



Intravascular Papillary Endothelial Hyperplasia: The Diagnostic Challenges

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ABSTRACT

Masson's tumor or intravascular papillary endothelial hyperplasia is a benign lesion of the skin and subcutaneous tissue. It is due to reactive proliferation of endothelial cells with papillary formations related to a thrombus. It poses an interesting diagnostic differential as the clinical signs and symptoms are nonspecific. The diagnosis is based on histopathology. We report a case of Masson's hemangioma occurring on the right ring finger.

Keywords: Masson's Lesion, Intravascular Papillary Hemangioma, Angiosarcoma, Endothelial Cells

Introduction

Masson's tumor or intravascular papillary endothelial hyperplasia (IPEH) is a rare type of vascular tumor usually found in the soft tissue and skin. [1] It is most common in the extremities. Due to its common presentation as a subcutaneous soft tissue swelling, histopathology forms the gold standard for the diagnosis. It is due to the reactive proliferation of endothelial cells with papillary formations related to a thrombus.[2] The treatment is surgical removal. It is a very rare benign lesion mimicking angiosarcoma and hence it is being reported here.

Case Report

A 76-year-old female came to the surgery outpatient unit with complaints of swelling over the right ring finger for the past 2 years. Patient started to feel pain over the swelling for the past 2 weeks.

On local examination, a 2x2cm swelling was present on the dorsal aspect on the base of the right ring finger. It was soft in consistency, mobile and partially compressible. Excision biopsy was done and sent for histopathology.

On grossing, we received a single grey brown soft tissue mass measuring 1.8x1x0.5cms. On cut surface, it was encapsulated and circumscribed, soft to firm in consistency with adjacent fatty tissue; grey brown nodules measuring 0.8 cm in diameter were seen.

On Microscopy, sections show many blood vessels of varying sizes, few with organizing thrombus (Figure 1). One of the large blood vessels show intraluminal papillations with hyalinized fibrous cores lined by hobnail plump endothelial cells (Figure 2). There is no necrosis nor increase in mitosis.

Special stains were done and Van Gieson and Periodic Acid Schiff confirmed the above findings with anastomosing vascular channels with plump endothelial cells (Figure 3). The diagnosis was given as Intravascular papillary endothelial hyperplasia (Masson's hemangioma).

Discussion

Masson's tumor is a rare, benign vascular lesion that accounts for approximately 2% of all vascular tumors of the skin and subcutaneous tissue.[1] In 1923, Dr. Pierre Masson first described an intravascular papillary proliferation formed within the lumen of inflamed hemorrhoidal plexus in a patient and named it as "Vegetant intravascular hemangioendothelioma".[2]

The pathophysiology of this unique entity is still a mystery. Although first described as a neoplasm formed by proliferation of endothelial cells into the vessel lumen by Dr. Masson, this theory was challenged by Dr. Clearkin and Dr. Enzinger in the year 1976. [3] They considered that it was due to the exuberant organization and recanalization of a thrombus and coined the term "Intravascular papillary endothelial hyperplasia".[4] Recently it is hypothesized to be a reactive vascular proliferation following traumatic vascular stasis. Hence, it is now known as Masson's tumor, Masson's hemangioma and more popularly as intravascular papillary endothelial hyperplasia (IPEH).

Primary or pure form arises within a normal blood vessel (most commonly in a vein), and is often located on the fingers, head and neck, and forearms. The secondary lesion or "mixed" form arises in the presence of a preexisting vascular malformation, such as a hemorrhoidal vein, cavernous hemangioma or pyogenic granuloma and may be present intramuscularly. [5] A rare "indeterminate" form



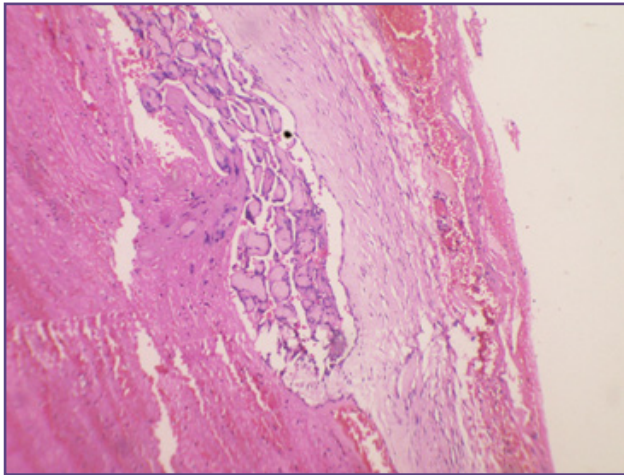


Fig. 1 : Low power view showing Large blood vessels filled with organizing thrombus showing papillations at the periphery.

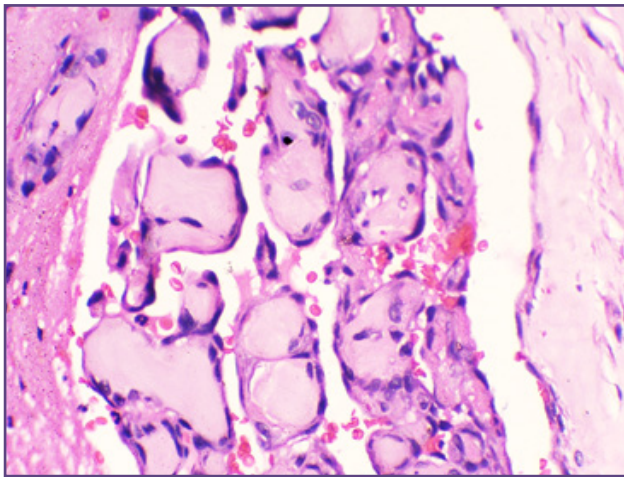


Fig. 2: High power view showing hobnail plump endothelial cells.

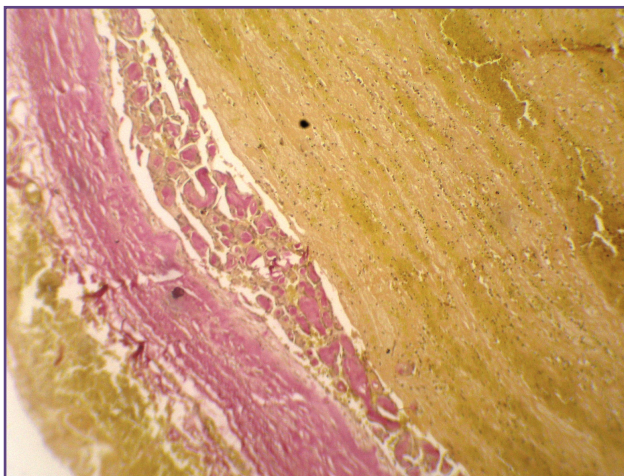


Fig. 3: Van Gieson Stain showing organizing thrombus with papillae.

that does not belong to either of the first two categories, and has an extravascular origin. [6] However, these different pathological forms do not carry any clinical importance, since the treatment is total excision.

Histopathology is essential and helpful for the diagnosis of this lesion. The papillary structure and exuberant endothelial proliferation of IPEH requires ruling out the much more common angiosarcoma.

Masson's lesion is often well circumscribed or encapsulated; the proliferative process is exclusively limited to the intravascular spaces; Papillary stalks composed of fibro-hyalinized (deeply eosinophilic) tissue of two or more endothelial cell layers. The endothelial cells are hyperchromatic, but extreme nuclear atypia and frequent mitotic figures will not be seen; There may be pseudo channels, but there are no irregular or anastomosing blood vessels in the stroma [7]; residual organizing thrombi is usually seen.

Angiosarcoma usually invades tissues outside the vascular channels. The papillae are covered by more than one or two layers of endothelial cells. Malignant features such as mitotic figures, necrosis, and nuclear pleomorphism can be seen on cytology. [8]

Immunohistochemistry may be required only if the endothelial origin of the lesion is in doubt. In such scenarios, endothelial cell markers, which would highlight the endothelial lining around the papillary tufts such as von Willebrand factor, CD31, factor XIIIa, and CD43 may be used. [9]

Simple excision is usually curative, although recurrence has been described. [10] Treatment of the lesion consists of conservative surgical excision with good outcomes in all cases except intracranial lesions, which have been reported to be fatal. [11]

Conclusion

The importance of IPEH or Masson's hemangioma lies in the fact that histopathology plays a vital role in the diagnosis of this benign lesion. It is also essential to differentiate IPEH from Angiosarcoma to avoid unnecessary surgery and radiation. [12]

Acknowledgements

We are grateful for the help of the technicians and Department of Pathology for providing support.

Funding

Nil

Competing Interests

None

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Date of Submission : 08/01/2020

Date of Final Revision : 16/02/2021

Date of Acceptance : 22/02/2021

Date of Publication : 30/03/2021

Financial or other Competing Interests: None.