



Mucoepidermoid carcinoma of the palate in a child

Catherine M. Flaitz, DDS, MS

Dr. Flaitz is a professor, Oral and Maxillofacial Pathology and Pediatric Dentistry, Department of Stomatology, University of Texas at Houston Health Science Center Dental Branch, Houston, Texas. Correspond with Dr. Flaitz at cflaitz@mail.db.uth.tmc.edu

Abstract

Salivary gland tumors are rare in children but when they involve the minor salivary glands, there is an increased risk that they will be malignant. The clinical and histopathologic features of a palatal mucoepidermoid carcinoma in an 8 year-old boy are presented. Differentiating this entity from common reactive and benign neoplastic lesions is discussed in order to prevent a delay in diagnosis and the potential for mismanagement. (Pediatr Dent 22:292-293, 2000)

Salivary gland tumors in children are rare with less than 5% of all these tumors occurring in this age group. The most common types of salivary gland tumors of epithelial origin are the pleomorphic adenoma and the mucoepidermoid carcinoma. These tumors are usually found in the parotid gland, while only a few pediatric cases have been reported intraorally. The purpose of this case report is to illustrate an example of a low grade mucoepidermoid carcinoma and to discuss a reasonable differential diagnosis, based on the age of the patient, history and clinical features.

Case History

An 8 year-old white boy was referred for the evaluation of a soft tissue enlargement of the palate, which had been slowly increasing in size for the past 9 months. Clinical examination revealed a localized submucosal nodule of the right posterior hard palate, measuring 2.0 X 1.5 cm (figure 1). The mucosal surface was smooth and intact with a faintly blue translucency. The asymptomatic swelling was soft and compressible with

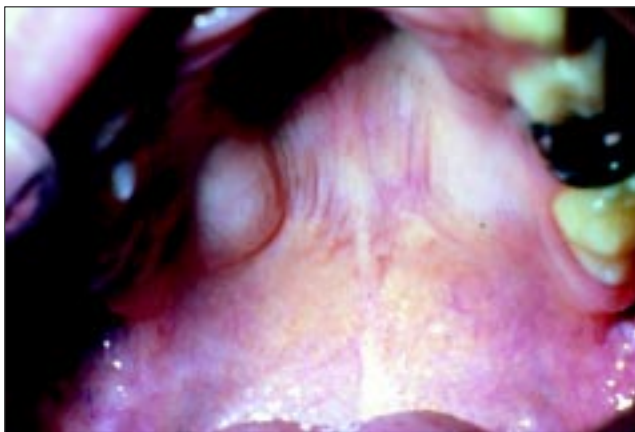


Fig 1. Mucoepidermoid carcinoma presenting as a unilateral enlargement of the posterior hard palate.

fluctuance. There was no mobility or displacement of the adjacent teeth and the median palatal raphe was clearly identified. Panoramic and periapical radiographs did not exhibit any resorption of the alveolar bone and the floor of the maxillary sinus was intact. No other abnormalities, including palpable submandibular and cervical lymph nodes, were found.

Clinical impression

Based on the persistent but slow growth pattern of this compressible palatal enlargement, a salivary gland tumor, in particular, a mucoepidermoid carcinoma should be the primary consideration in this child. The mucoepidermoid carcinoma is the second most common salivary gland tumor to occur in children, preceded only by the pleomorphic adenoma¹. This malignant salivary gland tumor occurs most frequently in the parotid gland, followed by the submandibular gland. When this tumor involves the minor salivary glands, it is the palatal region that is the most common site². Most mucoepidermoid carcinomas of childhood are diagnosed between 10 and 16 years of age with girls most often affected^{1,3}.

Intraorally, the typical clinical presentation of this malignancy is a painless, persistent enlargement, which is present for about a year. When the major salivary glands and tongue are involved, pain, paresthesia, and difficulty with swallowing are noted more frequently. Intraoral lesions appear as a localized fluctuant nodule with a bluish or reddish-purple, smooth, mucosal surface. In some cases mucus may be discharged from the tumor through a small sinus tract. Although most low grade tumors are soft and compressible, high grade lesions may be quite firm. Ulceration, resorption of bone and numbness of adjacent teeth are associated with more aggressive tumors.

Microscopic findings

These tumors are determined to be low, intermediate and high grade based on defined microscopic criteria, which are correlated with prognosis^{2,4}. Most pediatric cases of mucoepidermoid carcinoma are diagnosed as low or intermediate grade tumors^{1,3}. In the present case, the histopathologic findings revealed a low grade tumor, consisting of multicystic spaces and duct-like structures in a fibrous connective tissue. The cysts and small islands were composed of mucous, intermediate and epidermoid cells with evidence of mucus pooling (Fig 2). Mitoses were rare and cellular anaplasia, neural invasion and necrosis were not observed.

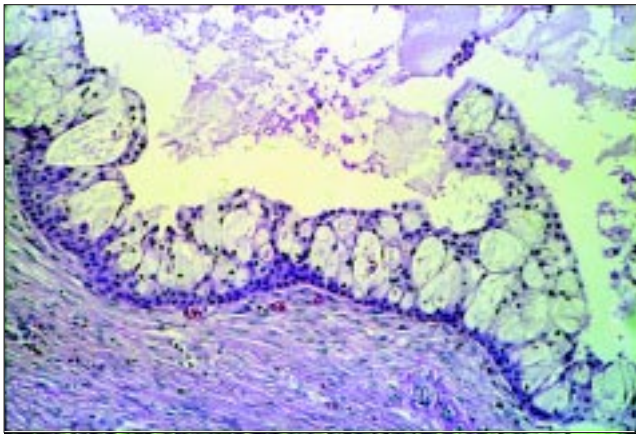


Fig 2. Photomicrograph of a low grade mucoepidermoid carcinoma, showing a cystic space with intraluminal mucus and lined by proliferating tumor cells (hematoxylin-eosin, original magnification X 100).

Treatment and prognosis

Management of the low grade mucoepidermoid of the minor salivary glands involves wide local excision with adequate tumor-free margins². If there is no evidence of bony involvement, as in the present case, the tumor is dissected down to the periosteum. When there is bony infiltration, then removal of a portion of the jaw is necessary. High grade tumors require more aggressive surgery with or without postoperative radiotherapy and chemotherapy^{3,5}. Individuals with low grade tumors have a good prognosis with greater than a 90% cure rate. In contrast, high grade tumors have a poor prognosis with only a 20 to 30% survival rate^{2,4}.

Differential diagnosis

In a child with a compressible palatal mass, reactive and neoplastic lesions should be included in the differential diagnosis. The most common of these entities is the palatal space abscess, which is characterized by a tender, diffuse, erythematous swelling of sudden onset. Typically, pulpal necrosis of a primary or permanent molar is the cause. Tooth mobility, suppuration, fluctuation in lesion size and radiographic evidence of inflammatory pulpal disease, excludes the palatal space abscess from consideration.

The mucocele is a common, reactive lesion of salivary glands that is seen frequently in children. This lesion presents as a dome shaped, translucent blue swelling that is fluid-filled. Mucoceles have a marked predilection for the lower labial mucosa, while the hard palate, retromolar pad and upper lip are uncommon sites of occurrence. Typically these reactive swellings periodically rupture and dispel the fluid contents, which results in a change in lesion size overtime. However, deeper mucoceles are often surrounded by a fibrous tissue wall and do not fluctuate significantly in size. Clinically, deep mucoceles located at sites other than the lower lip cannot be differentiated reliably from salivary gland tumors.

Since the mucoepidermoid carcinoma tends to have a blue or red surface hue, this salivary gland tumor may resemble a hemangioma. In the present case, the lack of gingival sulcular bleeding and the palatal location of the enlargement would not favor a vascular entity. Finally, benign neural tumors, including neurofibroma and schwannoma, may present as slow growing, dome shaped enlargements of the posterior hard pal-

ate. Typically these asymptomatic nodules are compressible to firm to palpation and pink in color, unless they are secondarily traumatized.

Pediatric significance

Although this salivary gland tumor represents a rare oral lesion in children, it should be considered when a lesion has a similar appearance as a mucocele but is found at a site other than the lower labial mucosa. Conservative treatment approaches advocated for the management of a mucocele by some clinicians, such as micro-marsupialization⁶, cryosurgery⁷, and laser therapy⁸ are contraindicated for this malignancy and may result in local spread of the tumor and more aggressive surgery in order to obtain disease-free margins.

Recent reports, concerning children with second malignancies, have described the development of a mucoepidermoid carcinoma in those who have received chemotherapy and cranial irradiation for a primary malignancy or aggressive disease, including acute lymphocytic leukemia and Langerhans cell histiocytosis^{3,9}. Although all reported cases have developed in the parotid glands, there is the potential risk for the intraoral minor salivary glands to be affected also.

In summary, pediatric salivary gland tumors are rare, but in contrast to adults, they are more likely to be malignant when they arise within the minor salivary glands. Clinically, this malignancy has an innocuous appearance, which mimics a reactive or benign salivary gland lesion. Significant delay in appropriate referral and treatment may occur, if the clinician does not consider this entity in the differential diagnosis of persistent fluctuant enlargements.

References

1. Luna M, Batsakis J, El-Naggar A: Salivary gland tumors in children. *Ann Otol Rhinol Laryngol* 100:869-71, 1991.
2. Auclair P, Ellis G: Mucoepidermoid carcinoma. In *Surgical Pathology of the Salivary Glands*. GL Ellis, PL Auclair, DR Gnepp EDS. Philadelphia: WB Saunders Co, 1991, pp 269-98.
3. Hicks J, Flaitz C: Mucoepidermoid carcinoma of salivary glands in children and adolescents: assessment of proliferation markers. *Eur J Cancer Oral Oncol*, 2000 (in press).
4. Hicks J, El-Naggar A, Flaitz C, Luna M, Batsakis J: Histocytologic grading of mucoepidermoid carcinoma of major salivary glands in prognosis and survival: clinicopathologic and flow cytometric investigation. *Head & Neck* 17:89-95, 1995.
5. Callender D, Franenthaler R, Luna M, Lee S, Goepfert H: Salivary gland neoplasms in children. *Arch Otolaryngol Head Neck Surg* 118:472-6, 1992.
6. Delbem A, Cunha R, Vierira A, Ribeiro L: Treatment of mucus retention phenomena in children by the micro-marsupialization technique: case reports. *Pediatr Dent* 22:155-58, 2000.
7. Twetman S, Isaksson S: Cryosurgical treatment of mucocele in children. *Am J Dent* 3:175-76, 1990.
8. Neumann RA, Knobler RM: Treatment of oral mucous cysts with an argon laser. *Arch Dermatol* 126:829-30, 1990.
9. Prasanna L, Pu A, Hoff P, Weatherly R, Castle V: Parotid carcinoma as a second malignancy after treatment of childhood acute lymphoblastic leukemia. *J Pediatr Hematol Oncol* 21:535-8, 1999.