Auris Nasus Larynx 40 (2013) 231–234

Contents lists available at SciVerse ScienceDirect

Auris Nasus Larynx

journal homepage: www.elsevier.com/locate/anl

# Malignant myoepithelioma of the soft palate

Irving J. Mejía-Hernández [a], Ana Ma. Cano Valdez [b,c], Denise De León-Trenado [b], Kuauhyama Luna-Ortíz [a,c,*]

[a] Department of Head and Neck Surgery, Instituto Nacional de Cancerología, México, D.F., Mexico
[b] Department of Pathology, Instituto Nacional de Cancerología, México, D.F., Mexico
[c] Pathology, Division de Estudios de Posgrado, Universidad Nacional Autonoma de México (UNAM), Mexico, D.F., Mexico

ARTICLE INFO

*Article history:*
Received 9 August 2011
Accepted 6 April 2012
Available online 31 May 2012

*Keywords:*
Malignant myoepithelioma
Soft palate
Pleomorphic adenoma
Carcinoma

ABSTRACT

Malignant myoepitheliomas (MM) (myoepithelial carcinomas) are rare tumors representing <1% of salivary gland tumors. They are characterized as being locally aggressive. Rarely do they present distant metastases; however, when they do metastasize the sites most affected are the lungs, liver, pleura, peritoneum and skin. They may originate *de novo* in a pleomorphic adenoma or a benign myoepithelioma. We report the case of a patient with a submucosal lesion of the soft palate measuring ~4 cm × 3 cm. The patient underwent transoral resection with a microscope and $CO_2$ laser. Histopathological report was MM originating in a pleomorphic adenoma. Management of this neoplasm is controversial. Myoepithelial carcinoma is a rare neoplasm whose diagnosis includes immunohistochemical (IHC) studies. Surgery is the cornerstone of treatment. Management with laser surgery may preserve the function of the soft palate without deterioration of the quality of life in these patients.


## 1. Introduction

Multiple authors have agreed that Sheldon et al. first used the term "myoepithelioma" in 1943 to describe three cases [1–3]; however, the first description of a clearly malignant myoepithelioma (MM) is attributed to Stromeyer et al. in 1975 [4]. Myoepithelioma (also called myoepithelial carcinomas) [5] are lesions characterized by presentation of exclusively myoepithelial differentiation with three main cell patterns: epithelioid (60%), plasmacytoid (12%), spindle (8%) or a mixture of these (20%) [6]. They are rare tumors that represent <1% of salivary gland tumors and only a few fulfill the criteria to be classified as malignant. These criteria are invasion and destructive growth, cellular pleomorphism and presence of mitosis [5–10]. MM are characterized by being locally aggressive. Rarely do they present distant metastases; however, in case reports of metastatic sites, they affect the lungs, liver, pleura, peritoneum and the skin [4,5,11]. MM, as well as benign myoepithelioma, may arise *de novo* or in a previously existing pleomorphic adenoma, as well as originating from a benign myoepithelioma [1,10,12]. These tumors generally present as a painful lesion with an evolution of months to years before diagnosis [2]. They are considered to have two stages of growth: a slow-onset followed by a phase of rapid and aggressive growth [1,2,8]. The average age of presentation is ~50 years (range: 18–64 years) without gender predominance [1,3,8]. The most common primary site is the parotid gland and the most common intraoral site described is the hard palate and, with less frequency, the larynx and the soft palate [1,4,11,12].

Currently, wide resection is accepted as an adequate treatment. The value of radiotherapy is unknown [10]. Our case report fulfills the histological criteria for a myoepithelial carcinoma, and we present $CO_2$ laser resection with intraoral approach as an appropriate treatment option with adequate oncological and functional results.

## 2. Case report

We report the case of a 73-year-old female who presented with a history of a painful lesion of 3 years evolution located in the soft palate, which increased in size during the past 2 months with increasing pain and dysphagia. The patient was seen at another hospital where she underwent a CT scan in which a soft palate tumor was observed. For this reason, she was referred to our institution. On examination we corroborated the findings and observed a firm, painful submucosal lesion of 4 cm × 3 cm localized in the soft palate to the right of the midline (Fig. 1a). There was no palpable adenopathy. We performed an MRI of the head and neck, which demonstrated the same solid mass under the soft palate (Fig. 1b and c). The tumor was resected using a microscope and $CO_2$ laser with a transoral approach under general

* Corresponding author at: Department of Head and Neck Surgery, Instituto Nacional de Cancerología, Av. San Fernando #22, Col. Sección XVI, Tlalpan, México, D.F. 14050, Mexico. Tel.: +52 55 26525037.
*E-mail addresses:* kuauhyama@yahoo.com.mx, kuauhyama@starmedia.com (K. Luna-Ortíz).

0385-8146/$ – see front matter © 2012 Elsevier Ireland Ltd. All rights reserved.
http://dx.doi.org/10.1016/j.anl.2012.03.002

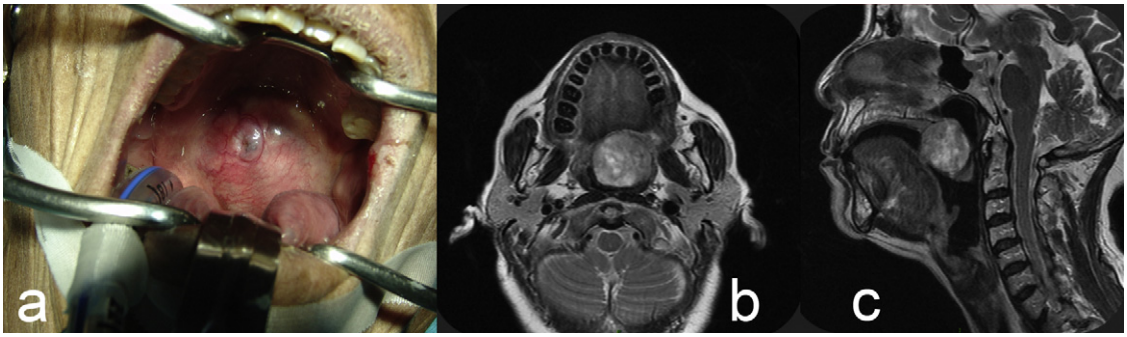


**Fig. 1.** (a) Preoperative view of the lesion. (b and c) Magnetic resonance imaging (MRI). Sagittal and axial cuts showing the lesion dependent on the soft palate.

anesthesia. There was no invasion to other sites, and the use of grafts was not necessary. Soft palate function was preserved intact (Fig. 2a). The patient showed no complications and was discharged on the same day of surgery. She had an excellent postoperative evolution with no alterations of phonation or swallowing.

## 3. Pathology

Macroscopically we found an encapsulated tumor of 4.3 cm × 3.3 cm × 3 cm weighing 20 g (Fig. 2b). The mucosa of the specimen showed normal characteristics. Upon dissection, the surface of the tumor was solid, irregular, yellowish-white in color, and with focal areas of hemorrhage. Microscopically, on routine H&E histological sections showed a neoplasm with a solid growth pattern and multinodular architecture. Tumor was composed of a polymorphous neoplastic population of spindled, stellate, epithelioid, clear and hyaline cells. Solid areas displayed foci of necrosis. Hyaline and clear cells most frequently were found in trabecular pattern. A transitional zone with ductal differentiation and chondromyxoid matrix in the same neoplasm was identified. Focal eosinophilic basal lamina material around tumoral nest was also observed. Mitotic index was low; nevertheless, it showed vascular permeation and infiltration to adjacent structures (Fig. 3a and b). The final pathology report was MM originating in the pleomorphic adenoma with vascular lymphatic permeation and necrosis. Surgical margins were free of neoplasm.

## 4. Immunohistochemistry

Neoplastic cells showed diffuse reactivity for S100 protein and focal positivity against calponin and p63, mainly in clear cell areas. Epithelial membrane antigen and cytokeratin 7 were positive in ductal component of the neoplasm (Fig. 3c and d). p53 was negative and showed no overexpression in neoplastic cells. CD-34 enhanced endothelial cells, becoming evident vascular invasion (Fig. 4a).

## 5. Electron microscopy

Ultrastructural study showed a reduplicated basal lamina between neoplastic cells, which were polygonal or stellate, with short, delicate processes and intercellular junctions. Irregular nucleus, rough endoplasmic reticulum, and filaments with focal densities were observed. Above features demonstrated myoepithelial differentiation (Fig. 4b).

## 6. Discussion

Myoepitheliomas are rare tumors of the salivary glands. Nagao et al. [5] described an incidence of 0.45% in a review of 1945 cases of salivary gland tumors that were included, for the first time, in the histological classification of the World Health Organization in 1991. The most common site of presentation is the parotid gland followed by the submandibular and minor salivary glands. Nevertheless, few cases of MM have been reported outside the oral cavity including the larynx. According to our review, only two cases have been reported in the soft palate, making this one the third case reported in the literature [1,4,11,12]. Clinical and biological characteristics of this tumor are variable. There are no histological features that correlate with its development [5]. Savera et al. [13] described that the atypical cellular structure correlates with poor results; however, other parameters such as tumor size, cell type, cytological grade, presence of a benign neoplastic precursor, mitotic index, necrosis and vascular-lymphatic permeation have proven not to be determinants in the prognosis of this disease [5,13]. DiPalma and Guzzo [1] and Nagao et al. [5] described MM as low-grade neoplasms characterized by multiple recurrences and a long history of evolution when they

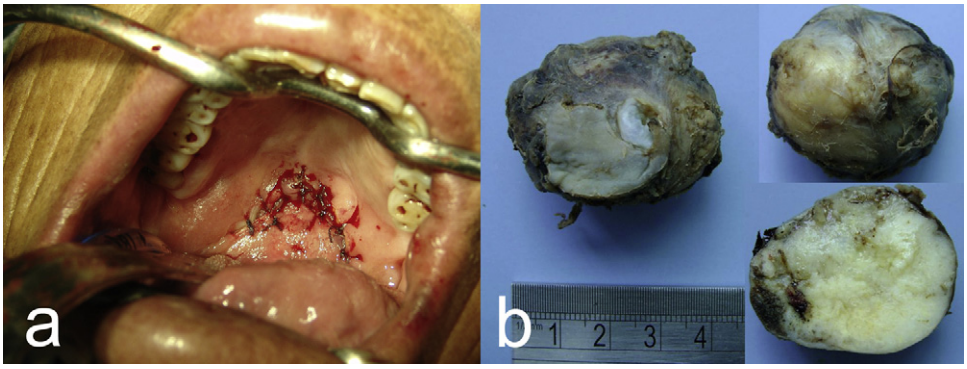


**Fig. 2.** (a) Soft palate reconstruction after resection. (b) Macroscopic view of surgical specimen of tumor (4 cm × 4 cm).