development of the IVMP.

Myopericytoma consists of a well-circumscribed, but not encapsulated, mesenchymal proliferation comprising areas of moderate cellularity with fusiform cells showing several layers of concentric growth around small vessels interspersed with other areas of abundant vascular slits of medium size and a hemangiopericytoid pattern. Very few mitotic figures are observed, and there is no necrosis. Immunohistochemistry of the lesion is positive for muscle-specific actin, SMA, h-caldesmon, and focally for desmin, but is negative for S-100 protein, CD31 antibody, CD34 antibody, factor VIII, human melanoma black 45, and cytokeratin AE1–AE3. Unlike myopericytoma, however, IVMP (as in the present case) resides completely within the lumen of the subcutaneous vein and is not positive for desmin (although McMenamin and Calonje⁸ reported that the muscular wall of the vein in which the tumor is found stained positively for desmin and SMA).

It is therefore suggested that there is a need to establish a differential histopathological diagnosis with other tumors and tumor-like lesions with which it shares clinical and histopathological features, such as perivascular myoma,^{17,18} angioleiomyoma,¹⁹ hemangiopericytoma,⁵ myofibroma and myofibromatosis,^{5,6,20,21} papillary endothelial hyperplasia,^{22,23} angiosarcoma,^{24,25} lobular capillary hemangioma (especially in its intravenous forms),²⁶ and intravenous glomus tumor,^{27,28} among others.

Diagnostic imaging does not show the vascular nature of IVMP, although both magnetic resonance imaging¹¹ and ultrasonography are useful for morphological assessment of the tumor and its location, and for designing the best surgical procedure. In the present case, the information provided by ultrasonography enabled a surgical approach to be taken that allowed outpatient postoperative recovery.

It is, however, likely that this particular IVMP is not the first example in the foot because others may have been diagnosed by histopathology alone without immunohistochemical tests. McMenamin and Calonje⁸ suggested that the case they presented was actually the second, with the first having been interpreted as an intravascular angioleiomyoma.²⁹

Recently, in October 2014, Kara et al³⁰ described “thin-walled vessels with a concentric, perivascular gathering of spindled myoid tumor cells” in hand. Furthermore, the possibility that these lesions may be malignant^{4,5} suggests that the histopathological study of vascular tumors should include immunohistochemical tests.

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