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# Intravascular Papillary Endothelial Hyperplasia (Masson's Hemangioma) Presenting as a Soft-tissue Sarcoma

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**Abstract.** *Intravascular papillary endothelial hyperplasia (Masson's hemangioma) is an unusual, benign, non-neoplastic, vascular lesion characterized histologically by papillary fronds lined by proliferating endothelium. It may appear as a primary or pure form developing in a distended vessel, or it can be associated with hemangiomas, pyogenic granulomas, or lymphagiomas. Nearly all lesions are intimately associated with a thrombus in various stages of organization. The main significance of intravascular papillary endothelial hyperplasia is its clinical and histological resemblance to soft-tissue sarcoma and possible misinterpretation as such. A case of intravascular papillary endothelial hyperplasia clinically diagnosed and treated as a low-grade angiosarcoma, in a 60-year-old man, presenting with a mass in the left thigh, is reported.*

First named "Hemangioendotheliome vegetant intravasculaire" by Masson (1) in 1923, Masson's hemangioma is an exuberant endothelial proliferation that requires differential diagnosis from angiosarcoma. Masson described it to be a form of neoplasm and explained the pathogenesis as proliferation of endothelial cells into the vessel lumen, followed by obstruction and secondary degeneration and necrosis. On the other hand, Henschen (2) depicted the lesion as a reactive process rather than a neoplasm. Kauffman and Stout (3) remarked that, although endothelial proliferation that can be easily mistaken for a characteristic of sarcoma is present, the endothelial layer of the lesion is composed of normal endothelial cells, the endothelial proliferation is of benign papillae pattern and the cells show no atypia. Today, it is considered to be a reactive vascular proliferation following traumatic vascular stasis.

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We describe a 60-year-old Caucasian male who underwent wide resection of a presumptive soft-tissue sarcoma of the thigh, with subsequent histopathological documentation of a benign intravascular papillary endothelial hyperplasia.

## Case Report

A 60-year-old Caucasian man was admitted to our service with a diagnosis of a suspicious mass in the lower third of his left thigh. The patient had no remarkable past medical history and his overall health condition was normal. Physical examination revealed a firm, smooth, rounded and slightly painful mass on the lateral surface of his left thigh, densely adhering to the underlying musculature, without any palpable adenopathy. The patient described a slow growing pattern of the mass over the previous 2 years. No previous injury was reported.

Initial laboratory workup was found to be within normal limits. Computerized tomography (CT) scan of the thigh showed a solid mass, measuring 6x4cm, extending into the overlying soft tissue and in close proximity to the femoral periosteum. Magnetic resonance imaging (MRI) scan disclosed a 5.5x3.5x3.5cm lobulated, heterogeneous, contrast enhancing soft-tissue mass involving the lateral head of the quadriceps muscle with nodular areas of increased density in delayed images (Figure 1). Both studies suggested the diagnosis of a soft-tissue sarcoma arising from the thigh musculature.

Preoperative fine-needle and core-needle aspiration were clinically significant for blood, fibrin and vascular channels, consistent with a vascular lesion, although no malignant cytology was confirmed.

A presumptive diagnosis of a low-grade angiosarcoma was made, and the patient underwent a radical, *en bloc* resection of the tumor, together with the lateral head of the left quadriceps muscle, including part of the underlying femoral periosteum.

Gross evaluation of the resected specimen revealed an expanding, fusiform, well-circumscribed mass, 5x3.5x3.5cm

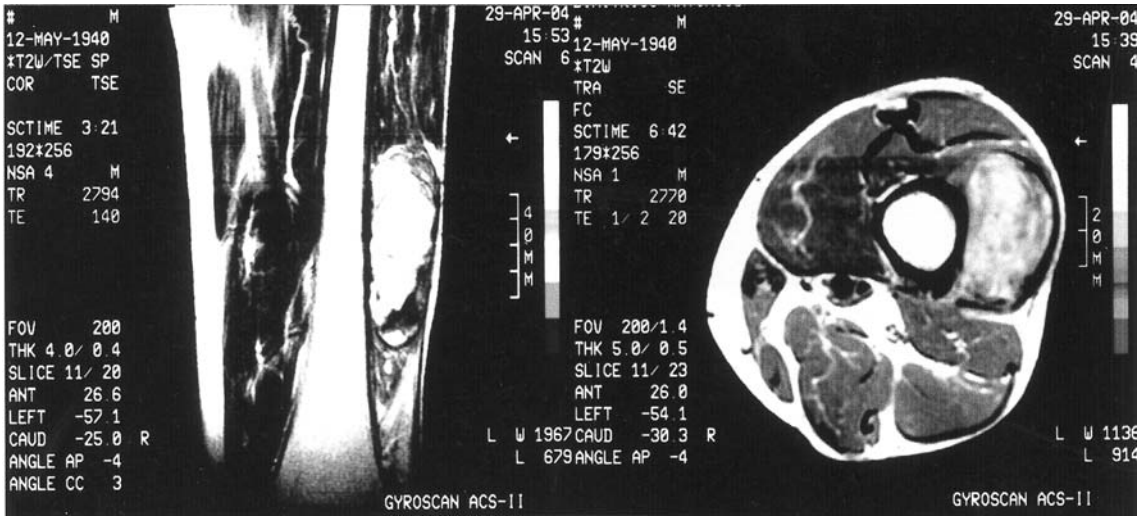


Figure 1. *MRI scan revealed a 5.5x3.5x3.5cm lobulated, heterogeneous, contrast-enhancing soft-tissue mass involving the lateral head of the quadriceps muscle with nodular areas of increased density.*

in size. Cut sections demonstrated a fibrous capsule surrounding locally hemorrhagic, tan friable material. Microscopic examination showed a pseudocapsule consisting of hyalinized, hypocellular, fibrous tissue. The cavity was composed predominantly of necrotic debris, fibrin and red blood cells. Cellular areas demonstrated endothelial cell proliferation, along with fibrin thrombi forming papillary fronds and vascular channels consistent with organizing thrombus with recanalization (Figure 2). The endothelium did not reveal any nuclear pleiomorphism, mitotic activity or significant necrosis (Figure 3). These findings were consistent with the diagnosis of intravascular papillary endothelial hyperplasia (Masson's disease) developed upon an underlying intramuscular hemangioma.

The patient tolerated the procedure well and had an uneventful course. He was discharged home on the seventh postoperative day and has had no signs or symptoms of locoregional recurrence for the last 15 months.

## Discussion

Intravascular papillary endothelial hyperplasia (IPEH) is a peculiar intravascular process that bears a remarkable resemblance to a hemangiosarcoma. In 1923, Pierre Masson (1) first described an intravascular papillary proliferation, formed within the lumen of inflamed hemorrhoidal veins in a 68-year-old man. He termed the lesion "Hemangio-endotheliome vegetant intravasculaire". He regarded it as the result of primary endothelial cell proliferation and remarked that, in its late stage, it could be confused with thrombi undergoing organization. He compared the architectural arrangement of the lesion to that of a mammary intraductal papilloma. Masson believed the lesion to result in the obliteration of the vascular lumen, causing degeneration and necrosis in the manner of secondary red infarct. Masson's observations were followed by the reports of Chagas (4) and Henschen (2). The latter, in 1932, portrayed a similar endothelial proliferation, but he regarded it more as a reactive process than an endothelial neoplasm. The lesion came to be called "L' endovasculite proliferante thrombopoietique", or Masson's intravascular vegetant hemangioendothelioma (5). Henschen described it in a variety of organs and lesions, such as the veins of the pelvic plexus, polyps of the nasal cavity, larynx, digestive tract, uterus, even in the cavernous hemangiomas of the liver or the extremities.

Dupont *et al.* (6) reported, in 1964, the case of a 10-year-old girl affected with multiple cutaneous angiomas and skeletal endochondromas in which the two cutaneous hemangiomas that were excised showed a marked papillary proliferation of the vascular endothelium. It seems clear from their description that they were describing focal Masson's pseudoangiosarcomatous changes in cavernous hemangiomas, similar to those seen in our report.

Through numerous further studies, researchers concluded that this lesion is a vascular proliferative reaction following traumatic vascular stasis, rather than a true neoplasm. This hypothesis formed a quick consensus and the term intravascular papillary endothelial hyperplasia (IPEH), first coined by Clearkin and Enzinger (7) in 1976, is most widely used instead of the misleading definition of Masson's pseudoangiosarcoma.

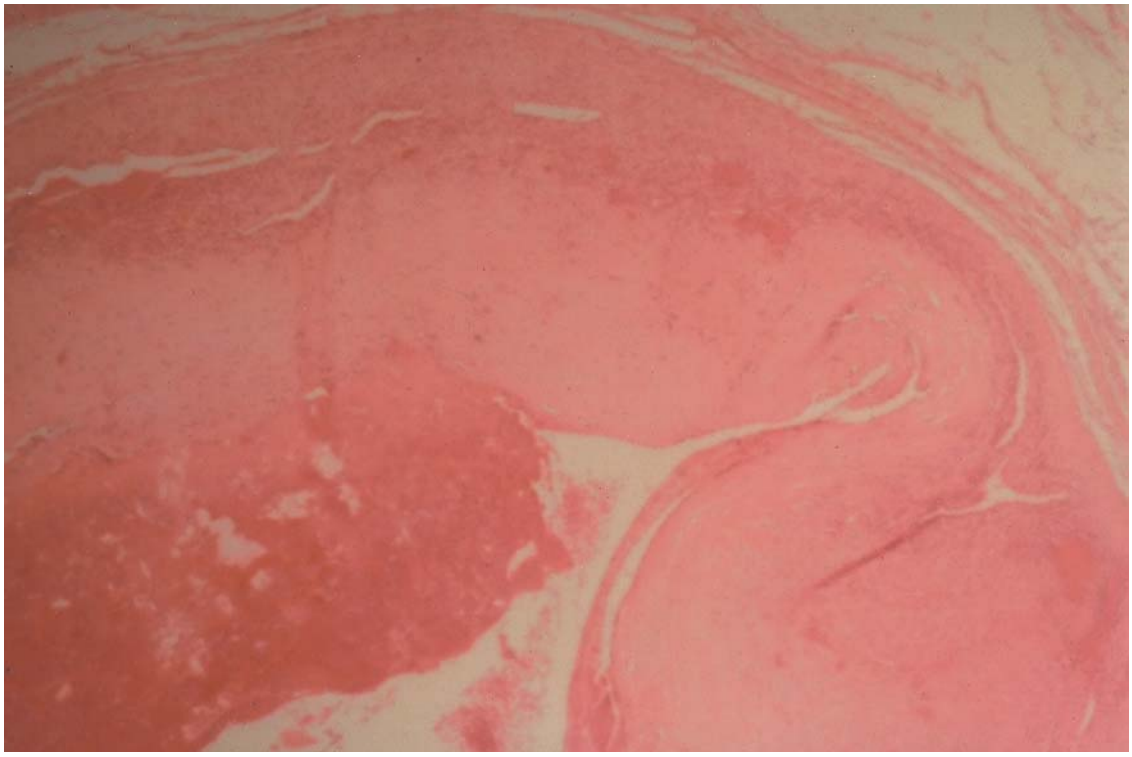

Figure 2. *Microscopic examination showed cellular areas with endothelial cell proliferation around organizing thrombus.*

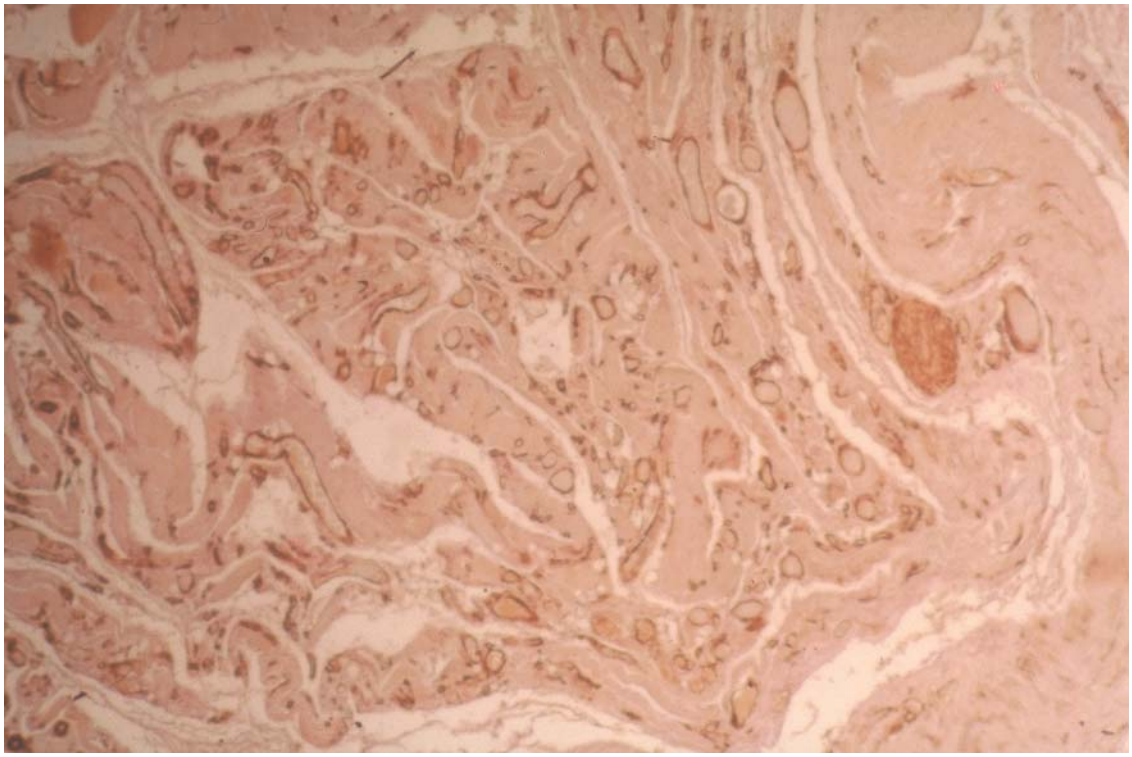

Figure 3. *SMA (smooth-muscle actin) staining disclosed areas of intramuscular hemangioma around the fibrous capsule. The endothelium did not reveal any nuclear pleiomorphism, mitotic activity or significant necrosis.*

According to the few reported cases to date, there is no gender or age predilection, as it occurs equally in males and females, ranging from 9 months to 80 years old. Clinically, IPEH occurs most frequently in the head and neck, fingers, trunk and cutaneous veins, as a small, firm, superficial mass with red to blue discoloration of the overlying skin (8, 9). Deep-seated IPEH of the extremities arising from pre-existing, intramuscular, cavernous hemangiomas, as in the described case, are considered to be very rare (10).

Histologically, IPEH has a characteristic exuberant endothelial proliferation within the lumen of medium-sized veins. Microscopically, the tuft-like or papillary proliferation of endothelial cells is nearly always associated with a thrombus and seems to represent a peculiar variant of an organizing process. The papillary structure and exuberant endotheliazation of IPEH necessitate ruling out the much more frequent angiosarcoma. The following features are important in the differential diagnosis: a) intravascular papillary endothelial hyperplasia is often well-circumscribed or encapsulated; b) the proliferative process is completely limited to the intravascular spaces; c) though the endothelial cells are hyperchromatic, extreme nuclear atypia and frequent mitotic figures cannot be seen; d) papillae are composed of fibro-hyalinized tissue of two or more endothelial cell layers without any covering; e) there is no true endothelial confirmation of IPEH; f) tangential sectioning may reveal pseudochannels, but no irregular or anastomosing blood vessels in the stroma; and g) necrosis is an unusual finding in IPEH (11, 12).

The importance of intravascular papillary endothelial hyperplasia or Masson's hemangioma resides in its capacity to simulate the growth pattern of a malignant soft-tissue sarcoma, such as a low-grade angiosarcoma. It should be in the surgeon's mind when dealing with equivocal cases of soft-tissue tumors and be recognized by the pathologist on the basis of its characteristic appearance and microscopic features. It should also be identified as a perfectly benign condition, as clearly evidenced by the presented case report.

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