

Intravascular Papillary Endothelial Hyperplasia (Masson's Tumor): Diagnosis the Plastic Surgeon Should Be Aware of

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Summary: Intravascular papillary endothelial hyperplasia (IPEH) or Masson's tumor is a rare benign entity commonly found on the head, neck, and upper extremities. It usually arises within a blood vessel but is considered to be a nonneoplastic reactive process often associated with vascular injury. Typically, IPEHs cause no symptoms and present as slowly growing soft-tissue masses. Given their prevalent location and indolent clinical presentation, the plastic surgeon should be familiar with this rare entity. We are presenting a case of IPEH of the forehead with unusual clinical and pathologic characteristics. Differential diagnosis, special considerations regarding preoperative work-up, and treatment options are discussed. (*Plast Reconstr Surg Glob Open* 2017;5:e1122; doi: 10.1097/GOX.0000000000001122; Published online 10 January 2017.)

Intravascular papillary endothelial hyperplasia (IPEH) or Masson's tumor is a rare type of vascular tumor usually found in the soft tissue and skin. It is more common in females and usually causes no symptoms. The treatment is surgical removal, and, given the location of these tumors, patients may be referred to a plastic surgeon for resection and/or reconstruction. To our knowledge, there are no cases of IPEH reported in the plastic surgery literature.

CASE REPORT

A 64-year-old man presented for evaluation of a pulsatile mass on the left lateral forehead. The patient reported a history of trauma in the area 2 months ago before presentation with hematoma formation. The wound was repaired with staples at an outside hospital. A few weeks later, he noted a steadily enlarging mass at the site of the repair. The patient reported associated pain and a "throbbing sensation," but he denied any overlying

skin changes, bleeding, or drainage. His medical, surgical, social, and family history was noncontributory.

Physical examination revealed a 2.5 × 2 × 1.75 cm pulsatile mass on the left lateral forehead (Figs. 1, 2). The mass was fully compressible, smooth, slightly mobile, and minimally tender to palpation, and a faint thrill was palpable. Transillumination was negative, and the overlying skin was thin and attenuated.

Based on the clinical presentation and physical examination findings, the most likely diagnoses included traumatic pseudoaneurysm of the temporal artery versus traumatic arteriovenous malformation. The patient was symptomatic and requested removal of the mass. A preoperative biopsy was deferred, and the patient was scheduled for surgical resection. A 3-cm incision was made within a natural forehead crease overlying the mass. A Doppler was used to identify the anterior branch of the superficial temporal artery proximal to the mass. The frontalis muscle was divided in a horizontal direction. Deep to the muscle, an artery with an associated vein leading into the mass was identified. Distal vascular structures were ligated, followed by proximal vessels (Fig. 3). The mass was completely removed and sent to pathology. The frontalis muscle was repaired, and a 2-layer closure was performed.

The pathology report showed a tan and red soft-tissue mass measuring 1.5 × 1.5 × 0.9 cm consistent with IPEH. The surface was serially sectioned to reveal fibrous cystic structure containing thrombi. Proliferation of endothelial cells with fibrin deposition was identified, with no evidence of atypia or mitoses (Fig. 4). The margins were negative.

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Fig. 1. Preoperative image of the lesion measuring 2.5 × 2 × 1.75 cm on the left lateral forehead.



Fig. 2. Another view of the lesion preoperatively.

The patient's postoperative course was uncomplicated. He had no sensory or motor deficits, and no recurrence has been identified 5 months postoperatively. The aesthetic result was excellent, despite the history of previous injury and type of initial wound repair (Fig. 5).

DISCUSSION

In 1923, Masson¹ first described a “vegetant intravascular hemangioendothelioma,” now known as IPEH. These lesions account for approximately up to 2% of vascular tumors of the skin and soft tissue and are more common in females of any age.² They can present anywhere in the body but are more common in the head, neck, and upper extremities. Less than 250 cases in the head and neck have been reported over the last 35 years, with less than 5% involving the forehead.³ There are no cases of IPEH reported in the plastic surgery literature.

The pathogenesis of this rare entity is poorly understood. Although IPEH arises within a vessel, it is not a primary vascular tumor. After vascular injury results

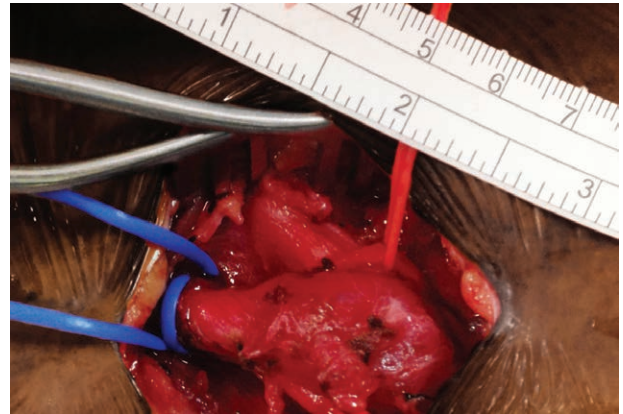


Fig. 3. Intraoperative picture of the lesion consistent with Masson's tumor.

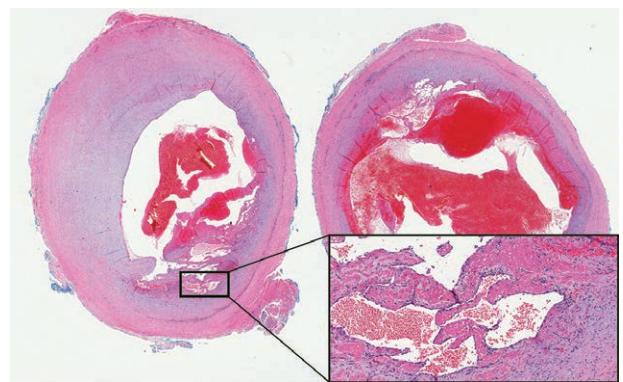


Fig. 4. Histological features of IPEH showing endothelial cell proliferation, fibrinous deposits, and dilated vascular spaces (H&E, low and magnified high power).



Fig. 5. Postoperative image at 4-week follow-up with excellent aesthetic outcome.

in thrombus formation inside a vessel, usually a vein, inflammation and vascular stasis can mediate endothelial cell proliferation.⁴ Chemo-attracted macrophages are thought to play a pivotal role by releasing endothelial basic fibroblast growth factor, which is thought to stimulate the formation of IPEH.⁵

Even though IPEH is a nonneoplastic reactive process, it can present similarly to other vascular benign or malignant neoplasms. The differential diagnosis includes angioma, vascular malformation, pyogenic granuloma, epithelioid hemangioendothelioma, angiosarcoma, Kaposi's sarcoma, and other more rare vascular tumors. Angiosarcoma does not typically present in the lumen of vessels, which differentiates it from IPEH.⁶

Histologically, IPEHs can be classified into 3 types. Primary or pure form typically presents within a dilated vein, rarely an artery, and most commonly arises in the subcutaneous soft tissue of the hand, digits, or the head and neck.⁷ Secondary or mixed IPEHs reside in preexisting vascular abnormalities, whereas extravascular IPEHs represent the least common type and arise in hematomas. Microscopically, they consist of papillae with a monolayer of plump endothelial cells, fibrous connective tissue core, and characteristic dilated vascular channels. IPEH is typically positive for CD31, CD34, SMA, and factor VIII-related antigen.⁴ CD105 is expressed on vascular endothelial cells and is positive in primary vascular neoplasms, differentiating IPEH from angiosarcoma.^{8,9} Atypia, pleomorphism, and necrosis are typically absent. In our case, pathology report showed primary IPEH, likely arising from a vein.

Clinically, IPEHs usually present as well-defined, oval, red or purple superficial papules or deep nodules. Typically, they are small in size, measuring 0.5 to 5 cm. They usually grow in the skin, dermis, or subcutaneous tissues but have been also found in the oral and lingual mucosa, heart, and small bowel. Preoperative ultrasound can detect one or more vessels associated with the lesions, differentiating IPEHs from other soft-tissue masses.²

Treatment consists of complete surgical resection. There is no indication for wide margins of excision; however, incomplete resection may result in recurrence.¹⁰ There are no cases of malignant transformation reported to date. We performed the mass resection with a 1 to 2 mm margin of surrounding healthy soft tissue.

Our case demonstrates some unusual findings. Our patient presented with a pulsatile soft-tissue mass, which is not typical for this type of tumor, especially because pathology showed venous origin. Additionally, our patient had a history of trauma in the area with subsequent hematoma formation. Even though extravascular IPEHs typically arise in hematomas, the pathologic analysis revealed primary IPEH, indicating that even with the history of hematoma, a primary intravascular IPEH can still develop after vascular trauma.

CONCLUSIONS

The plastic surgeon should be aware of this rare tumor given the common involvement of the face and the hand. IPEHs are benign tumors that can mimic other more common entities, including hemangiomas and vascular malformations. Complete surgical resection is considered therapeutic; however, if a malignant process is suspected, a more cautious approach with preoperative biopsy and more extensive margins of resection is recommended. Preoperative ultrasound can be utilized to differentiate IPEH from other soft-tissue masses.

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PATIENT CONSENT

The patient provided written consent for the use of his image.

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