Intravascular papillary endothelial hyperplasia

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Summary

A case of intravascular papillary endothelial hyperplasia occurring in a 35 year old female is reported. The diagnostic histological features, differential diagnosis, and aetiology of this uncommon benign endothelial proliferation are discussed.

Introduction

Intravascular papillary endothelial hyperplasia also known as intravascular angiomatosis, was first described by Masson in 1923[1], who regarded it as a true neoplasm displaying degenerative changes including necrosis and thrombosis, and designated it, "vegetant intravascular haemangioendothelioma"[2]. It is currently thought to represent an unsual form of organizing thrombus[2]. It is important to distinguish this benign lesion from an angiosarcoma. To the best of our knowledge, this is the first report of this entity in Sri Lanka.

Case report

A 53 year old hypertensive female, presented with a soft lump on the dorsum of the first web space of the right hand, of one years duration. She gave a history of a needle prick at the site of the lesion. It was excised once, but recurred.

The macroscopy specimen consisted of an apparently encapsulated brownish mass, measuring 36 × 24 × 8 mm. The cut surface was congested and displayed numerous patent vascular spaces. A central paler area of approximately 4 mm in diameter was present.

The H & E stained sections revealed a lesion composed of a thin fibrous pseudo capsule, enclosing anastamosing vascular channels, and numerous small delicate papillary tufts, with central hyalinized cores. These were lined by a single layer of plump endothelial cells devoid of pleomorphism and mitotic activity. The intervening stroma was composed of hyalinized connective tissue which was more abundant towards the centre of the lesion, where there were only a few slit like vascular spaces.

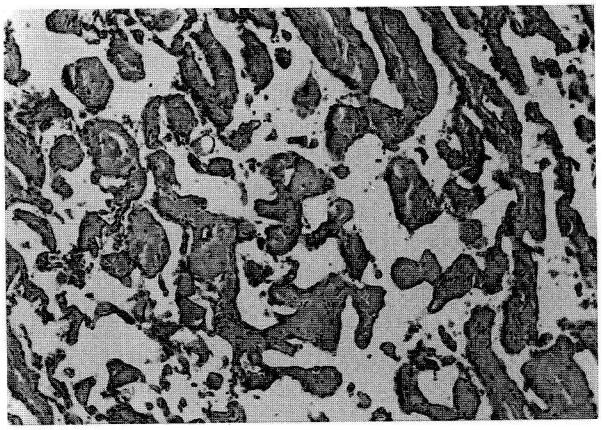

Figure 1. Papillary tufts with hyalinized cores, lined by endothelial cells (H & E × 100)

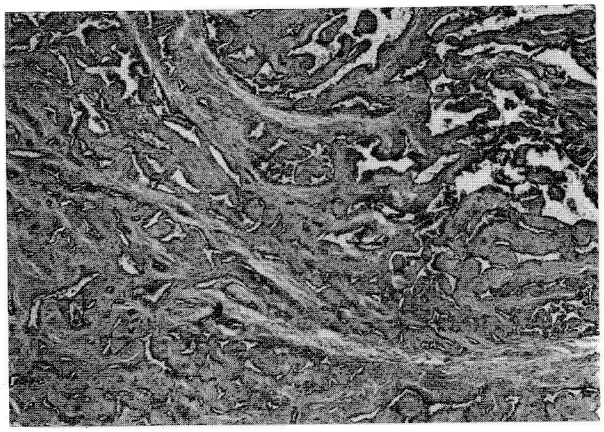

Figure 2. Papillary tufts and hyalinized area with vascular spaces (H & E × 100)

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Discussion

The histological features were diagnostic of intravascular papillary endothelial hyperplasia. Intravascular papillary endothelial hyperplasia is a benign endothelial proliferation which is thought to represent exuberant organization and recanalization of a thrombus. It may arise in previously normal vessels or in pre-existing vascular lesions, such as varices, haemorrhoids, haematomas, pyogenic granulomas, haemangiomas, and angiosarcomas[3]. A history of trauma although not usually elicited is present in this case. Most lesions are intimately associated with thrombus material which in this case is represented by the pale hyalinized central area. In the early lesion the ingrowth of the endothelium along contours of the thrombus partitions it into coarse papillae with fibrous cores. In the well established lesion numerous small declicate papillae project into the lumen, which later clump and fuse to form an anastamosing network of vessels[2].

It is important to distinguish this lesion from an angiosarcoma which it may simulate due to the presence of papillary structures, anastamousing vascular channels and plump endothelial cells. The intravascular location, and absence of necrosis, bizarre cells and atypical mitoses favour the diagnosis of intravascular papillary endothelial hyperplasia[3]. As the prognosis of intravascular papillary endothelial hyperplasia is excellent, and cure achieved by simple excision[2], this distinction is of utmost importance. The recurrence of the lesion in this patient, following initial surgery is unsual. It could have been due to incomplete excision or to the presence of an underlying vascular lesion. There was however, no evidence of the latter in the specimen that we received.

References

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