Intravascular Papillary Endothelial Hyperplasia (Masson's Tumor) of the Mouth – A Case Report

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Key Words

Intravascular endothelial hyperplasia · Mouth · Masson's tumor · Oral mucosa · Angiosarcoma

Abstract

We report a rare case of intravascular papillary endothelial hyperplasia (IPEH) of the oral mucosa. This neoplasm, known as Masson's tumor, is an unusual vascular lesion of proliferating endothelial cells. It is usually confined to the lumen of preexisting vessels or vascular malformations. The principal significance of IPEH is its microscopic resemblance to angiosarcoma and possible misdiagnosis as such. Achieving a correct diagnosis is essential to avoid subjecting a patient to unnecessarily aggressive therapy. For this reason, awareness of this lesion is very important for dermatologists and dentists. In this article, we discuss the clinical features, histopathological characteristics, and management of IPEH and review the pertinent literature.

Introduction

Intravascular papillary endothelial hyperplasia (IPEH) is an unusual benign vascular lesion comprising approximately 2% of the vascular tumors of the skin and subcutaneous tissue [1]. It was first described by Masson [2] in 1923 as a 'hemangioendotheliome végétant intravasculaire', and has subsequently been described by a variety of names. The term intravascular papillary endothelial hyperplasia is the most descriptive and least confusing and is the one most frequently used in the English literature [3].

Although the most common sites of IPEH are head and neck, fingers and trunk, these tumors may occur in any blood vessel. However, occurrence of IPEH in the oral cavity is extremely rare. A review in the accessible literature showed less than 80 cases of IPEH in the oral mucosa and lips.

Although benign, this lesion is clinically important because it presents as a mass lesion that may be mistaken histologically for angiosarcoma, and it tends to recur if incompletely resected [4, 5]. Correct diagnosis of the lesion is essential to prevent overly aggressive treatment.

In this article, we present a new case. Furthermore, we discuss the clinical features, histopathological characteristics, and management of IPEH and review the pertinent literature.

Case Report

A 70-year-old woman presented to the Stomatology service of the Dermatological Institute of Guadalajara for an asymptomatic, unique, nodular lesion of approximately 1 cm in diameter in the right side of the internal mucosa of her cheek. The lesion was slightly elevated with an oval form, firm consistency, and red-bluish color ([fig. 1](#)). She attributed the lesion to injury due to the constant friction in that zone. The patient had no family history of similar lesions.

The patient underwent an excisional biopsy. The sample was fixed in formol, and subsequent histopathology revealed a Masson tumor. Microscopically, the most prominent features of the lesion were papillary projections within dilated vascular spaces ([fig. 2a](#)). The papillary projections, which were composed of plump endothelial cells around a fibrinous core, were either attached to the vascular wall or floated freely in the lumen ([fig. 2b](#)). In the deeper areas, proliferating endothelial cells formed solid islands. They showed no significant nuclear atypia, hyperchromasia or mitotic figures and no necrosis or invasion of surrounding tissues. In the adjacent areas, an organizing thrombus was observed.

Three months after resection, the patient had no evidence of recurrence.

Discussion

Three different types of IPEH have been reported: (a) a primary (pure) form where changes are observed in a distended vessel; (b) a secondary (mixed) form that occurs in preexisting varices, hemangiomas, pyogenic granulomas, or lymphangiomas; and (c) an uncommon type in an extravascular location [1].

The pathogenesis of IPEH is poorly understood. One possible mechanism is a benign neoplastic process involving endothelial cell proliferation and papillary formation in the vascular lumen that undergoes degeneration and necrosis in the manner of a red infarct. Alternative mechanisms include a benign endothelial proliferation arising from a thrombus as a variant of angiolymphoid hyperplasia with eosinophilia; a reactive process of endothelial cells induced by blood stasis and perivascular inflammation; and a pseudotumoral lesion caused by endothelial proliferation with papillary formation proceeded by an accumulation of thrombotic material, which serves to facilitate development of the lesion [6].

The benign behavior of these lesions is emphasized throughout the literature. The vast majority of lesions present as a slowly growing mass that can be cured by local excision [4, 6].

Ki-67 (MIB-1) is a large nuclear protein preferentially expressed during all active phases of the cell cycle but absent in resting cells. Avellino et al. [4] have demonstrated the

presence of a small number of Ki-67 (MIB-1) positive cells in IPEH tumors by immunohistochemistry, suggesting that these are slow-growing benign lesions and not reactive growths.

IPEH of the oral mucosa and lips occurs more commonly in females than males [3]. A possible hormonal role has been suggested based on this gender difference, and local angiogenic growth factors may contribute to endothelial proliferation [3]. This lesion is more common in older patients (6th decade of life) [1, 7, 8]. Our patient, a 70-year-old female, is a typical example.

In the oral cavity, IPEH presents as a slow-growing, firm, reddish-blue mass with slight elevation. The most frequent locations are the lower lip, tongue, buccal mucosa, upper lip, mandibular vestibule and angle of the mouth [3].

The lesion has been clinically mistaken for mucocele, hemangioma, lymphangioma, hematoma, Kaposi sarcoma, hemangioendothelioma, thrombosed vein, traumatic fibroma, pyogenic granuloma, angiosarcoma and salivary gland tumor [3]. Since these growths lack highly specific clinical characteristics, the final diagnosis can only be made after biopsy and microscopic examination [8]. Histologically, IPEH is characterized by a papillary proliferation of endothelial cells forming vascular channels, commonly associated with thrombus (fig. 2a). IPEH may be clinically and histopathologically mistaken for an angiosarcoma. Although hematoxylin and eosin staining is sufficient for the diagnosis of IPEH confined to a dilated vessel, the CD105 (named endoglin, a transmembrane protein that is highly expressed on human vascular endothelial cells) immunohistochemistry technique could be helpful in the case of extravascular location of the IPEH lesion. CD105 staining will differentiate IPEH from angiosarcoma, since this molecule is overexpressed only in angiosarcoma-associated endothelial cells [8, 9]. The best treatment is a total excision-biopsy with healthy margins. When resected completely, recurrence is extremely rare.

Conclusions

This lesion is clinically important because it presents as a mass lesion that may be mistaken histologically for angiosarcoma. Awareness of this lesion will prevent incorrect diagnoses and overly aggressive treatment. Thus, it is very important for dermatologists and dentists to recognize this lesion.

Fig. 1. The lesion was a firm, reddish-blue, slightly elevated mass in the oral mucosa resembling a hemangioma, mucocele or Kaposi sarcoma.

Fig. 2. Histological features of IPEH, showing: **a** papillary projections that are composed of plump endothelial cells around a fibrinous core within dilated vascular spaces (hematoxylin and eosin, 200×); **b** proliferating endothelial cells that form solid islands in the deeper areas (hematoxylin and eosin, 400×).

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