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Presentation and Discussion of Masson's Vegetant Intravascular Hemangioendothelioma in the Case of Recurrent Blunt Force Trauma

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Background	Intravascular papillary endothelial hyperplasia (IPEH) is a benign neoplasm of the endovasculature, preferentially selecting the areas of the head and neck. IPEH represents a thrombogenic proliferation of endothelial papillae that results in appreciable mass causing cosmetic more than physical deformity. Although manifestations appreciable on gross physical exam suggest a malignant course, histopathological analysis reveals a consistently benign development of endothelial papillary projections that promote thrombogenesis, thus conveying the gross physical manifestations of IPEH. Correct identification relies on histopathologic assessment, and thorough surgical dissection is necessary for curative treatment.
Summary	A 31-year-old man of Hispanic descent reported to our clinic complaining of a six-month course of painless mass along the right forehead. Surgical excision and histopathological analysis revealed dense endoluminal papillary projections consistent with IPEH. A subsequent mass three months later was identified on the contralateral forehead and was further identified as a less histopathologically differentiated case of IPEH. The patient had a history remarkable for recurrent blunt physical trauma secondary to prior altercations.
Conclusion	We present two instances of IPEH arising in one individual with a history of recurrent trauma. The unique endovascular manifestations of the disease combined with the thrombogenic nature of recurrent trauma suggest a likely course of disease progression. Furthermore, the histopathological differences between the two specimens suggest a plausible time course for evaluating changes at the microscopic level.
Key Words	Masson's tumor; intravascular papillary endothelial hyperplasia

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Case Description

A 31-year-old Hispanic man was referred to the plastic surgery division with a six-month history of progressively enlarging mass of the right forehead. Although painless throughout the day, the mass caused the patient significant discomfort when sleeping on the affected surface. Physical examination revealed a firm, mobile, non-pulsatile area along the right temporal hairline without overlying skin changes, discolorations, or puncta (Figure 1). Past medical history was noncontributory except for previous physical altercations resulting in recurrent trauma to the affected area.

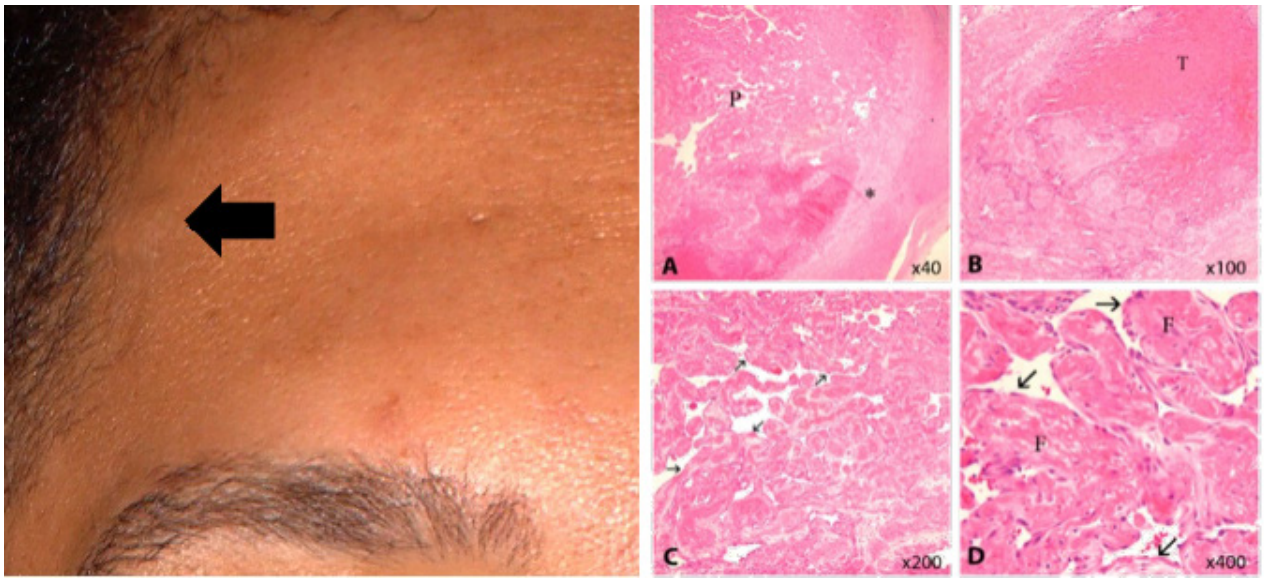
The surgery team suspected the mass to be an inclusion cyst or lipoma and elected against further preoperative imaging based on symptomatology. The patient underwent elective excisional biopsy, during which the specimen was isolated in the subcutaneous plane and removed intact. On gross examination, the well-encapsulated mass measured 1.5 cm × 1.0 cm × 0.5 cm and was composed chiefly of organized thrombus and intravascular papillary endothelial hyper-plasia characteristic of a Masson's pseudoangiosarcoma (Figure 1A–D).

One month postoperatively, the patient noticed a second enlarging mass on the contralateral forehead and returned to the clinic for further evaluation (Figure 2). Elective resection of this new lesion revealed a well-encapsulated mass similar to the previous pseudoangiosarcoma measuring 1.3 cm × 0.8 cm × 0.5 cm with mixed hemangiomatous structure. Pathological analysis of the specimen identified varicosity of the involved blood vessel with scant papillary formation (Figure 2A–D). The postoperative course has been benign, with no signs of recurrence at five-month follow-up, and the patient has been discharged from our clinic without complaint.

Discussion

Intravascular papillary endothelial hyperplasia (IPEH), originally identified as vegetant intravascular hemangioendothelioma and discussed elsewhere as Masson's pseudoangiosarcoma or Masson's tumor, was first described by the French physician Claude L. Pierre Masson. In 1923, Masson identified an unusual series of anastomosing vascular proliferations with endothelial surfaces rich in dense papillary projections that formed an interconnected network previously unidentified in vascular medicine.[1] Although

Figure 1. Initial Instance of Intravascular Papillary Endothelial Hyperplasia (IPEH). Published with Permission



A) H&E stain of specimen under 40x magnification; B) H&E stain of specimen under 100x magnification; C) H&E stain of specimen under 200x magnification; D) H&E stain of specimen under 400x magnification. Large arrow = clinical aspect of the mass on physical examination
** = endovascular epithelium, smaller arrow = papillary epithelial lining, P = papillae, T = thrombus, F = fibrostromal core*

initially classifying the lesion as a tumor, in 1932, Masson's successor Dr. Folke Henschen disputed this claim, instead insisting the papillae resulted from a proliferative thrombopoietic endovasculitis.[2]

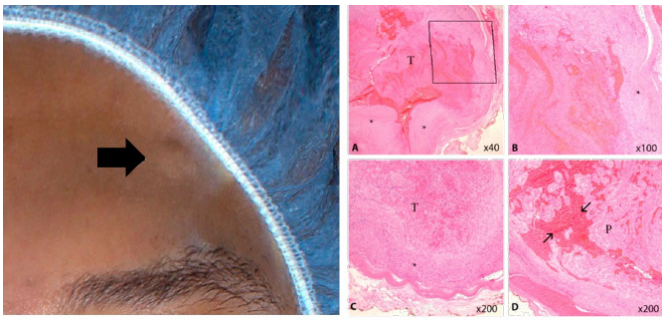
It would not be until the mid-1970s when Clearkin and Enzinger would popularize the modern terminology of intravascular papillary endothelial hyperplasia and solidify the current nomenclature.[3] Classification proposed by Hashimoto et al. divides IPEH among Type I primary, or pure, lesions often limited to the small veins of the head, neck, fingers, and trunk with Type II secondary manifestations reported occurring within preexisting angiomata, vascular malformations, and varicosities.[4–6] A rare Type III form, although noted, is not discussed in great detail.[6]

IPEH represents a unique subset of benign head and neck vascular tumors, consisting of approximately 2–4 percent of all vascular neoplasms of the skin and subcutaneous tissue.[7] The underlying pathology is attributed to the dysregulated proliferation of endovascular endothelium, resulting in the formation of a dense network of papillae capable of promoting thrombogenesis.[8] The subsequent thrombus and eventual fibrous capsule encircling the lesion convey the palpable mass appreciated on physical exam. Despite the rapidity with which these masses can form, they lack malignant potential and often present cosmetic rather than life-threatening concerns for the affected patient.

Early lesion pathology is characterized by endothelial cell proliferation with enhanced cellular mitotic activity that creates a fibroepithelial papillary protrusion emerging from the vessel wall into the vascular lumen. Invariably, the disruption of endoluminal lamellar flow promotes thrombogenesis and subtotal or total occlusion of the blood vessel (Figure 1). Papillae ultimately fuse into an anastomotic network of vessels in a loose connective tissue. In the final stages, the lesion surrounds itself with a fibrous sclerotic scar, contributing to the firmness appreciated on physical exam.[4,9,10]

In clinical appearance, IPEH presents characteristics resembling both benign as well as malignant vascular lesions. Growth and development of the palpable mass have been attributed to progressive thrombogenesis secondary to the turbulent flow created by the papillae and not by the endothelial protrusions themselves.[3,8] Histological examination of the papillae does not reveal the anticipated hallmarks of malignancy, with no atypia, pleomorphism, or necrosis appreciated (Figure 2).[11] However the clinical course may strongly parallel malignancy with recurrence of the palpable mass possible secondary to the reformation of thrombus.[8] Due to the benign nature of the lesion, its outcomes are more similar to those of synovial cysts, mucoceles, lipomas, pyogenic granulomata, and hemangiomata rather than their malignant counterparts.[12]

Figure 2. Follow-Up Occurrence of IPEH. Published with Permission



*Evolution of second mass with similar symptomatology on left forehead three months following surgical evaluation of initial IPEH; histology notes significantly lighter density of endovascular papillae compared to previous lesion; A) H&E stain of specimen under 40x magnification; B) H&E stain of specimen under 100x magnification; C) H&E stain of specimen under 200x magnification; D) H&E stain of specimen under 400x magnification (Large arrow = clinical aspect of the mass on physical examination, * = arteriolar wall, structure between smaller arrows = red blood cells, P = papillae, T = thrombus)*

Ultrasound, while valuable, proves difficult to distinguish between a simple cyst and IPEH. Doppler sonography presents one diagnostic possibility based on its ability to clearly demonstrate lesion flow and hypervascularity while ruling out thrombus formation; however, the relatively small size of IPEH at initial encounter makes flow an unreliable measure of assessment.[13] Craig et al. propose useful metrics for assessing IPEH based on a small sampling of thirteen cases where magnetic resonance (MR) with and without ultrasonography showed characteristically high T2 peripheral signal with variable central T2 density on MR and hypoechogenicity on ultrasound.[14] The majority of patients with IPEH, therefore, do not undergo diagnostic imaging as the small size of the lesion and the superficial location within the deep dermis or subcutaneous tissue present challenges for minimally invasive techniques.[4,13]

A biopsy is necessary for accurate diagnosis. Immunohistochemical studies have in special cases proven useful where endothelial cells lining the nascent papillae have expressed CD31 and CD34. However, this is not a consistent or standardized method for positive identification.[9] Excisional biopsy and primary wound closure remain the

gold standard of treatment for IPEH, as the small size and superficial location of the malformation make additional wound closure techniques unnecessary. Recurrence, when present, is attributed to a failure to entirely excise the primary lesion or, more commonly, in failing to manage secondary lesions in the context of comorbid vascular disease, which facilitate neothrombogenesis.[4]

This case demonstrates a patient with two successive lesions in anatomically similar distribution secondary to recurrent physical trauma. In this case, the lesions provide meaningful insight into the timeline of the pathogenesis in IPEH, supporting the conventional theory that this pathology is likely a reactive process secondary to traumatic thrombus formation. The exact nature of this trauma may not be immediately evident to the patient, as it is reported that most individuals have no recollection of a specific inciting event before the onset of observable physical symptomatology.[4] In this instance, the first mass was excised six months after manifestation as a mature IPEH with the characteristic histology of densely populated mature papillae. This contrasts with the second lesion, which was evaluated only three months into the pathological course and was noted to have substantially lower papillary density. Due to the thromboproliferative nature of the endovascular papillae, it is reasonable to assume additional thrombogenic events would prove contributory to the progression of this condition.

Conclusion

We conclude that due to the nature of the lesion and the time required for proliferation of endovascular papillae, some minor precipitating traumatic thrombus formation perhaps associated with activities of daily living may be enough to affect as yet ill-defined individuals with a predisposition to IPEH. Based on the previous history of the first lesion, we suspect that given adequate time the second mass would have followed identical histologic findings of the first and represents an earlier form of IPEH. Although more research is required to characterize its evolution, this case report provides valuable information regarding the probable cause, progression, and possible underestimated incidence of Masson's psuedoangiosarcoma.

Lessons Learned

We present a case in which a patient with a history significant for recurrent blunt force trauma secondary to physical altercation was able to manifest two instances of IPEH in different stages of development. We discuss the potential clinical significance of the proliferative nature of this disorder and current recommendations in the analysis and treatment of IPEH.

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