



INTRAVASCULAR PAPILLARY ENDOTHELIAL HYPERPLASIA (IPEH) - A REVIEW ARTICLE

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ABSTRACT

IPEH, is defined as an unusual benign non-neoplastic vascular reactive lesion characterized histologically by papillary fronds lined by proliferating epithelium. IPEH was first described in 1923 by Pierre Masson in haemorrhoidal veins, under the term – hemangioendothelioma vegetant intravascularie. It is a non-specific disease entity, with a distinct histologically mimic angiosarcoma. The term IPEH was coined by Clerkin and Enzinger in 1976, since it was more descriptive and less confusing.

INTRODUCTION

Also known as; Hemangioendothelioma vegetant intravascularie, Masson's Lesion Tumor, Intravascular angiomatosis, Intravenous vascular proliferation, Intraendothelial hyperplasia, Masson's pseudo angiosarcoma. Amerigo (1980) reported that IPEH represents approximately 2% of all Benign and Malignant vascular tumors of skin and subcutaneous tissues after a retrospective study of 1,217 cases of IPEH. IPEH is generally associated with a thrombus which precedes the formation of papillary fronds which serves as a matrix for subsequent organization and recanalisation. The fragmentation of thrombus may result from the forces of intravascular pressure as well as its contraction leading to recanalisation [1,2].

Etiopathogenesis

The exact Etiopathogenesis is still unclear. IPEH is thought to represent an unusual, exuberant response of endothelial cells to an organizing thrombus. The exact mechanism is unknown: Masson, (1923): Considered IPEH as a true neoplasm with an intravascular endothelial

cell proliferation from vein walls. Henschen, Considered IPEH, as a hyperplastic reaction induced by blood stasis and perivascular inflammation and named it as Thrombopoietic proliferations endovascularitis. Silver and Salyer [3] (1985): Considered IPEH, as atypical manifestations of an organizing thrombus, which is more a pseudotumoral lesion and called it intravascular angiomatosis. Clerkin and Enzinger [4] Supported Salyer's theory and suggested the term IPEH. They stated that the altered blood flow induces formation of thrombus with a subsequent organization and recanalisation. Mc Clatchy [5] described that IPEH, was due to a reactive proliferation of endothelial cells, associated with exuberant organization and recanalisation of thrombus. Lever [6] suggested that the reactive and exaggerated attempt to recanalise in thrombus to be due to a basic fibroblast growth factor. The vasoformative cells of IPEH are thought to originate either from blood monocytes or the thrombosed vessel endothelium. A lymphatic vessel counterpart was reported in 1979 in a cystic lymphatic malformation.

Some authors have suggested the role of trauma as a predisposing factor especially at sites susceptible to trauma such as lips and tongue. However, such an

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association has been seen in only 4-7% of case of oral due to a hormonal influence on endothelial proliferation by locally produced antigenic growth factors.

Clinical features

They are Non Specific and include: AGE : 9 – 80 years, average: 34 years, SEX : 2:1 Female predilection, SIZE: 0.2 to 2 cm SITE: Skin and Subcutaneous tissues of fingers, head and neck, extremities and trunk where VARICES usually occur. In the Oral Cavity IPEH occurs in the Deep mucosa without any involvement of overlying tissue. Lower Lip > Tongue > Upper lips > Angle of mouth. The first oral lesion, located on the lower labial mucosa was reported in 1978 by Heyden et al. Occasional cases in lymphatic channels and maxillary sinus is also reported. Clinically, IPEH presents as a solitary, asymptomatic, slow growing (duration 10 days to 20 years), Bluish to reddish nodule of firm consistency which rarely exceeds 20 mm in diameter. Patients usually consult the clinician for functional or esthetic concerns.

Histologic features

The lesion is well circumscribed or encapsulated with the proliferative process entirely limited within the vascular wall and characterized by papillary fronds, which appear as bundles, lined by one or two endothelial cell layers of the distended tumour of medium sized veins. In certain areas, the tips of the papillae seen to float freely into the vascular lumen. The stalks of the papillae consist of a central core of fibrin or hyalinised C.T. sometimes surrounded by fibrosis [7].

Histologic differential diagnosis

Angiosarcoma, Intravenous atypical vascular proliferation, Spindle cell hemangioendothelioma, Malignant endovascular papillary angioendothelioma/ DABSKA's tumor, Intravenous pyogenic granuloma

Histologically, IPEH can be subdivided into Primary /

Pure form: It appears in dilated vascular spaces usually in the veins of head or upper limbs and appears as a solitary, slowly growing often painful nodule located within a dilated dermal or S.C. vein.

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lesions. A female predilection is suggested to be probably

- Secondary / Mixed Form: These occur in a pre-existing vascular lesion such as Hemangioma, Pyogenic granuloma, Lymphangioma etc.
- Extravascular form: They are seen arising from hematoma / lymphatic channels [8].

Important Characteristics to Distinguish IPEH from Angiosarcoma

- Grossly well circumscribed / encapsulated
- Proliferation confined to intravascular spaces
- Endothelial cells may be hyperchromatic, with absence of nuclear atypia and mitoses.
- Papillae consist of fibrohyalinised tissue covered by not more than 2 endothelial cell layers.

There are no true endothelial fronds.

- Irregular and anastomosing blood vessels are absent in stroma, although pseudochannels may be present due to Tangential sectioning.

Necrosis is unusual.

- Management is Surgical excision while prognosis is excellent. Its recurrence is rare ranging from 0.14 to 3% possibly due to incomplete excision or regrowth of underlying tissues.

Immuno histochemistry

Albrecht and Kahn showed a progression of the immunophenotype of the lining cells from histiocytic to endothelial. The cells initially reacted with antibodies for the histiocytic marker Ferritin in the earliest stages, followed by positive reaction for VIMENTIN and at the final stages for FACTOR VIII related antigen, which was similar to the progression seen in ordinary organizing thrombus which further supports the hypothesis of organizing thrombus and recanalisation

CONCLUSION

Masson's tumor is an uncommon lesion thought to be an atypical manifestation of organizing thrombus which is clinically nonspecific and histologically resembles angiosarcoma which necessitated accurate microscopic diagnosis and close clinicopathologic correlation.

