Masson's Tumor – Not Rare Anymore?

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Abstract: Masson's tumor is an unusual intravascular hemangioendothelioma, a rare condition affecting the vessels and known to simulate an angiosarcoma, or lesions like intravascular pyogenic granuloma and intravascular fasciitis. Despite being an uncommon tumor, we present 3 cases of Masson's tumor – one each in the nasal cavity, cheek and neck. [N Rattan Natl J Integr Res Med, 2020; 11(1):88-89]

Key Words: Intravascular hemangioendothelioma, angiosarcoma, papillary

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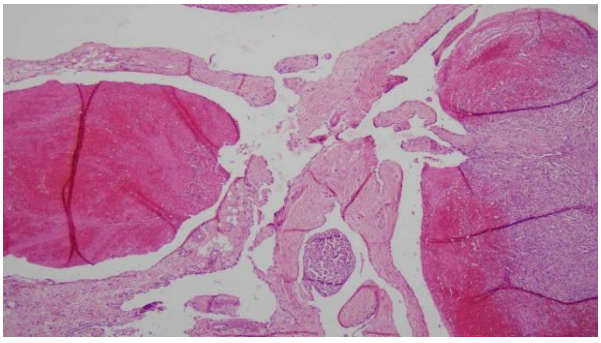
Introduction: Vegetant intravascular hemangioendothelioma described by Pierre Masson in 1923[1] is an intravascular papillary endothelial hyperplasia (IPEH) also known as intravascular angiomatosis or Masson's pseudoangiosarcoma. It is an unusual benign vascular lesion comprising approximately 2% of vascular tumors of the skin and subcutaneous tissue[2]. The lesion is characterized by proliferation of endothelial cells with papillary formation associated to a thrombus and fibrin or collagenous connective tissue deposits leading to vascular obliteration. Diagnosis of Masson tumor can be difficult with broad differential diagnosis including intravascular fascitis, intravascular pyogenic granuloma and angiosarcoma[3]. We describe 3 cases of intravascular endothelial papillary hyperplasia, one each in the neck, cheek and nose.

Case Reports: Case 1: A 70 year old male had a single cystic lesion on the left side of the neck. Excision biopsy revealed a grey brown to grey black cystic structure measuring 2.5cm in diameter. Cut sections revealed grey brown areas filled with hemorrhagic fluid. Histopathological examination showed an intravascular lesion having papillary formation with hyaline and fibrous cores, anastomosing vascular channels and plump endothelial cells. Lumen showed evidence of thrombus [Figure I].

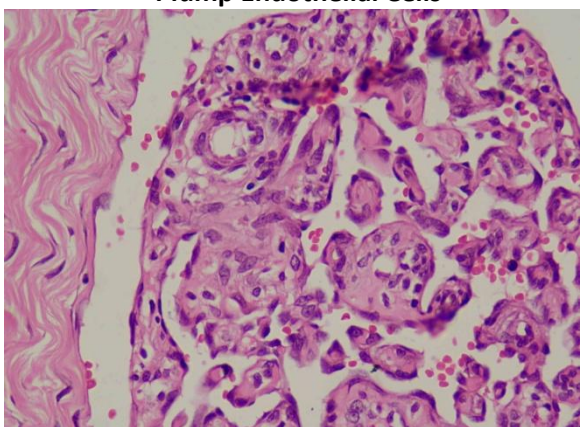
Figure 1: H&E (4X) Showing Anastomosing Vascular Channels With Thrombus



Chronic inflammatory cells with hemosiderin laden macrophages were seen in the periphery. No evidence of anaplasia, mitotic figures or necrosis was noted.

Case 2: A 40 year old female had cystic swelling right cheek. Excision biopsy done revealed a soft tissue piece measuring 1.6x1x0.7cm. Histopathology showed multiple vascular channels with intravascular papillary stalk, a fibrous core and lined by plump endothelial cells [Figure II]. There was no evidence of nuclear atypia, mitosis or necrosis .

Figure 2: H&E (40X) Intravascular Papillae With Plump Endothelial Cells



Case 3: 55 year old female patient had a right nasal cavity mass occupying the entire nasal cavity attached at the junction of bony and cartilaginous part of the septum. Endoscopic removal of the mass was done, which showed grey black soft tissue pieces collectively measuring 4x4x1cm.Histopathology showed multiple capillary sized blood vessels and irregularly anastomosing vascular channels lined by plump endothelial cells. Large areas of hemorrhage and thrombotic foci were noted. Focal areas had formation of papillary structures confined to vascular spaces with a hyalinised stalk lined by plump endothelial cells. The polypoidal mass was lined by focal respiratory

epithelium with large areas of ulceration. No atypia or mitosis was noted.

Discussion: In the year 1983, Hashimoto et al[4] described three subtypes of IPEH : (1) a primary (pure) form occurring in the dilated vessels, (2) a secondary (mixed) form existing in the varices, hemangioma and lymphangiomas and (3) an extravascular form occurring infrequently[5] and developing in hematomas. All 3 subtypes described involve the proliferation of endothelial cells around a thrombus in a setting of venous stasis. Females are affected more frequently than males and generally affect middle age[6]. Although the pathogenesis of IPEH is to date speculative, Masson in 1923 regarded it as a true neoplasm with secondary thrombosis[7].

However, considering that thrombi are frequently found in these lesions, some authors do believe that IPEH is an unusual form of thrombus organization[8-9].It is now believed that the thrombus serves as a matrix for ingrowth of papillary structures[10]. The differential diagnosis of the lesion, particularly in respect to angiosarcoma is emphasized. Angiosarcoma never occurs within the lumen of a vessel whereas Masson's tumor remains confined to the lumen of a vessel. Its differential diagnosis from other conditions like pyogenic granuloma, Kaposi's sarcoma, hemangioma and atypical vascular proliferation needs to be entertained and needs to be re-emphasized. Characteristics particular to IPEH are in the form of it being an intraluminal lesion, papillary formation related to thrombotic material, a papillary stalk which is fibrohyalinized with little or no cellular pleomorphism or mitotic activity. IPEH is known to recur when incompletely resected[10].

Conclusion: Masson's tumor is a rare benign tumor which affects all ages. This study of 3 cases had a female preponderance with age ranging from 40 to 70 years. The cases showed a predilection for involvement of the head and neck region. Clinical settings alone do not serve as the sole criterion for diagnosis, and a histological examination is considered essential to arrive at the diagnosis and thus, ensure that there is no possibility of it being a malignant lesion is not overlooked.

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