



Intravascular Papillary Endothelial Hyperplasia (Masson's Tumor) of Wrist

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ABSTRACT

Nonspecific slow growing lesions of hand and wrist can be difficult to diagnose. We present a case of intravascular papillary endothelial hyperplasia (IPEH), or Masson's tumor of the wrist. Present case is a 23-year-old female presented with a vague, soft, mobile and mildly tender swelling over the ventral aspect of the right wrist. The present case highlights the importance of excision and histopathological examination along immunohistochemistry in these lesions.

Keywords: Vascular Tumor, Masson's Tumor, Intravascular Papillary Endothelial Hyperplasia, Wrist

Introduction

Intravascular papillary endothelial hyperplasia (IPEH) is a benign intravascular lesion. It mimics other benign as well as malignant vascular proliferations. It is mostly found in head and neck region and the extremities. ^[1] The classical histological features of this is multiple papillary structures covered by hyperplastic endothelial cells in the vascular lumen. Along with this intra-luminal location of the lesion and a close association with organizing thrombus, the absence of necrosis, cellular pleomorphic, and mitotic activity are helpful in clinching the diagnosis. In addition, immunohistochemistry helps to demarcate its vascular origin and proliferative potential. ^[2] We present a case where we have evaluated histological and immunohistochemical markers in Intravascular papillary endothelial hyperplasia. It is important to not mistake this clinically benign lesion for other well differentiated tumors of vascular origin.

Case Report

Present case was a 23-year-old female who presented with a slowly growing mass on the ventral aspect of the right wrist. There was no history of trauma. The mass was slow growing over the past few years, but started to hinder the day-to-day movements. The lesion was approximately 2 cm × 1.5 cm, soft, mobile and slightly tender. The patient requested the mass to be removed as it was causing problem in her writing work.

On gross examination a grey brown, irregular tissue piece was identified. H & E-stained section, revealed, dilated vessels with intraluminal papillary formations. Some areas show desquamating endothelial cells and focal thrombi (fig 1&2). Immunohistochemical stain for CD34 highlighted

the endothelial lining. A final histological diagnosis of IPEH was rendered.

Discussion

Intravascular Papillary Endothelial Hyperplasia commonly called as Masson's tumor after its original description by Masson who called it vegetant intravascular haemangioendothelioma. ^[1]

It is a rare benign vascular endothelial proliferation representing about 2% of all the vascular tumors in skin. ^[2]

Pathogenesis of this lesion is still unknown, but it is considered as reaction and eventual proliferation of endothelial cells of normal blood vessels in response to a variety of chemical signals probably hormonal. ^[3-5]

Masson in his original description regarded it as a true neoplasm arising from an ulcerated haemorrhoidal vein, which showed degenerative changes including necrosis and thrombosis. ^[1, 6]

It usually presents in the third and fourth decades of life, most commonly occurs in the skin and subcutis of the fingers and head-neck regions as "pure" lesions ^[7]. Although this lesion can occur in virtually any vessel, it is mostly located in veins of head-neck, fingers and trunk. Clinically it appears as a superficial subcutaneous swelling with overlying skin showing blue or red tinge.

Three distinct types are described 1 primary "pure" or intravascular form which arises in dilated blood vessels, usually veins. 2 a secondary or "mixed" form 40% which originates within a pre-existing varix like aneurysm, haemangioma, arteriovenous malformation, lymphangioma and pyogenic granuloma. 3 an extravascular form which

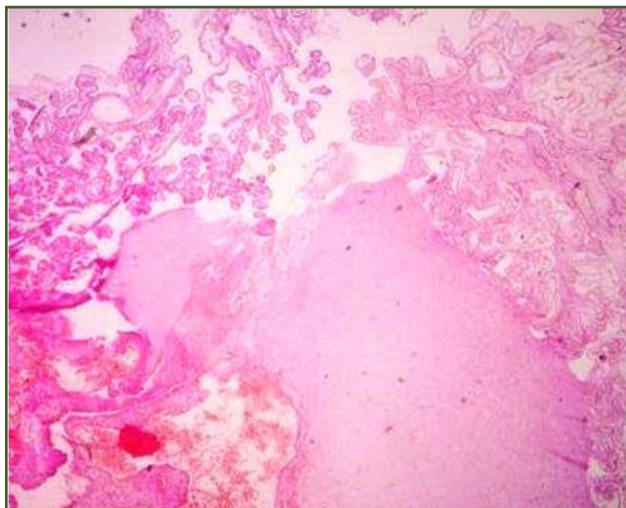


Fig. 1: Photomicrograph showing desquamating endothelial cells and focal thrombi (H&E, 100x).

arises in hematomas.^[8, 9] Although any of this form is not related to previous history of trauma.

Pure lesions most commonly occurs in skin and subcutaneous tissue of fingers, head and neck regions.^[2-5, 10], while mixed lesions tend to occur intramuscular region^[11]. Other common locations of this lesion are cranial cavity, mandible, maxillary sinus, tongue, orbital fossa, thyroid, triceps, superior vena cava, lung parotid, cervix, liver, adrenal gland renal vein.

The differentials of this entity include various other benign as well as malignant vascular proliferative lesions. Particularly those in which there is fusion of papillae to form anastomotic pattern which may mimic angiosarcoma^[12-15]. IPEH has characteristic papillary proliferation of reactive endothelial cells. These papillae are composed of hypocellular and hyalinised cores which are lined by one to two layers of plump endothelium. It is commonly associated with thrombus. IPEH does not have any cellular atypia, atypical mitosis or infiltrative growth pattern, all these features which are present in malignancy.^[16]

Low grade angiosarcoma shows an infiltrating pattern of growth along with nuclear hyperchromasia, atypical cells, mitosis and necrosis^[17-19].

Immunohistochemistry is also helpful in detection and categorization of this lesion. Cellular markers of proliferation Ki-67 may help identifying the biological behaviour of this lesion^[16]. Various other immunohistochemical markers like CD34, CD31, CD105 are also helpful. CD105 is help is differentiation from angiosarcoma in which it is overexpressed^[18]. IPEH is also show varying positivity with vimentin, laminin, podoplanin and type IV collagen.^[14, 20]

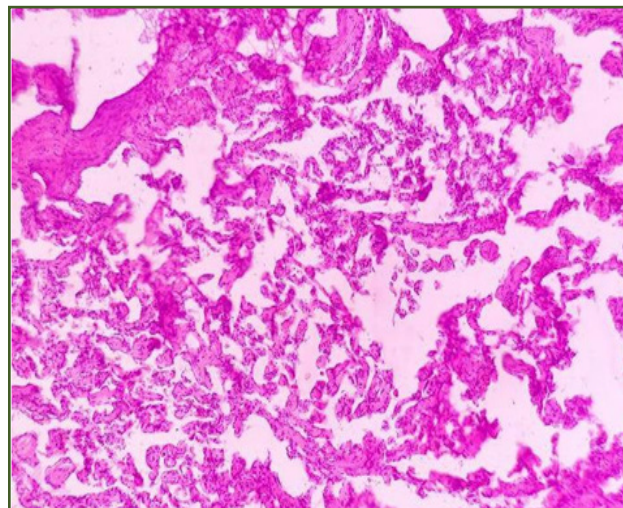


Fig. 2: Photomicrograph showing dilated vessel with intraluminal papillary formations (H&E, 100x).

The treatment of Masson's tumor is conservative surgical excision. It has excellent prognosis except in cases with intracranial involvement. Recurrence is very rare except when it is arising in primary vascular lesion which itself is recurring. The role of radiation therapy is not clear but there are some reports of successful treatment in cases of recurrent IPEH, both intracranially and in digits^[2-5, 10].

Conclusion

In conclusion clinical features of Masson's tumour are nonspecific. Histopathological examination and immunohistochemistry, is helpful for diagnosis. Due its close morphological resemblance with low-grade angiosarcoma care must be taken to prevent misdiagnosis and unnecessary aggressive treatment.

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