Unusual presentation of intravascular papillary endothelial hyperplasia (Masson's tumor)

Marco Breccia MD\textsuperscript{a}, Mariangela Novello MD\textsuperscript{b}, Marco Galli MD\textsuperscript{a}, Antonella Coli\textsuperscript{b,*}

\textsuperscript{a}Department of Geriatrics, Neurosciences and Orthopedics, Catholic University, Rome, Italy
\textsuperscript{b}Institute of Anatomic Pathology, Catholic University, Rome, Italy

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Abstract

Intravascular papillary endothelial hyperplasia is a rare, exuberant form of reactive endothelial proliferation which can mimic benign and malignant vascular tumors. In this report, we describe a 22-year-old man presenting with a 2.5 cm nodule on his left foot, near the first metatarsal head. The patient underwent total excision of the lesion, with subsequent histological diagnosis of intravascular papillary endothelial hyperplasia arising within a thrombosed periosteal vein. After nine months from surgery, the patient is alive and well, with no evidence of local recurrence.

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1. Introduction

Intravascular papillary endothelial hyperplasia, firstly described by Masson in 1923 as “hémangioendothéliome végétant intra-vasculaire” in hemorrhoidal vessels [1], is a rare intravascular benign lesion reported in various locations, most frequently occurring in the skin and subcutaneous soft tissues of the head, neck, finger and trunk as a small, red to blue superficial mass [2,3]. The foot is rarely affected [2–11]. This lesion, which histologically mimics a vascular neoplasm, is commonly found in strict association with thrombotic material, and is currently considered as an organization process of a thrombus with endothelial proliferation, rather than a true intravascular tumor [3]. The histological examination easily discloses the reparative nature of the process, although cases have been reported in which a differential diagnosis with a vascular, possibly malignant tumor is mandatory, due to an unusual exuberant proliferation of endothelial cells.

2. Case report

2.1. Clinical history

A 22-year-old Caucasian male presented with a palpable, subcutaneous mass located in the first metatarsal head of the left foot. The patient reported the appearance of a painless swelling of 8 months’ duration, without any previous trauma. Physical examination revealed a roundish subcutaneous nodule, about 3 × 2.5 cm, located on the dorsal forefoot, at the first metatarsal head, medially to the extensor hallucis longus tendon. The lesion was mobile, painless and mildly...
tender to palpation, with no alterations of the overlying skin. Blood laboratory tests were normal. Biplanar radiographs showed no gross involvement of the tubular bones; only a thin sclerotic periosteal reaction being observed on the medial cortex of the first metatarsal head, near the soft tissue mass. Ultrasound scan revealed a solid, capsulated, elongated lesion measuring 2.7 × 2.2 × 0.9 cm, which was homogeneously hypoechoic with intrallesional vascular signal, and magnetic resonance imaging (MRI) showed a well-defined, ovoid mass in the subcutaneous area, with well-defined margins, homogeneously isointense on T1-weighted images and heterogeneously hyperintense on T2-weighted images (Fig. 1A). Owing to the clinical suggestion of a benign lesion, an excisional biopsy was performed, with complete removal of the lesion (Fig. 1B). Nine months from surgery, no evidence of local recurrence was observed.

2.2. Histology and immunohistochemistry

The tissue biopsy was fixed in 10% buffered formalin and routinely processed for paraffin embedding. Four micrometer sections were stained with hematoxylin and eosin, Weigert’s elastic van Gieson and Masson’s trichrome. Additional immunohistochemical analyses were performed by the Dako AutostainerLink 48 (Dako, Carpinteria, CA, USA), using the following primary antibodies: CD31 (Dako, clone JC70A, 1:50), CD34 (UCS Diagnostic, Rome, Italy, clone QBEND/10, 1:100), FLI1 (UCS

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**Fig. 1** Intravascular papillary endothelial hyperplasia in a 22-year-old man. A, A T2-weighted magnetic resonance imaging demonstrates a hyperintense mass on the left dorsal forefoot, at the first metatarsal head. B, The sectioned surface of the mass reveals a well-defined, intraluminal pinkish-red almost solid lesion with thrombotic areas. C, Photomicrograph showing endothelium-lined vascular spaces on the left, with transition to densely-packed papillary endothelial proliferation on the right (H&E, original magnification ×100). D, At higher magnification, note the small papillary structures supported by delicate hyaline core and lined by a single layer of hyperplastic endothelial cells (H&E, original magnification ×200). The endothelial cells show positive cytoplasmic reactivity for CD31 (E) and nuclear reactivity for FLI-1 (F).
Diagnostic, polyclonal, 1:50), smooth muscle actin (SMA, Dako, 1:50) and desmin (Dako, 1:100). For CD31 and FLI1 antibodies, heat-induced epitope retrieval was performed using the PT Link treatment (Dako).

3. Pathological findings

Macroscopically, the specimen consisted of a red-bluish and well circumscribed mass, measuring 2.5 cm in greatest diameter. Histologically, the nodule, which appeared attached to the wall of a dilated vascular space, showed a central area consisting of thrombotic material surrounded by a florid proliferation of densely packed, small papillary structures with a hyaline core, lined by plump endothelial cells apparently floating in a lumen (Fig. 1C and D). We also detected solid areas composed of clusters of hyperplastic endothelial cells with prominent nuclei, in which the vascular nature was not promptly apparent. Occasional mitoses, but not atypia, were found; tissue necrosis was absent. At the periphery, a collagenous tissue was present with features of organizing thrombus, intersected by numerous, irregular anastomosing vascular spaces with a thin wall and showing transition to densely packed papillary endothelial proliferation. Elastic van Gieson and Masson’s trichrome stains confirmed that the lesion was entirely confined within a large venous vessel. Immunohistochemically, the endothelial cells showed cytoplasmic reactivity for CD31 (Fig. 1E) and CD34, and nuclear reactivity for FLI1 (Fig. 1F). Pericytic cells, lining the outer surface of endothelial cells, were stained by SMA and, with a less extension, by desmin. Although the histological features raised the possibility of a proliferative vascular neoplasm, the presence of an entirely intravascular lesion without signs of extension in the surrounding tissues and, lastly, the absence of true features of malignancy suggested the final histological diagnosis of an intravascular papillary endothelial hyperplasia arising in a dilated, thrombosed vein.

4. Discussion

Intravascular papillary endothelial hyperplasia (also known as Masson’s tumor or Masson’s hemangioma) is an exuberant proliferative process occurring in previously normal, dilated vessels (de novo or “pure” form), generally in the distal extremities, or in preexisting vascular lesions (“mixed” form), as hemorrhoids, varices, hematomas, pyogenic granulomas and hemangiomas [2,3]. In 1923 Pierre Masson first described this lesion, and named it “hémangioendothéliome végétant intra-vasculaire” [1]. The lesion observed by Masson presented as a rapidly growing and painful mass within hemorrhoids of a 68-year-old man. In his work, Masson illustrates, also by fine drawings, an intravascular florid hyperplasia of endothelial cells with a fibrinous background, forming packed vegetations. He considered this entity, clinically suspected for a malignancy, as a benign vascular tumor of endothelial nature [1]. Actually, intravascular papillary endothelial hyperplasia is considered a very unusual form of exuberant thrombus organization. This lesion is commonly intravascular, although in markedly dilated vessels, smooth muscle or elastic tissue of the preexisting vessel wall is barely demonstrable [4]. Rarely, this process may extend into surrounding soft tissues following rupture of the vessel of origin; nevertheless, the intravascular location of the lesion is usually yet demonstrable in most cases [3].

The characteristic morphological appearance of this peculiar benign process is the presence of small papillary structures supported by delicate stromal stalks and lined by a single layer of hyperplastic endothelial cells projecting into a vascular lumen. The endothelial cells appear plump or swollen, thus simulating the growth pattern of a vascular tumor and sometimes suggesting malignancy, but usually lack pleomorphism and mitotic activity. Histological studies reviewing cases recorded as vascular neoplasms or tumor-like lesions show that intravascular papillary endothelial hyperplasia may also be confused with neoplastic processes, not rarely with a malignant tumor, particularly if the pathologist is unaware of this entity [2,4,9]. Indeed, a clear distinction between the small papillary structures with a hyaline core, lined by plump endothelial cells of this reactive disease and the tuft-like structures of an angiosarcoma may sometimes be difficult, especially in fine needle biopsies [12]. Furthermore, the plump, hyperchromatic endothelial cells of the angiosarcoma often pile up along the lumens, creating the pattern of papillary growth also observed in papillary endothelial hyperplasia [3]. The intravascular location, the presence of fibrin and features of organizing thrombus, the lack of true atypias and necrosis, and the hyaline core of the papillae are useful diagnostic findings.

The peculiarity of this case rests on the very large size of the lesion, the extremely florid endothelial proliferation also showing solid areas and the distinctive location. In fact, intravascular papillary endothelial hyperplasia has been rarely described in the foot, with 14 reported cases in the literature [2–11]. In most of these reported cases the lesion occurred as small nodules in the subcutaneous soft tissue of the planter region [4–6,8,10,11], while in the present case the lesion was located on the dorsal forefoot, near the periosteum of the first metatarsal head. Also, the large size of this benign process was an unusual feature.

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References