

CASE REPORT

## Plasmacytoid myoepithelioma of the soft palate: a review of the literature and report of a case with immunohistochemical findings

Yumuşak damakta plazmasitoid miyoepitelyoma: İmmünohistokimyasal bulguları ile birlikte bir olgu sunumu ve literatürün gözden geçirilmesi

Mustafa Fuat AÇIKALIN, M.D.,<sup>1</sup> Özgül PAŞAOĞLU, M.D.,<sup>1</sup> Hamdi ÇAKLI, M.D.,<sup>2</sup> Evrim ÇİFTÇİ, M.D.<sup>1</sup>

Myoepitheliomas of salivary glands are rare neoplasms, accounting for less than 1% of all salivary gland tumors. They are classified into four cell types: epithelioid, spindle, clear, and plasmacytoid. Among them, the plasmacytoid cell type appears to have a predilection for the oral cavity, especially the palate. We presented a 32-year-old man with plasmacytoid myoepithelioma arising in the soft palate, together with a brief discussion of the clinicopathologic features and differential diagnosis in the light of the literature.

**Key Words:** Myoepithelioma/pathology/surgery; palatal neoplasms/pathology; salivary gland neoplasms/pathology.

Tükürük bezlerinin miyoepitelyomaları nadir tümörlerdir ve tükürük bezi tümörlerinin %1'inden azını oluştururlar. Miyoepitelyomalar epitelioid, iğsi, şeffaf ve plazmasitoid olmak üzere dört hücre tipinde sınıflandırılır. Bunlar arasında, plazmasitoid hücrelerden oluşanlar, damak başta olmak üzere, oral kavitede yerleşme eğilimi gösterir. Bu yazıda, yumuşak damakta plazmasitoid miyoepitelyoma saptanan 32 yaşında bir erkek olgu sunuldu ve bu tümörlerin klinikopatolojik özellikleri ile ayırıcı tanısı literatür bilgileri eşliğinde tartışıldı.

**Anahtar Sözcükler:** Miyoepitelyoma/patoloji/cerrahi; damak neoplazileri/patoloji; tükürük bezi neoplazileri/patoloji.

Myoepithelial cells are found in numerous tissues, including major and minor salivary glands, secretory portion of sweat glands, around ducts of mammary glands, and Bartholin's glands.<sup>[1]</sup> Myoepithelial cells in the salivary gland are located around secretory acini and the intercalated duct, and are a component of several types of benign and malignant salivary gland tumors, particularly benign mixed tumors, adenoid cystic carcinoma, and terminal duct carcinoma.

Tumors composed entirely or predominantly of myoepithelial cells are referred to as myoepitheliomas. Myoepitheliomas of salivary glands are rare neoplasms and make up less than 1% of all salivary gland tumors.<sup>[2-4]</sup> Histologically, myoepitheliomas of salivary gland origin are classified into four cell types: epithelioid, spindle, clear and plasmacytoid.<sup>[3]</sup> Among them, the type appears to have a predilection for the oral cavity, especially the palate.<sup>[2,3,5-7]</sup>

◆ Departments of <sup>1</sup>Pathology and <sup>2</sup>Otolaryngology, Medicine Faculty of Osmangazi University (Osmangazi Üniversitesi Tıp Fakültesi, <sup>1</sup>Patoloji Anabilim Dalı, <sup>2</sup>Kulak Burun Boğaz Hastalıkları Anabilim Dalı), Eskişehir, Turkey.

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◆ Correspondence (İletişim adresi): Dr. Mustafa Fuat Açıkalın. Gültepe Mah., Üniversite Evleri, C 3 Blok, Daire 6, 26040 Eskişehir, Turkey.  
Tel: +90 222 - 239 29 79 / 4533 Fax (Faks): +90 222 - 239 37 72 e-mail (e-posta): acikalın@ogu.edu.tr

In this report, a case of plasmacytoid myoepithelioma arising in the soft palate with immunohistochemical confirmation, is described together with a review of the literature.

### CASE REPORT

A 32-year-old man presented with symptoms of upper respiratory tract infection. During the oral examination, a firm, painless, asymptomatic submucosal swelling was discovered in the right side of midline of the soft palate. The overlying mucosa was normal. There was no regional lymphadenopathy. The remainder of his physical examination was non-contributory and routine laboratory tests were within normal limits. Magnetic resonance imaging of the head and neck region revealed a well-circumscribed, solid mass in the soft palate (Fig. 1). After the clinical diagnosis of "possible minor salivary gland tumor" was made, the tumor was surgically removed with a rim of surrounding uninvolved mucosa under general anaesthesia. Postoperatively, no complications were encountered such as velopharyngeal insufficiency and fistulae. The patient was alive and well with no recurrence after eight months.

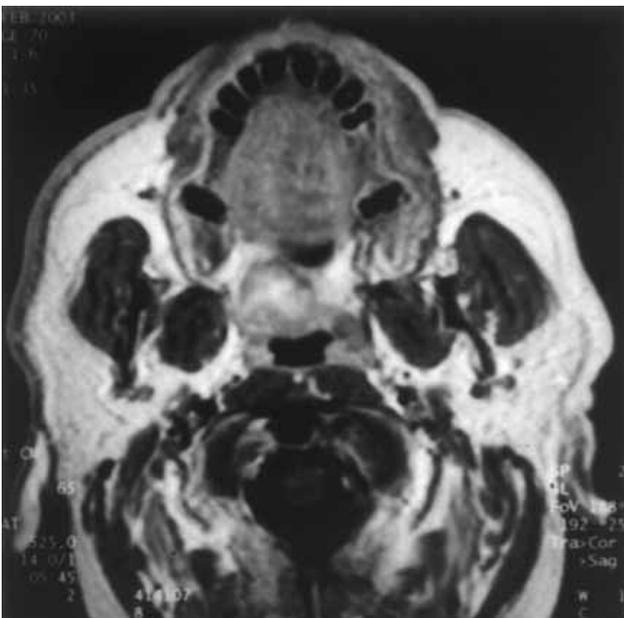
#### Pathological findings

Macroscopically, the surgical specimen consisted of a rounded tumor measuring 2.5x2.3x2.3 cm. The

tumor had a rubbery consistency. The cut surface was solid, whitish and glistening, with focal areas of hemorrhage (Fig. 2). There was no necrosis.

Microscopically, the tumor was surrounded by a thin fibrous capsule. The tumor composed entirely of plasmacytoid cell population with eosinophilic cytoplasm and eccentric, round to oval nuclei. The nuclei were generally vesicular and contained prominent nucleoli. Some cells had a large and pleomorphic nuclei; however, mitoses and necrosis were not seen. The cells were in either sheets or as loose aggregates lying in a myxoid matrix (Fig. 3a, b). The intervening myxoid stroma stained positively with Alcian blue stain. Neither ductal architecture nor areas of chondroid differentiation was observed. Destructive invasive growth was not seen. The surgical margins of the specimen were free of disease.

Immunoperoxidase staining was performed on sections of paraffin embedded tissue using monoclonal antibodies for vimentin (Neomarkers, Fremont, USA), S-100 protein (Neomarkers, Fremont, USA), cytokeratin AE1, AE3 (Neomarkers, Fremont, USA), glial fibrillary acidic protein (DAKO, Carpinteria, USA), desmin (Novocastra, Newcastle, UK), smooth muscle actin (Novocastra, Newcastle, UK), muscle-specific actin (HHF-35) (DAKO, Carpinteria, USA), and Ki-67 (using the mAb MIB-1, Novocastra, Newcastle, UK). The plasmacytoid cells were intensely and diffusely positive for vimentin and S-100 protein. Some of the tumor cells were reactive for cytokeratin and glial fibrillary acidic protein. Desmin, smooth muscle actin and muscle-specific actin (HHF-35) gave negative results. The immunoreactivity of Ki-67 was sporadic.



**Fig. 1** - Postcontrast injection Magnetic Resonance Imaging of the head and neck region revealed a well-circumscribed, solid mass in the right of midline of the soft palate.



**Fig. 2** - The cut surface of the tumor was solid, whitish and glistening, with focal areas of hemorrhage.

From these histopathological and immunohistochemical findings, the diagnosis of "benign plasmacytoid myoepithelioma" was made.

### DISCUSSION

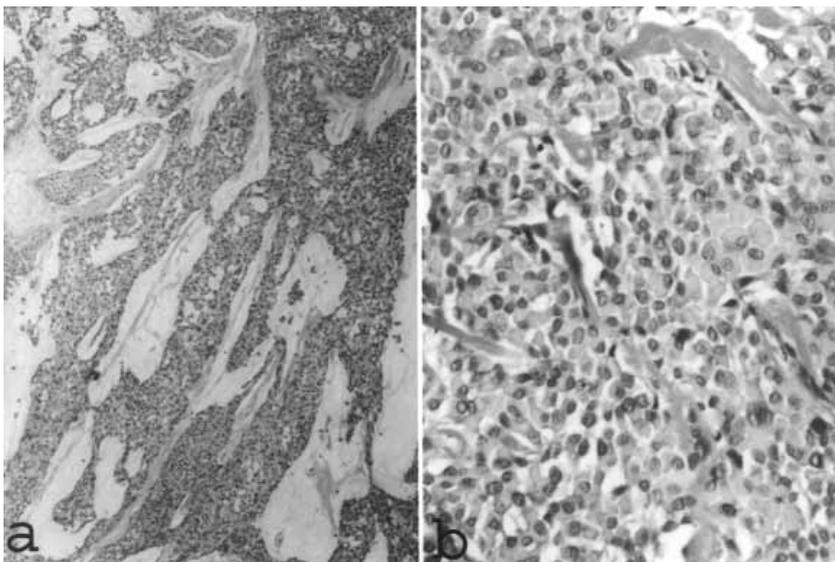
Myoepitheliomas of salivary glands are rare neoplasms and make up less than 1% of all salivary gland tumors.<sup>[2-4]</sup> Men and women are affected with equal frequency. The clinical presentation is similar to other benign salivary gland tumors—a nonpainful mass that slowly enlarges over the course of several months to years.<sup>[3]</sup>

Myoepitheliomas were first classified under benign salivary gland tumors and later as a variant of pleomorphic adenoma.<sup>[1]</sup> However, in the latest edition of the World Health Organization Histologic Classification of salivary gland tumors, myoepitheliomas are separated from pleomorphic adenoma and are classified as a distinct clinicopathologic entity.<sup>[8]</sup>

Approximately 50% of all such lesions involve the parotid gland, 40% arise in minor salivary glands, and a few affect the submandibular salivary gland.<sup>[9]</sup> According to the review of Kanazawa et al.,<sup>[3]</sup> a total of 9 cases of ultrastructurally or immunohistochemically confirmed myoepithelioma of minor salivary gland origin have been reported up until 1999, and in all of these cases, the tumors were located in the oral cavity. The most frequent site of origin of intraoral minor salivary gland lesions is the palate.<sup>[2,3,5-7]</sup>

Histologically, myoepithelioma is a tumor composed of epithelioid, spindle, clear or plasmacytoid cells with ultrastructural and immunohistochemical features of myoepithelial differentiation.<sup>[3]</sup> A single tumor may consist of myoepithelial cells showing more than one morphological type. In a large series consisting of 23 cases, the spindle cell type formed the significant majority (70%) of cases.<sup>[1]</sup> Most of the reported cases of spindle cell and clear cell myoepitheliomas have occurred in the parotid gland,<sup>[1,3]</sup> whereas plasmacytoid myoepitheliomas tend to occur more frequently in the oral cavity, especially the palate.<sup>[2,3,5-7]</sup> Among 9 cases of myoepitheliomas of minor salivary gland origin reviewed by Kanazawa et al.,<sup>[3]</sup> 8 exhibited plasmacytoid morphology. In the plasmacytoid variant, the tumors are composed of cells with eccentric nuclei, some degree of pleomorphism and hyperchromasia, but scanty or no mitotic activity. The cells are distributed in nests and groups separated by an abundant myxoid stroma that contains hyaluronic acid and lacks mucin. Chondroid areas and ductal differentiation are never found. In our case, the tumor was located on the soft palate, and the histopathological and immunohistochemical findings were consistent with plasmacytoid myoepithelioma.

Although most myoepitheliomas behave in a benign fashion, a few cases of malignant myoepithelioma have been reported.<sup>[4]</sup> The malignant counterpart of myoepithelioma may arise de novo or develop within a preexisting pleomorphic adenoma or benign myoepithelioma.



**Fig. 3 -** (a) Sheets and loose aggregates of tumor cells lying in a myxoid matrix (H-E, original magnification x 40). (b) Plasmacytoid cells population with eosinophilic cytoplasm and eccentric, round to oval nuclei (H-E, original magnification x 200).

The differential diagnosis of myoepitheliomas depends on the predominant cell type. For the spindle cell variant, the differential diagnosis includes benign fibrous histiocytoma, leiomyoma, extracranial meningioma, peripheral nerve sheath tumors, and smooth muscle neoplasms. Plasmacytoid cell type myoepitheliomas should be distinguished from extramedullary plasmacytoma, pleomorphic adenoma, and myoepithelial carcinoma. The plasmacytoid myoepithelioma can be differentiated from extramedullary plasmacytoma by the larger size of the cells and the absence of both perinuclear clear zone and cytoplasmic immunoglobulins.<sup>[2]</sup> Additionally, unlike the plasma cells, the cells of plasmacytoid myoepitheliomas are immunoreactive for cytokeratin, vimentin, S-100 protein, and in some cases, muscle-specific actin and GFAP. At times pleomorphic adenoma may be difficult to distinguish from plasmacytoid myoepithelioma. In contrast to pleomorphic adenoma, myoepitheliomas show myoepithelial but not ductal differentiation and is without chondroid or myxochondroid foci. Myoepithelial carcinoma differs from its benign counterpart by the presence of cytologic atypia, mitotic activity and infiltrative growth pattern. Occasionally, confusion may arise with well-differentiated myoepithelial carcinoma when the latter is devoid of invasion or metastases. For the problematic cases, assessment of the proliferative activity of the tumor may be helpful in establishing the correct diagnosis. In a recent study, cellular proliferative activity assessed by mitotic count and the Ki-67 labeling index was found to be significantly higher in myoepithelial carcinomas when compared with benign myoepitheliomas.<sup>[4]</sup> In this study, a Ki-67 labeling index of more than 10% or mitotic figures more than 7 per 10 high-power fields are reportedly diagnostic of myoepithelial carcinoma. In our case, some cells had a large and pleomorphic nuclei, but mitoses, necrosis, and destructive invasive growth were not seen. The immunoreactivity of Ki-67 was

sporadic. The patient is alive and well with no recurrence after 8 months.

The treatment of myoepithelioma is similar to that used for any other benign salivary gland tumors and is by total excision.

In conclusion, plasmacytoid myoepitheliomas are rare tumors and tend to occur more frequently in the oral cavity. The histologic diagnosis of plasmacytoid myoepithelioma is often problematic, however, the awareness of this entity and careful interpretation of histopathologic and immunohistochemical features are helpful to arrive at the correct diagnosis.

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