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INTRAVASCULAR PAPILLARY ENDOTHELIAL HYPERPLASIA (MASSON'S TUMOR)- A RARE FINDING IN THE ORBITAL REGION

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ABSTRACT Intravascular papillary endothelial hyperplasia (IPEH) is a reactive proliferation of endothelial cells in the blood vessel. It typically present as a small, firm, painless lesion with red to blue discoloration. Most of the reported cases were usually located in the skin and subcutaneous tissue of the head and neck and extremities. Ocular involvement is very rare, and mostly involves the eyelids; whereas orbital lesions occur even less commonly. Here we report a rare case of IPEH in a 38 year old male who presented with solitary orbital mass in right eye with proptosis and diminished vision. Histopathological examination revealed papillary and tufted capillary endothelial hyperplasia with no atypia of endothelial lining. The hyperplastic endothelium showed immunohistochemical positivity for CD34 and vimentin. The diagnosis of IPEH (Masson's tumor) was made. Prognosis of IPEH is excellent, if the lesion is completely excised. Although IPEH is benign, clinically this lesion is important because it present as a mass lesion or may be mistaken for angiosarcoma; it should be considered in the list of differentials of proptosis.

KEYWORDS : Intravascular papillary endothelial hyperplasia, Masson's tumor, Orbit

INTRODUCTION:

Unexplained swellings in the orbital region pose a diagnostic dilemma for clinicians. One of the rarely found entities is Masson's hemangioma or intravascular papillary endothelial hyperplasia (IPEH). It was first described in 1923 by Pierre Masson as neoplastic proliferation of the endothelium, which he termed "hemangioendotheliome vegetant intravasculaire".[1] However, it is now believed to be a benign reactive vascular proliferation following trauma or vascular stasis. The current terminology IPEH was given by Clearkin and Enzinger in 1976 and is now most widely used.[2] Most of the reported cases in head and neck region affect the skin and subcutaneous tissue.[3] Ocular involvement is very unusual, and typically affects the eyelids.[4] The median age of patients is reported to be 45 years.[5] Here we present a case of solitary orbital mass in a middle aged male who presented with proptosis and dimness of vision. The diagnosis was confirmed as Masson's tumor on histopathological and immunohistochemical (IHC) examination.

Case Report:

A 38 year old male presented with diminished vision and painless, slowly progressive proptosis in the right eye since 10-12 months. There were no complaints with his left eye. There was no history of trauma, swellings in other parts of the body and weight loss. There was no significant past medical history. Ocular motility was mildly restricted in right eye with visual acuity of counting fingers at 2 meters. Fundus examination showed mild edema of disc with blurred margins but the vessels and macula were normal. Visual acuity of the patient's left eye was 6/6. Intraocular pressure was normal in both the eyes. Systemic examination was normal. Excision biopsy was done. Multiple grey white to grey brown soft tissue pieces measuring 0.6x0.3x0.2 cm were received. Microsections examined showed a dilated vein with partially organized thrombus (Fig.1). The lumen was filled with tufted, papillary projections lined by single layer of plump endothelial cells without atypia (Fig. 2). On IHC, the hyperplastic endothelium was positive for CD34 and vimentin (Fig.3,4). Based on histopathological and IHC findings a diagnosis of IPEH (Masson's tumor) was made.

DISCUSSION:

IPEH was first described by Pierre Masson in 1923. He described it as a true neoplasm, due to proliferation of endothelial cells into the lumen of vessel with subsequent obstruction, necrosis and secondary degeneration.[1] It has been described by various names including Masson's hemangioma, Intravscular endothelial proliferation, Masson's tumor, reactive papillary endothelial hyperplasia, and intravascular angiomatosis. It constitutes about 2% of the benign and malignant tumors of skin and subcutaneous tissues.[3] In order of decreasing frequency, it involves the fingers, head and neck, trunk, lower and upper extremities; the rare sites affected are thyroid, orbit, parotid gland, nose, mandible, pharynx, paranasal sinuses and central nervous system.[6] Ocular involvement is very rare, and mostly involves the eyelids; whereas orbital lesions occur even less commonly.[4]

It is now believed to be a reactive proliferation of endothelial cells that is associated with organization and recanalization of thrombus. The pathogenesis of IPEH is poorly understood. It has been proposed that IPEH formation is triggered off with release of basic fibroblast growth factor (bFGF) by the invading macrophages to the trauma site with proliferation of endothelial cells. Further release of more bFGF by the proliferating endothelial cells occur which further causes positive feedback of endotheial cell proliferation.[7] Three different types of IPEH have been discussed: (a) a primary (pure) form where changes are observed in a distended vessel; (b) a secondary (mixed) form that occurs in preexisting varices, hemangiomas, pyogenic granulomas, or lymphangiomas; and (c) an uncommon type in an extravascular location.[8] In our case IPEH is considered to be a mixed type.

Differential diagnosis of IPEH includes angiosarcoma, mucocele, intravenous pyogenic granuloma, spindle cell hemangioendothelioma, malignant endovascular papillary angioendothelioma and intravascular endothelioma. Angiosarcoma is a major differential of IPEH. IPEH is usually well-circumscribed, entirely lined by vascular wall with characteristic

papillary fronds. However, angiosarcoma invades tissue outside the vascular wall and has more than one to two layers of endothelial cells covering the papillary formation. It also show nuclear pleomorphism, numerous mitotic figures and necrosis.[9] The gold standard for diagnosis being histopathological examination.

Magnetic resonance imaging may be helpful as this lesion appears to enhance on T2- weighted images secondary to hameorrhage with in the lesion. Prognosis of IPEH is excellent. Follow up of large series showed no evidence of local invasion or metastasis.[10] However, IPEH may recur if it arises in a primary vascular lesion which may itself recur or if the lesion is incompletely excised.

Conclusion:

The clinical significance of IPEH is its resemblance to a variety of benign and malignant diseases. It should be considered in the list of differentials of proptosis due to mass lesion. The gold standard for diagnosis being histopathological examination. It tend to recur if incompletely resected. Correct diagnosis of the lesion is essential to prevent aggressive or inadequate treatment.

Figure 1: Dilated vein with partially organized thrombus (H&E 4x).

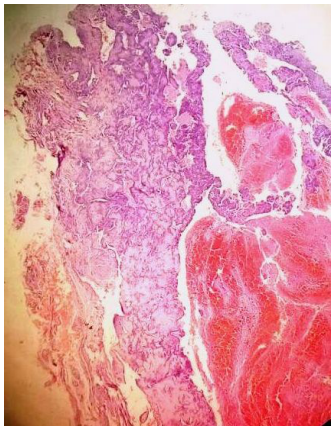


Figure 2: Lumen filled with tufted, papillary projections lined by single layer of plump endothelial cells without atypia (H&E 40x).

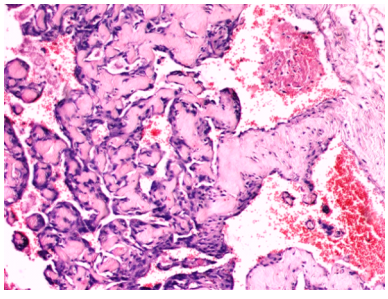


Figure 3: Hyperplastic endothelium positive for CD34 and vimentin respectively (IHC 40x, 100x)

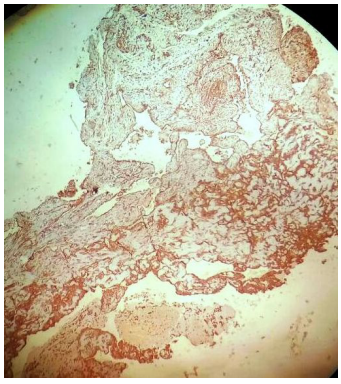
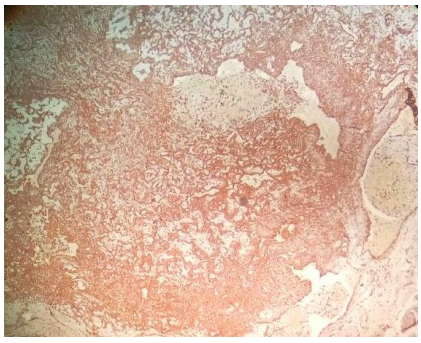


Figure 4: Vimentin positivity revealing mesenchymal origin of the lesion (IHC 10x)



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