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Case Report

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Plasmacytoid Myoepithelioma Of The Palate In An Adolescent – A Case Report And Review Of Literature.

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Abstract
Myoepitheliomas are rare benign tumors that may generally arise from the minor or major salivary glands. It was considered to be a variant of pleomorphic adenoma (PA), but myoepitheliomas are now believed to be relatively aggressive tumors. This tumor should be differentially diagnosed along with several benign and malignant soft tissue neoplasms. This paper presents a case report of plasmacytoid myoepithelioma of the hard palate in a 15-year-old adolescent as an addition to the previously documented cases and review of literature.

Key Words
Myoepithelioma, plasmacytoid, hard palate, adolescent.

Introduction

Myoepithelioma is a rare benign tumor of salivary glands and represents 1-1.5% of all salivary gland neoplasms[1]. Myoepithelioma once considered a variant of Pleomorphic adenoma is now considered as a distinct pathological entity[2]. It is composed of various proportions of plasmacytoid, spindle, epitheloid, and clear cells that exhibit myoepithelial differentiation and generally lack ductal differentiation[3]. It occurs mainly in parotid gland and rarely in minor salivary glands[4].

Plasmacytoid myoepithelioma is extremely rare in children and adolescents and to our present knowledge only 7 well documented cases of myoepithelioma in patients below 18 years of age have been reported in the literature.We present this case of plasmacytoid myoepithelioma of the hard palate in a 15-year-oldadolescent because of the rarity of this tumor in this age group with the objective of contribution to the better understanding of this tumor leading to its timely diagnosis and early management. In the case reported herein, we discuss clinical, histopathlogical and immunohistochemical findings along with differential diagnosis and management.

Case Report

A 15-year - old femalereported with a swelling of six months duration in posterior part of the hard palate. The lesion was non tender, firm in consistency, measuring 3x2cm, with normal overlying mucosa w.r.t color, texture & temperature. Deglutition was normal. No regional lymphadenopathy was found. Radiographically, no bone resorption was found. Routine laboratory investigations including complete blood counts were within the normal values. No fluid was found on aspiration of the lesional mass. Fine needle aspiration cytology (FNAC) was done but the results were inconclusive. Incisional biopsy was done under LA and the histopathology report revealed myoepithelioma.

Surgical excision extending down to periosteum and including the overlying mucosa along with a margin of uninvolved healthy tissue was done under general anesthesia **(Fig. 1A-D)**.The operation, as well as the postoperative course under antibiotic cover was uneventful, and the patient was discharged home after 5 days. Histopathological report of the excised tissue confirmed the diagnosis of myoepithelioma. The immuno histochemical analysis was done and was found to be positive for S-100 protein but the results were inconclusive. The patient was followed up regularly for two years and six months with no evidence of tumor recurrence **(Fig. 1E)**.

Discussion

Tumors of salivary glands are rare [5] constituting only 3% to 5% of all salivary gland neoplasms in children and adolescents[3]. Myoepitheliomas are located most frequently in parotid gland and less frequently in minor salivary

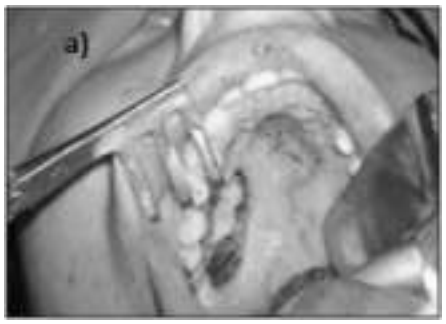

Fig 1 a) Intraoperative photograph of the patient showing surgical resection

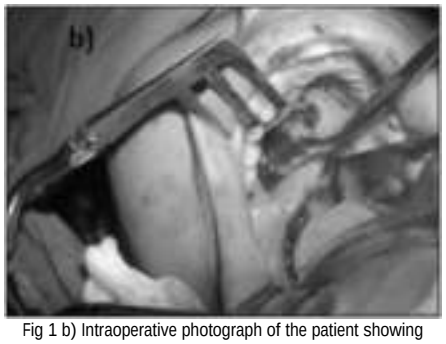

Fig 1 b) Intraoperative photograph of the patient showing surgical resection

Fig 1 c) Showing the excised tumor

Fig 1 d) Showing the excised tumor

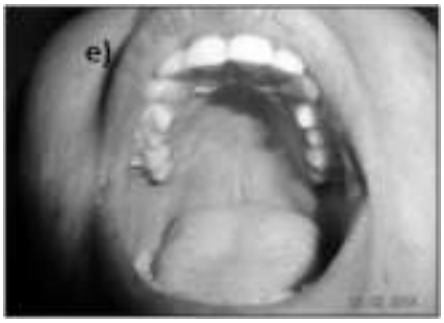

Fig 1 e) Postoperative photograph of the patient after two years and six months with no sign of recurrence

Table 1 : Reported Cases Of Myoepithelioma Of The Palate In Adolescents.

Sn	Reference	Age/Gender	Site Of Lesion	Size	Treatment
1	Kahn & Schoub 18 (1973)	17/F	Hard Palate	3.0cm	Surgery
2	Nesland Et Al 17 (1981)	18/F	Soft Palate	1.5cm	Surgery
3	Lins & Gnepp 19 (1986)	8/F	Soft Palate	1.0 Cm	Surgery
4	Arkuszewski Et Al 20 (2005)	12/M	Soft Palate	-	-
5	Nwoku Et Al 21 (2005)	11/M	Palate	3.5cm	Surgery
6	Perez Et Al 5 (2007)	13/M	Soft Palate	4.0cm	Surgery
7	Santos Et Al 2 (2011)	15/M	Hard Palate	3.5cm	Surgery
8	Present Case (2014)	15/F	Hard Palate	3.0cm	Surgery

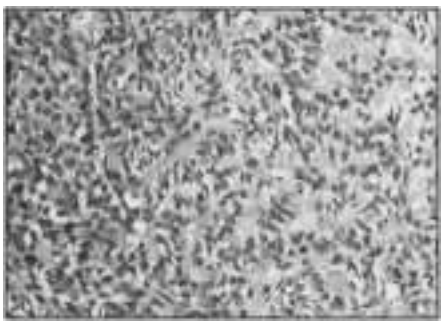

Fig.2 Histopathology Showed Tumor Cells Arranged In Nests And Cords With Intervening Hyalinized Stroma. (H&E × 200)

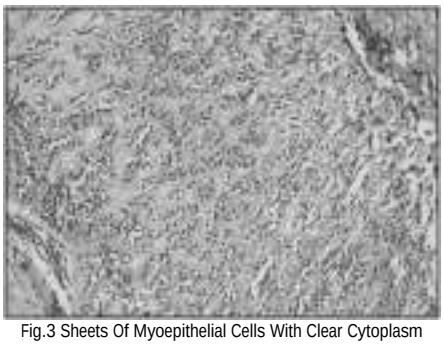

Fig.3 Sheets Of Myoepithelial Cells With Clear Cytoplasm And Occasional Plasmacytoid Cells With Eosinophilic Cytoplasm And Eccentrically Placed Nuclei. (H&E × 400)

glands of the oral cavity[6]. Myoepitheliomas are most common in the age group of 30 -50 years and very rare in individuals less than 18 years of age.No gender predilection has been reported in literature however male to female (M: F) ratio of 1: 2 was reported by Zelaya et al 4. To our present knowledge, only 7 well documented cases of myoepithelioma of palate in children and adolescents have been reported in literature. **(Demographic data presented in Table 1)**.

According to Dardick et al[7] Myoepitheliomas are benign or malignant lesions in which the tumor cells are either spindle shaped or have the range of cytological features seen in the myoepitheliomatous component of typical pleomorphic adenomas[7]. However, in myoepitheliomas there are few (less than 5%) if any luminal epithelial cells forming obvious duct-type structures seen by light microscopy. Tumors in which the ducts comprise of more than 5-10% of the sections under review are classed as pleomorphic adenomas[8]. However, Zelaya et al [4] suggested an alternative diagnostic parameter based on cellular predominance according to which, if myoepithelial predominance is found then it should be named myoepithelioma and if ductal prominence then it should be named pleomorphic adenoma[4].

The clinical presentation as found in the present case is asymptomatic, slow growing sub mucosal mass[1] with no evidence of ulceration[9],[10].Thus the clinical characteristics and overall biologic behavior of Myoepithelioma is similar to Pleomorphic adenoma[3]. Furthermore, it is also essential to know the biologic behavior of its malignant counterpart. Malignant myoepithelioma of the intraoral minor salivary glands is generally a low grade malignant tumor with little propensity for regional or distant metastasis and low recurrence[11].

Histopathologically, myoepithelioma is composed almost exclusively of sheets, islands or cords of cells with myoepithelial differentiation that may exhibit spindle, plasmacytoid, epitheloid or clear cytoplasmic features[11]. Plasmacytoid cells are polygonal cells with eccentric nuclei and dense, nongranular or hyaline, abundant eosinophilic cytoplasm. Plasmacytoid cells are found more often in tumors arising in the minor salivary glands than in the parotid gland. These hyaline cells may simulate neoplastic plasma cells, skeletal muscle or "rhabdoid" cells[10]. Although the diagnosis of myoepitheliomas should be made primarily on the basis of their histomorphology, immunohistochemical study can assist in arriving at final diagnosis, as myoepithelial cells are difficult to identify in routine histopathological examination or electron microscopy[6],[7]. In the present case, histopathological examination revealed tumor cells distributed in nests, groups that were separated by myxoid stroma. The individual cells were polygonal to round and had clear to homogeneous eosinophilic cytoplasm, at places showing sheets of plasmacytoid morphology with eccentrically placed nuclei **(Fig. 2 & 3)**.

The tumor markers recommended for diagnosing myoepithelioma are S-100 protein, Vimentin, α-smooth muscle actin (α-SMA), Cytokeratin 14 (CK14) and Glial fibrillary acidic protein (GFAP)[2],[7]. Normal myoepithelial cells show myogenic differentiation, which is revealed by the presence of actin filaments, as well as filaments of cytokeratin. However, tumoral myoepithelial cells rarely show the same cytoskeleton as normal cells therefore it is suggested that tumor myoepithelial cells might exhibit different stages of differentiation. CK 14 is a useful marker of normal myoepithelial cells that is responsible for the anchorage of myoepithelial cells to the basement membrane. It is usually unexpressed in tumor cells, unless those cells present terminal differentiation so lack of CK14 expression is supposed to be expected[2]. Myoepitheliomas show strong positivity for S-100 protein and is also positive for GFAP[2],[5],[12]. Immunohistochemical expression of vimentin may indicate that myoepithelial cells in tumors such as myoepitheliomas do not reach complete differentiation. The negativity for myogenic markers is expected in plasmacytoid variant so α SMA is

negative[2],[7]. In this case report immunohistochemical analysis was found to be positive for S-100 proteinbut the results were inconclusive as these markers are also positive for pleomorphic adenoma. Immunohistochemical staining can assist in diagnosing myoepitheliomas but histopathology still remains the gold standard for diagnosis.

Myoepitheliomas should be differentiated from benign and malignant tumors such as pleomorphic adenoma, adenocarcinoma, nerve sheath tumors, fibrous histiocytoma, nodular fasciitis, synovial sarcoma, leiomyoma, leiomyosarcoma, hemangiopericytoma, solitary fibrous tumor and paraganglioma. Furthermore, myoepitheliomas should be differentiated from malignant neoplasms [13] as the incidence of malignant epithelial salivary gland tumors is almost same as that of benign tumors in children and adolescents[5]. It can be misdiagnosed as a malignant tumor because of the variable architectural pattern with diverse cell types, and frequently increased cellularity and unfamiliar growth patterns [5],[7]. Soft tissue myoepithelial tumors with at least moderate cytologic atypia are clinically malignant [14].

The biological behavior of myoepitheliomas appears to parallel that of the pleomorphic adenoma and, as such, should be treated accordingly[10] .As performed in the current case ,surgical excision is considered the treatment of choice[5]. Most morphologically benign or low-grade myoepithelial neoplasms of soft tissue behave in a benign fashion with a low, but unpredictable risk for local recurrence(approximately20%) [14]. Although these tumors do not present high levels of the recurrence [13] but they can undergo malignant transformation especially in long standing tumors or in tumors with multiple recurrences[15]. In malignant lesions a wider excision must be planned, and lymph node dissection should be considered[16].The possibility of recurrence should be discussed with the patient and parents and regular and long term follow up should be carried out.

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