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Case Report

Masson's Hemangioma of the Urethra: A Case Report

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Abstract

Intravascular papillary endothelial hyperplasia (IPEH) is an uncommon benign vascular disease characterized by endothelial cell proliferation and papillary formation within the lumen of blood vessels arising from an organizing thrombus. The occurrence of this uncommon lesion is about 2% of all vascular tumors. IPEH mostly occurs in the 5th decade of life, and there is no gender or age predilection. Nevertheless, some studies have suggested that IPEH is more common in women than in men. The clinical features are mostly asymptomatic, and the lesion is typically characterized as a small, firm, slightly elevated mass with red to blue discoloration of the overlying skin. The main diagnosis of the lesion is based on histological examination. The most common locations of IPEH are head and neck, fingers, and trunk. The occurrence of IPEH in the urethra is extremely rare, with only a few cases having been reported in the literature. We describe a 70-year-old woman with a complaint of dysuria and urethral bleeding of 3 days' duration. Clinically, the lesion was a palpable firm mass in the urethra. Urethrocystoscopy illustrated a dark mass, a few millimeters in size, with bleeding. Histologic findings revealed dilated blood vessels with multiple papillary projections into the lumen. Thus, based on these findings, a final diagnosis of Masson's tumor was confirmed. According to clinical and radiographic findings, this lesion is similar to malignant lesions and its accurate diagnosis is based on microscopic findings. Therefore, awareness of the microscopic characteristics of this tumor is very important for clinicians to rule out the presence of malignant vascular lesions and to avoid unnecessarily aggressive therapy.

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Keywords ● Thrombosis ● Endothelial hyperplasia ● Urethra ● Endothelial cells

What's Known

- Intravascular papillary endothelial hyperplasia is a benign vascular disease characterized by endothelial cell proliferation and papillary formation within the lumen of blood vessels arising from an organizing thrombus.
- Henschen depicted the lesion as a reactive process rather than a neoplasm.
- Although endothelial proliferation, which can be easily mistaken for a characteristic of sarcoma, is present, the endothelial layer of the lesion comprises normal endothelial cells, the endothelial proliferation is of benign papillae pattern, and the cells show no atypia.

What's New

- According to the reported cases so far, the lesion occurs in the fingers, head and neck, trunk, lower extremities, and upper extremities, in descending order of frequency.
- Occurrence of this lesion in the urethra is extremely rare, with only a few cases having been reported in the literature.

Introduction

Intravascular papillary endothelial hyperplasia (IPEH) (Masson's hemangioma or Masson's tumor) is an unusual benign vascular disease accounting for approximately 2% of all vascular tumors of the skin and the subcutaneous tissue.[1] IPEH was 1st described by Pierre Masson in 1923, and the term "IPEH" was 1st defined by Clearkin and Enzinger.[2] Clinically, the most common locations of IPEH are the head and neck, fingers, and trunk. Clinically, the lesion has the feature of a small, firm superficial mass with red to blue discoloration of the overlying skin.[3] IPEH is characterized by endothelial papillary structures with abundant endothelial

proliferations in normal blood vessels with an organized pattern surrounding a thrombus.[4] The clinical features are mostly asymptomatic, and the diagnosis is based on histopathology examination. The clinical and histopathological features are similar to benign and malignant vascular lesions such as angiosarcomas.[5] In IPEH, correct diagnosis is necessary to avoid subjecting the patient to unnecessarily aggressive therapy. A few cases of IPEH involving the urinary tract have been previously reported.[3] Our literature review yielded reported cases of Masson's hemangiomas in the urethra by Barua et al.[2] in 1983, Nevin et al.[4] in 2006, and Hevia[1] in 2016.

In this report, we describe a 70-year-old woman with IPEH of the urethra.

Case Report

A 70-year-old woman was referred to our hospital with a complaint of dysuria and urethral bleeding of 3 days' duration. The patient had no significant medical history and her general health condition was normal. In addition, her history was uneventful. Laboratory tests showed no abnormal findings. In clinical examination, there was a palpable firm mass in the urethra. Urological sonography revealed no abnormality. Urethrocystoscopy was performed and it illustrated a dark mass, a few millimeters in size, with bleeding. According to the clinical findings, an initial diagnosis of hemangioma was made. The patient underwent surgical excision of the mass. The resected mass measured 2.1×1.7×1.0 cm in size, and it was sent for histopathological evaluation. Histologic findings revealed dilated blood vessels with multiple papillary projections into the lumen. These papillae were lined by a single layer of plump endothelial cells with a hyalinized core. There were thrombi in the lumen (figures 1 and 2). No evidence of mitotic activity and atypia of the endothelial cell was observed. Based on the clinical and histopathological findings, a final diagnosis of IPEH/Masson's tumor was confirmed. After surgery, the patient was hospitalized for 3 days and was given antibiotics for a week. She has been under regular follow-up over the last 10 months, and there has been no evidence of recurrence. Informed written consent was obtained from the patient for this case report.

Discussion

IPEH is an uncommon benign vascular disease of the skin and the subcutaneous tissue with abundant endothelial proliferations in

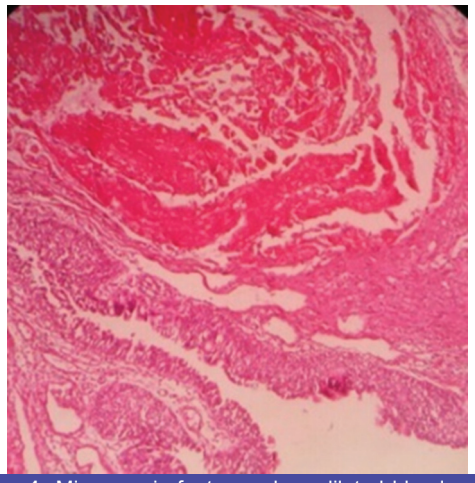

Figure 1: Microscopic features show dilated blood spaces with papillary projections into the lumen, lined by a single layer of plump endothelial cells around a fibrinous core. The lumen shows thrombi (hematoxylin and eosin, ×200).

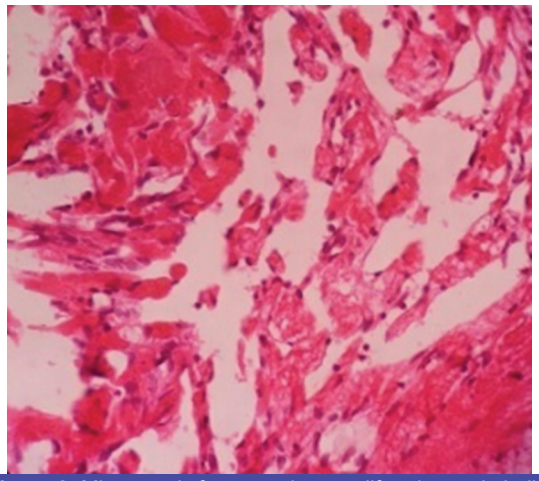

Figure 2: Microscopic features show proliferating endothelial cells with papillary projections and capillaries with blood cells.

normal blood vessels.[6] The occurrence of this uncommon lesion is about 2% of all vascular tumors.[4] The most common locations of IPEH are the head and neck, fingers, and trunk. The occurrence of this lesion in the urogenital system is extremely rare.[2] A review article showed that only 3 cases of IPEH relating to the urethra had been reported. Barua et al.[2] were the 1st to report an IPEH case in the urethra in 1983 and then Nevin et al.[4] in 2006 and finally Hevia[1] in 2016.

IPEH mostly occurs in the 5th decade of life and there is no gender or age predilection, although some studies have suggested that it is more common in women than in men.[7] Clinically, the lesion is mostly asymptomatic and it is characterized as a small, firm, slightly elevated mass with red to blue discoloration of the overlying skin. The main differential diagnoses are pyogenic granulomas, hemangiomas, lymphangiomas, thrombosed

veins, hematomas, Kaposi's sarcomas, and angiosarcomas.[8] Ultrasonography, advanced imaging modalities such as computed tomography and magnetic resonance imaging, and color Doppler sonography can identify vascular lesions. There are 3 variants of IPEH: 1) primary (intravascular), 2) secondary (mixed), and 3) extravascular (unusual type).[9] Histologically, this lesion is well-circumscribed and it is characterized by a papillary proliferation of endothelial cells forming vascular channels. The papillary structures projecting into the lumen are usually associated with thrombi. These papillae are lined by single layer of plump endothelial cells with a hyalinized core.[7,10] Histopathologically, IPEH can be mistaken for an angiosarcoma. Immunohistochemistry could be useful for the differentiation of IPEH from angiosarcomas.[5] CD105 (endoglin) staining is highly expressed only in angiosarcoma-associated endothelial cells.[7] Also, IPEH reacts with vimentin, α-SMA, factor VIII, factor XIII A, CD31, CD34, and ULEX confirmed lesions having a mesenchymal origin. The pathogenesis of IPEH is still unclear. Previous research has suggested a few possible mechanisms for the pathogenesis of IPEH: 1) degeneration and necrosis in the manner of a red infarct, 2) a thrombus as a variant of angiolymphoid hyperplasia with eosinophilia, and 3) pseudotumoral lesion formation by an accumulation of thrombotic material.[8,9] Various terms are used to describe this lesion such as papillary fibroendothelioma, intravascular endothelioma, papillary proliferation of the endothelium, papillary endothelioma, hémangioendothéliome végétant intravasculaire, l'endovasculite proliférante trombopoiétique, intravenous atypical vascular proliferation, intravascular angiomatosis, IPEH, Masson's vegetant intravascular hemangioendothelioma, Masson's pseudoangiosarcoma, intravascular endothelial hyperplasia, Masson's lesion, and papillary endothelial hyperplasia.[11]

IPEH has a good prognosis and the best treatment is a simple excision biopsy. Additionally, other methods are drawn upon to treat this lesion such as sclerotherapy, endoscopic surgery, and β-adrenergic antagonist nebivolol.[12] The recurrence of IPEH is extremely rare, and the follow-up lesions do not show evidence of metastasis.[11]

Our patient underwent surgical excision and for the last 10 months since then she has exhibited no evidence of recurrence at regular follow-ups.

Conclusion

Clinically, Masson's tumors of the urethra may resemble urethral stones and benign or malignant vascular lesions. The gold standard diagnosis of IPEH is histopathological features; therefore, awareness of microscopic characteristics of this tumor is very important for clinicians to rule out the presence of malignant vascular lesions and to avoid unnecessarily aggressive therapy.

Conflict of Interest: None declared.

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