CASE REPORT

Angiomyolipoma of the palate displaying growth potential

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Summary

Angiomyolipoma of the oral cavity is an extremely rare benign hamartomatous tumour that occurs separately, unrelated to other conditions. As the name implies, the microscopic features comprise of mature adipose tissue, thick walled blood vessels, and smooth muscle cells which stain positive for smooth muscle actin. Our case involved a 54 year-old female who presented with a long-standing, slow growing, and asymptomatic mass on the palate. Medical history revealed no contributory factors to her condition, and the lesion doubled in size in a 12 month period. Surgical excision is the treatment of choice for this entity.

KEYWORDS

Angiomyolipoma; Palate; Oral

Introduction

Angiomyolipoma is a rare variant of lipomatous tumours characterized histologically by a triad of mature adipose tissue, thick walled blood vessels, and spindle-shaped or/and epithelioid smooth muscle cells.1 It is a benign neoplasm that arises primarily in the kidney and manifests either as a solitary mass or multiple lesions, with approximately one third of cases being associated with tuberous sclerosis.2 Angiomyolipomas involving the liver, the second most frequent site of occurrence, rarely show an association with tuberous sclerosis.3 Within the head and neck region, angiomyolipomas have been reported to occur in the oral cavity, nasal cavity, parotid gland and ear lobe.4–8 In this article, we present a new case of solitary angiomyolipoma occurring in the palate, which doubled in size in a 12 month period.

Case report

A 54 year-old healthy Caucasian female presented with an asymptomatic soft tissue mass located on the left hard palate adjacent to the maxillary second premolar. The patient reported a history of 20 years’ duration of this lesion, and had previously been examined independently by an oral pathologist and an oral and maxillofacial surgeon, both whom did not undertake any treatment. No specific aetiological factors could be elicited.

Clinically, the lesion measured approximately 1.0 cm in diameter and was red/purple in colour. The mass was covered by normal mucosa, felt soft on palpation, and blanched slightly on pressure (Fig. 1A). Clinical differential diagnosis included haemangioma or angiogranuloma, and the patient...
was advised to have the lesion surgically removed. The patient did not return for treatment, and attempts to contact her were unsuccessful. She presented 12 months later complaining of an increase in the size of the lesion, at which stage the mass measured approximately 2.0 cm in greatest dimension, and the overlying mucosa was ulcerated (Fig. 1B). An excisional biopsy was performed under local anaesthesia, and histopathological examination revealed a well-circumscribed lobulated solid tumour composed of lobules of adipose tissue mixed with sheets of smooth muscle and irregularly arranged, thick walled blood vessels (Fig. 2). The smooth muscle cells showed positivity for smooth muscle actin (Fig. 3) and vimentin, while S100 staining was negative. These features were consistent with an angiomyolipoma. On follow-up, there has been no recurrence.

Discussion

There have been four cases of intraoral angiomyolipoma described to date in the English literature. Additionally, five other cases have been described in the Japanese literature as reviewed by Ide et al. None of these cases were associated with tuberous sclerosis, in contrast to renal angiomyolipoma. Five cases overall have occurred in the hard palate, two cases each have occurred on the buccal mucosa and the lips, and a single case has occurred on the tongue.

We describe an additional case of angiomyolipoma occurring on the palate, which showed great growth potential in a 12 month period.

All the reported intraoral angiomyolipomas presented clinically as slow growing painless masses, and developed in adults between the fourth and eighth decades, with no sex predilection. The size of these lesions has varied from 0.5 to 4.0 cm in diameter, with patient’s reporting a duration of 2 months to 20 years. No aetiological or precipitating factors have been documented, and angiomyolipomas seem to follow a completely benign course.

The microscopic appearance of the angiomyolipoma of the oral cavity is identical to lesions that arise in the skin and nasal cavity and are grouped together as mucocutaneous angiomyolipoma. The tumour is well-demarcated and non-aggressive, distinct from the kidney lesion that has shown lymph node involvement albeit rarely. The smooth muscle cell component exhibits negativity for HMB-45 staining, in contrast to renal and hepatic angiomyolipoma which are typically immunoreactive, and HMB-45 positivity is often used as a diagnostic marker for this entity in such locations. The smooth muscle cells and vascular structures show positivity for smooth muscle actin, vimentin, and to a lesser degree desmin.

A variant of intraoral angiomyolipoma was described by Ide and co-workers in which the lesion was reportedly poorly circumscribed and not encapsulated. The presence of angiomatosus proliferation similar to cellular angiolioma...
was also demonstrated. They have recommended the designation of angiomyolipomatous hamartoma instead, because of a lack of typical histological features of oral angiomyolipoma. In our case, the tumour mass was encapsulated.

The histological differential diagnosis of angiomyolipoma includes angioleiomyoma, angiomyoma, leiomyosarcoma, rhabdomyosarcoma, liposarcoma, and other reactive processes, such as fibrolipomatous hyperplasia. The histologic resemblance of mucocutaneous angiomyolipoma to angioleiomyoma is controversial. Adipose tissues have been identified in angioleiomyoma, with only 3% of angioleiomyomas containing fat, and all arising in the head region, with adipose tissue representing less than 1% of the tumour mass. The predilection of angioleiomyoma for the dermis and subcutis further complicate the distinction between these tumours.

Although not much is currently known about oral angiomyolipoma, additional cases will contribute to further understanding the clinical and histopathologic features of this rare tumour.

References