Intravascular Papillary Endothelial Hyperplasia on the Hard Palate in a Young Korean Female Patient: a case report and literature review

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Intravascular papillary endothelial hyperplasia (IPEH) is a rare benign vascular lesion that rarely occurs in the oral cavity. Its clinical features are similar to neoplasms, which are easily identified in the oral cavity, and it can be misdiagnosed as an angiosarcoma. Therefore, it is important to recognize the characteristics of the lesion in order to both diagnose and treat it properly. We report a case of IPEH in a young Korean female patient, as well as discuss its differential diagnosis and treatment using a review of the related literature.

Key words: Hyperplasia, Hard palate, Neoplasm, Angiosarcoma

I. INTRODUCTION

Intravascular papillary endothelial hyperplasia (IPEH) is a rare, benign vascular lesion often caused by traumatic vascular stasis. Intravascular papillary endothelial hyperplasias have been reported to occur in the skin, subcutaneous tissue, thyroid, orbit, internal auditory canal, and parotid gland, but few cases have been reported in the oral cavity.

The clinical features of IPEH are similar to those of the neoplasms frequently found in the oral cavity. As IPEH rarely occurs in the oral cavity, however, it can be misdiagnosed as a neoplasm. In addition, because the growth pattern of IPEH is similar to that of soft tissue sarcomas such as angiosarcoma, the misdiagnosis of IPEH as a malignancy can lead to unnecessarily aggressive treatment. Therefore, an accurate diagnosis of IPEH is important for both oral and maxillofacial surgeons and pathologists.

In this paper, we report a case of IPEH that occurred on the hard palate of a young Korean female patient, discuss its differential diagnosis and treatment, and provide a review of the related literature.
Fig. 1. Clinical image showing a red, soft mass on the midpalatal area.

Fig. 2. Panoramic radiograph shows no remarkable pathologic findings.

Fig. 3. Clinical image obtained immediately after excisional biopsy.

Fig. 4. Excised mass.

Fig. 5. A. Microscopic examination reveals extensive intravascular proliferation with papillary in-growth, (H&E staining, x40) B. Higher magnification shows papillary fronds (long arrows) and endothelial cells (arrow heads). Red blood cells are seen, indicating a vascular lumen, (H&E staining, ×200)

Fig. 6. Three-month follow-up clinical image shows no recurrence.
II. REPORTS OF CASE

A 14-year-old female patient visited our clinic complaining about a mass that had formed 2 years earlier on the hard palate. Examination revealed a 1.5-cm pinkish papillomatous mass in the mid-palatal area (Fig. 1). The patient had not felt pain from the soft mass. The patient's medical history was unremarkable, and there were no abnormal findings in the panoramic radiograph (Fig. 2). Since the mass had not changed in size since it formed, we assessed that the probability of malignancy was low and recommended periodic follow-up. At the 1-month follow-up visit, however, the patient complained of dislodged small particles and bleeding during tooth brushing. We thus decided to remove the lesion to both treat the patient's complaint and make an accurate diagnosis. The mass was excised completely (Figs. 3, 4), with no abnormal bleeding during excision, and we applied hyaluronate gel to the excision site with a wafer.

The initial clinical impression was papilloma or pleomorphic adenoma based on the shape of the mass and the presence of many minor salivary glands on the palate. In the histopathological examination, extensive intravascular proliferation with papillary ingrowth was observed (Fig. 5A); higher magnification showed papillary fronds, endothelial cells, and red blood cells, indicating a vascular lumen (Fig. 5B).

The pathologic diagnosis was IPEH. There was no recurrence as of the 3-month follow-up after excision (Fig. 6), but we plan to perform long-term follow-up for this patient.

III. DISCUSSION

Intravascular papillary endothelial hyperplasia, first described by Pierre Masson in 1923, has also been designated as Masson’s tumor, Masson’s lesion, and Masson’s pseudoangiosarcoma. It seldom occurs in the oral cavity, especially on the hard palate. Previous reports have reported IPEH occurrence on the skin, subcutaneous tissue, fingers, and trunk, but only 5.8% of vascular malformations of the oral cavity were diagnosed as IPEH. According to Sarode et al, there has only been one reported case of IPEH on the hard palate. Therefore, we consider this case unusual and worth reporting.

Its rarity and similarity to other common neoplasms in the oral cavity can make the exact diagnosis of IPEH difficult. Intravascular papillary endothelial hyperplasia manifests as a soft to firm, painless reddish-blue mass approximately 0.5 to 1.8 cm in diameter. The lesions show a clear margin against the adjacent mucosa and are slightly raised. Mostly solitary nodules can be seen without imaging. The mean age of reported occurrence is usually during the third to fourth decade, but reports vary. The growth rate is slow, ranging from 15 days to 6 years, and 7% of reported lesions are related to minor trauma. Such characteristics of IPEH are similar to common neoplasms in the oral cavity such as traumatic fibroma, papilloma, and mucocele, and the growth pattern resembles that of angiosarcoma. In this case, we first assumed the mass was a papilloma, considering the color, clear demarcation, and slow growth rate, as well as a pleomorphic adenoma based on the high instance of minor salivary glands on the hard palate. Intravascular papillary endothelia hyperplasia can be related to traumatic vascular stasis, but the patient reported no specific trauma history. Consequently, it was difficult to diagnose the lesion through patient history and initial clinical examination.

Imaging tools play little role in the diagnosis of IPEH. Lee et al, in studying the relationship between the imaging and the pathological findings of IPEH, concluded that findings from magnetic resonance imaging (MRI) of the pure form of IPEH are nonspecific, particularly since they
can be confused with sarcoma due to imaging enhancement\textsuperscript{2}. A case of IPEH in the ramus of the mandible presented as a well-circumscribed, expansile osteolytic cystic mass with peripheral enhancement\textsuperscript{3}\textsuperscript{)}, whereas computed tomography (CT) scans showed homogenous, non-homogenous, contrast enhancing, and non-enhancing masses\textsuperscript{1).} Sonography can help surgeons assess the vascularity of the lesion, but plays little role in diagnosis\textsuperscript{14).} We did not perform imaging studies in this case because the small lesion size counterindicates the use of imaging techniques. That being said, MRI or sonography can help diagnose IPEH in cases involving large lesions or when there is a high risk of intraoperative bleeding.

Histopathological examination is crucial for the differential diagnosis of IPEH. Intravascular papillary endothelia hyperplasia shows multiple papillary fronds lined by either a single or double layer of proliferating endothelial cells; cores of papillae are composed of fibrous connective tissues\textsuperscript{1);} thrombi and red blood cells are present; and hemosiderin can be observed in the connective tissue cores\textsuperscript{10).} Such characteristics of IPEH are pathognomonic and can aid in the differential diagnosis between IPEH and other neoplasms such as papilloma and pleomorphic adenoma. While several reports have indicated that IPEH can sometimes be confused with angiosarcoma\textsuperscript{2,5,5,8),} IPEH shows specific histopathological features that distinguish it from angiosarcoma: (1) lack of cellular anaplasia, mitotic activity, pleomorphism, and infiltrative pattern of growth; (2) proliferation limited to the intravascular space; (3) presence of thrombi covered by no more than two layers of endothelial cells in association with the papillae; (4) hyperchromatic endothelial cells with extreme nuclear atypia; and (5) absence of tissue necrosis\textsuperscript{1).}

Intravascular papillary endothelial hyperplasia can be successfully treated by simple total excision. Vascular lesions such as pyogenic granuloma or hemangioma have recently been successfully treated using minimally invasive therapies such as cryotherapy or sclerotherapy instead of total excision; IPEH has also been treated by sclerotherapy\textsuperscript{40).} In addition, a report indicate that IPEH has been successfully treated using the beta-adrenergic antagonist nebivolol to induce rapid regression of infantile hemangioma\textsuperscript{15).} Most of the IPEH cases we reviewed showed no local invasion and metastases; however, IPEH can recur if it arises in a primary vascular lesion\textsuperscript{10} or if the lesion is not excised completely\textsuperscript{17).} In our patient, we performed a total excision of the lesion, and there were no remarkable abnormal signs or recurrence at the 3-month follow-up visit.

IV. REFERENCES
