Pediatric myoepithelioma of the palate

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ABSTRACT

Myoepithelioma is a rare benign tumor. There are controversial subtypes that lack myogenic differentiation. A 2003 literature search listed only 12 cases of myoepithelioma of the maxillofacial region. This paper describes one case of pediatric myoepithelioma as an addition to the previously documented cases, and a review of the literature.

Saudi Med J 2005; Vol. 26 (6): 999-1002

It is well known that in general, salivary gland neoplasms, benign or malignant, are rarely found in children. According to various studies, the incidence is put at ≤5%. However, a report of a large series of salivary gland tumors showed that only 37 of the 2,878 tumors (2.5%) were found in patients younger than 20 years old, of which 25 were malignant and only 5 were located in the minor salivary glands. Although myoepithelial cells are an important element in many salivary gland tumors, pure myoepitheliomas are rare, accounting for <1% of all salivary gland tumors.[1,2] Furthermore, the precise pathologic definition of myoepithelioma remains a matter of controversy due to its morphologic variations in the neoplastic myoepithelial cells. Described first by Sheldon in 1943,[3] myoepithelioma is defined as a neoplasm composed completely or almost completely by myoepithelial cells. A Medline search for reports describing myoepithelioma revealed only 12 reported cases **(Table 1)**. Even as the frequency of this rare neoplasm makes assembly of an ample amount of experience at one institution difficult, we report a case of benign myoepithelioma in the palate, verified histopathologically and confirmed by immunohistochemical and structural analysis, with a review of the literature.

Case Report. An 11-year-old boy born to Filipino parents, with no significant medical or dental history, was referred to our outpatient clinic after a lesion was found on the palate. The history was that of a painless slow-growing palatine mass, now causing some discomfort during eating. On examination, an oval lesion measuring approximately 3.5 cm in diameter was found on the right side at the junction of the hard and soft palate. The consistency was firm, painless, reddened central portion but otherwise normal mucosal surface, and immovable against the underlayer **(Figure 1).** The upper cervical lymph nodes were not palpable on either side of the neck. Routine laboratory results, including complete blood counts and liver enzymes, were within normal values. A CT scan showed a circumscribed mass of the hard and soft palate producing a swelling of the palatal vault without mucosal or bone invasion. A fine needle aspiration biopsy revealed a probable diagnosis of myoepithelioma of hyaline variant. The patient was admitted, and the following day, under oroendotracheal anesthesia, the lesion was excised with a clear margin of uninvolved soft tissue. Following excision and hemostasis, the defect was concomitantly reconstructed with buccal fat pad. The operation, as well as the postoperative course

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Received 4th December 2004. Accepted for publication in final form 19th March 2005.

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under antibiotic cover was uneventful, and the patient was discharged home after 5 days. A 2-year follow-up has shown the patient asymptomatic with no evidence of tumor recurrence **(Figure 2)**. Histologic examination showed highly cellular and large number of myoepithelial components with plasmacytoid appearance and glassy cytoplasm. The nuclei are round to oval with homogenous chromatin distribution. Fragments of intensely metachromatic stroma are noted focally, confirming the diagnosis of myoepithelioma **(Figures 3 & 4)**.

Discussion. Biologically, myoepitheliomas are benign in most cases, but occasionally infiltrate locally and metastasize. Indeed, differential diagnosis between benign myoepithelioma and malignant myoepithelioma and myeloma can be problematic unless there is precise information regarding specific diagnostic features. Myoepitheliomas have been frequently mistaken for cellular pleomorphic adenomas, as both tumors contain abundant myoepithelial cells.[4] Several investigations[1] have indicated that myoepithelioma is a variant of pleomorphic adenoma. However, the latest World Health Organization Salivary Gland Tumor Classification clearly separated myoepithelioma from pleomorphic adenoma. However, a slowly growing clinical profile and absence of pain may suggest the correct diagnosis. Various authors[5] point out that ultrasound should be the initial modality in the special investigation of salivary gland masses in children, because, in most cases, it enables differentiation between intraglandular and extraglandular lesions and may even suggest the final diagnosis. In our case, we employed a CT scan to identify the precise area of involvement. It is claimed that myoepitheliomas generally show a benign clinical course; the common presentation being that of a nonpainful swelling or mass that slowly increases in size over the course of several months to years.[4] The most frequent site of origin is the palate. However, on rare occasions, the tumor arises in the intraoral minor salivary glands with locations at the floor of the mouth[2] and tongue.[6] Other sites include soft tissue, breast, and lung.[7]

Histologically, myoepitheliomas of the salivary gland origin are classified into 3 cell types: epitheloid, spindle and plasmacytoid.[1] Occasionally epitheloid cells may have clear cytoplasm, abundant acellular mucoid stroma and hyaline cells as in our case. This is in agreement with description offered by various authors. Fine needle aspiration cytology (FNAC) is felt by some to be the most definitive procedure for pre-surgical diagnosis;[8] however, some other authors argue that although indispensable when deciding on the proper treatment, it is not fully reliable. In the present case, we were able to establish a diagnosis with FNAC, which was confirmed histologically, using immuno histochemical and structural analysis. Immunocytochemical analysis of salivary gland lesions, the presence of parathyroid hormone-related protein (PTHrP) in salivary glands and in myoepithelial cells surrounding the mucous alveoli of the glands may indicate accessory role of PTHrP in the secretory processes in the glands.[9] In myoepithelioma almost all tumor cells demonstrate metallothionein reactivity and a restricted positivity for PTHrP.[10]

The treatment in our case consisted of a wide local excision with one cm free margin, which was

Table 1 - Reported cases of myoepithelioma of the palate.

Author	Gender	Age	Site	Cell type
Kahn and Shoub 1973[11]	Female	17	Hard palate	Plasmacytoid
Luna et al 1973[12]	Female	30	Hard palate	Spindle
Sciubba and Goldstein 1976[13]	Male	22	Palate	Plasmacytoid
Nesland and Sobrinho-Simoes 1981[14]	Female	18	Soft palate	Plasmacytoid
Barnes et al 1985[15]	Female	24	Hard palate	Plasmacytoid
Enomoto et al 1985[16]	Female	58	Soft palate	Plasmacytoid
Ellyn and Grepp 1986[17]	Female	8	Soft palate	Plasmacytoid
Kawabe et al 1991[18]	Male	53	Soft palate	Plasmacytoid
Bombi et al 1996[19]	Male	31	Palate	?
Katsuyama et al 1997[20]	Female	67	Soft palate	Plasmacytoid
Kanasawa et al 1999[4]	Female	42	Hard palate	Plasmacytoid
Lopez et al 2000[5]	Male	46	Soft palate	Plasmacytoid
Present study* 2002	Male	11	Soft palate	Plasmacytoid

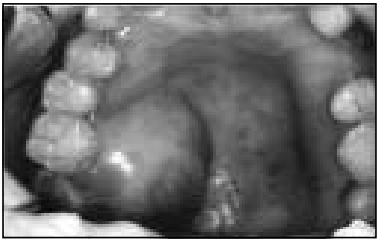

Figure 1 - Intraoral view showing a well-circumscribed palatal mass with central shallow ulceration.

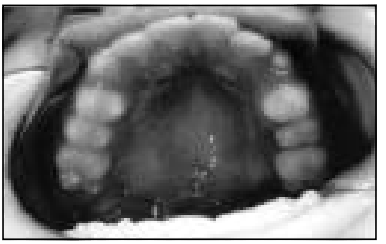

Figure 2 - Postopereative condition of palate following tumor excision and simultaneous reconstruction with buccal fat pad 6 weeks after operation. Note the highly esthetic result of defect repair.

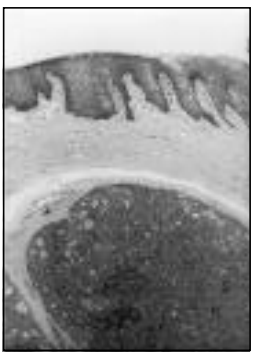

Figure 3- Photomicrograph showing tumor mass with plasmacytoid myoepithelial cells and loosely arranged intervening, acellular mucoid stroma (hematoxylin and eosin staining, magnification x 4).

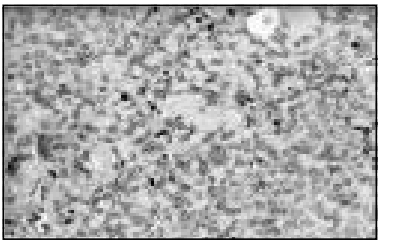

Figure 4 - Higher magnification showing majority of plasmacytoid cells, with eccentrically located nuclei (hematoxylin and eosin x 40).

monitored by frozen section, followed by closure with the use of buccal fat pad. On the palate, local palatal flaps, tongue flap or facial artery muscle-myocutaneous flap can easily be employed for closure. Two years after the operation, the patient has remained asymptomatic with no evidence of recurrence.

Acknowledgment. The authors express their grateful thanks to Dr. Abdulaziz Al-Ajlan, Consultant, Department of Pathology, Armed Forces Hospital, Riyadh, Kigndom of Saudi Arabia for his painstaking preparation and production of the histomicrophotographs. Our thanks also to Mrs. May Flores for typing the manuscript.

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