Cutaneous Intravascular Papillary Endothelial Hyperplasia of the Forearm: A Case Report
J.M. Farah, N. Sawke, G.K. Sawke

Department of Pathology, People's College of Medical Sciences & Research Centre, Bhanpur, Bhopal-462037

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Abstract:
Intravascular papillary endothelial hyperplasia (IPEH) or reactive vascular endothelial lesion is a rare benign condition and is also known as Masson's haemangioma. Blood vessel injury or venous thrombosis may predispose to this condition. Distinction of reactive entities from neoplastic condition like angiosarcoma is important. A case of cutaneous IPEH in forearm of an adult male is being reported here.

Key Words: Masson’s Haemangioma, Angiosarcoma, Intravascular papillary endothelial hyperplasia (IPEH).

Introduction:
It was first described by Masson (1861) in hemorrhoidal vessels as “Vegetant intravascular hemangioendothelioma.” Albrecht & Kahn (1990) studied immunohistochemistry of IPEH and concluded that it is an exuberant organization and recanalization of a thrombus. Kuo et al (1976) noted its occurrence in previously normal vessels or in varices, hemorrhoids, hematomas, pyogenic granulomas and hemangiomas. Intravascular papillary endothelial hyperplasia is considered to be a reactive vascular proliferation following traumatic vascular stasis. A clinico-pathologic study of 91 cases done by Hashimoto et al (1983) noted that it simulates angiosarcoma because of the presence of papillary formation, anastomosing vascular channels and plump endothelial cells. It is identified because of the exclusive intravascular nature of the process, the lack of necrosis, bizarre cells and atypical mitosis, the characteristically fibrinous and/or hyaline (deeply eosinophilic) appearance of the papillary stalks and the frequent finding of residual organizing thrombi. It is a rare benign condition simulating angiosarcoma hence it is being reported here.

Case Report:
A 21 years old male presented in surgical OPD with verrucous growth on left forearm. Physical examination revealed no other abnormal finding. The vital signs were all normal.

All laboratory findings were within normal limit. Exision of tumour mass was done and sent for histopathology examination.

On gross examination several irregular black to brownish tissue pieces collectively measuring 1x0.8x0.5c.m. were received.

Histological examination revealed large to medium sized blood sinuses with organising thrombus, within which anastomosing vascular proliferating channels were also seen. These channels were lined by plump endothelial cells. At places, papillary appearance with fibrinous and hyaline stalks were also seen. After extensive examination in multiple sections, necrosis, bizarre cells or atypical mitosis were not seen. After exclusion of a malignant vascular lesion, a diagnosis of intravascular papillary endothelial hyperplasia (Masson’s hemangioma) of the fore arm (Fig. I & Fig. II) was made.

Corresponding Author: Dr. Meenai FJ, “Falak Villa” Riyaz Manzil Compund Adjacent to Khanu Gaon, VIP Road, Bhopal
Phone No.: 91-9826665344
E-mail: zafarah@rediffmail.com

Fig. I : Dilated blood vessel with intravascular papillary formation (H & E, 100X).
Discussion:

Intravascular papillary endothelial hyperplasia or Masson’s hemangioma is now considered as benign intravascular process that have resemblance to a angiosarcoma. Masson (1861) considered the lesion as a neoplastic condition as it was a proliferative lesion of endothelial cells. Clearkin & Enzinger (1976) termed this lesion as “intravascular papillary endothelial hyperplasia” (IPEH). There are numerous synonyms for IPEH such as intravenous atypical vascular proliferation, intravascular angiomatosis, intravascular endothelial proliferation, Masson’s pseudoangiosarcoma and Masson’s lesion (Murugaraj et at, 2010; Guvena et al, 2008). However, its exact pathogenesis remains uncertain. Gupta et al (2005) suggested that some of the hyperplastic endothelial cells in IPEH are vascularized and granulation tissue is formed during the process of organization of thrombus. In the present case lesion occurred in thrombosed vein of forearm. Barr et al (1978) observed that IPEH has characteristic exuberant endothelial proliferation within the lumen of medium sized veins. Microscopically, the tuft like papillary proliferation of endothelial cells was always intimately associated with a thrombus. It seem to represent a peculiar structure and exuberant endothelialization of intravascular papillary endothelial hyperplasia which necessitate ruling out angiosarcoma. The following features are important in the differential diagnosis:

1. Intravascular papillary endothelial hyperplasia is often well circumscribed or encapsulated.
2. The proliferation process is limited completely to the intravascular spaces.
3. Though the endothelial cells are hyperchromatic, extreme nuclear atypia and frequent mitotic figures are not seen.
4. Papillae are composed of fibro-hyalinized tissue of two or more endothelial cell layers without any covering.
5. Tangential sectioning may reveal pseudochannels but no irregular and anastomosing blood vessels are seen in the stroma.
6. Necrosis is an unusual finding in intravascular papillary endothelial hyperplasia.

In literature desmographic predilection were not reported so far. But Makos & Nikolaidou (2004) studied intravascular endothelial papillary hyperplasia in oral mucosa and noted that the lesion was more frequent in female than in males with females to male ratio being 1.3:1. In the present case it was in a male patient. Kuo et al (1976) and Park et al (2000) stated that IPEH occurs most frequently in the head and neck, fingers, trunk and cutaneous veins as a small firm, superficial mass with red to blue discoulourtion of the overlying skin. In the present case it was present at an unusual site ie. forearm and was presented with discoulouration of the overlying skin.

Intravascular endothelial hyperplasia in forearm is unusual and histopathology is essential to distinguish it from more alarming lesion of angiosarcoma.

References:

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