

Myoepithelioma of Soft Palate: Radiologic-pathologic Correlation

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ABSTRACT

Myoepithelioma is a very rare benign neoplasm, which is composed entirely of myoepithelial cells and accounts for <1% of all salivary gland tumors. Of all the myoepitheliomas arising from salivary glands, 26% involve the minor salivary glands of the oral cavity, where the palate is the most common origin of intraoral myoepithelioma. Final diagnosis of myoepithelioma can only be done by histopathological examination and cannot be predicted preoperatively solely on clinical or radiographic grounds. We are presenting a case of myoepithelioma of soft palate in a 35-year-old female patient with emphasis on its radiological and histopathological appearances.

Key words: Benign tumor; myoepithelioma; salivary gland tumor; soft palate

Introduction

Myoepitheliomas are rare benign neoplasm composed of ectodermally derived contractile smooth muscle cells, that is, myoepithelial cells which lack ductal differentiation. About 50% of salivary gland myoepitheliomas arise in parotid gland followed by sublingual gland (33%) and submandibular gland (13%).^[1] Myoepitheliomas arising in the oral cavity are very rare constituting about 1.5% of all salivary gland tumors. Myoepithelioma occurring in minor salivary glands of oral cavity accounts for 26% of all salivary gland myoepitheliomas with the palate being the most common site. The term myoepithelioma was first introduced by Sheldon in 1943.^[1,2] Myoepithelial cells are situated between the basal lamina and acinar ductal cells of salivary glands and other exocrine organs. Myoepithelioma classically presents as an asymptomatic mass that slowly enlarges from over a period of months to years in the patient with an average age in the third decade. However, recurrent and malignant ones have also been described.^[2,3] They are more likely to occur in the parotid gland, but rare cases arising from minor salivary glands are also reported in

the literature.^[4] We report a case of myoepithelioma of the soft palate in a 35-year-old female patient and correlated the radiological and histopathological findings.

Case Report

A 35-year-old female patient was referred to the Department of Oral and Maxillofacial Surgery for evaluation of an intraoral swelling. The lesion had been slowly increasing in size since it was first noticed about 1 year previously, causing difficulty in speech and dysphagia. There was no history of associated trauma, pain, paresthesia or lymphadenopathy. The mass was soft, and the overlying mucosa was normal in color and texture, nontender to palpation, and mobile arising from soft palate thus causing obliteration of oropharynx [Figure 1]. Computed tomography (CT) scan and magnetic resonance imaging were requested and they both showed a well-defined soft tissue mass arising from soft palate (measuring around 4.5 cm × 5 cm × 4 cm) causing significant narrowing of the oropharyngeal airway with no obvious involvement of hard palate. The CT showed no calcification, noncystic component, and no fat within the mass [Figures 2-5]. Based on clinical and radiological investigations, a provisional diagnosis of benign tumor arising from minor salivary glands of the soft palate was made. Incisional biopsy and fine needle aspiration cytology were done under general anesthesia for final diagnosis based on histopathological findings. Histopathological examination revealed islands of plasmacytoid myoepithelial cells with round eccentric nuclei distributed over myxoid connective tissue matrix suggestive

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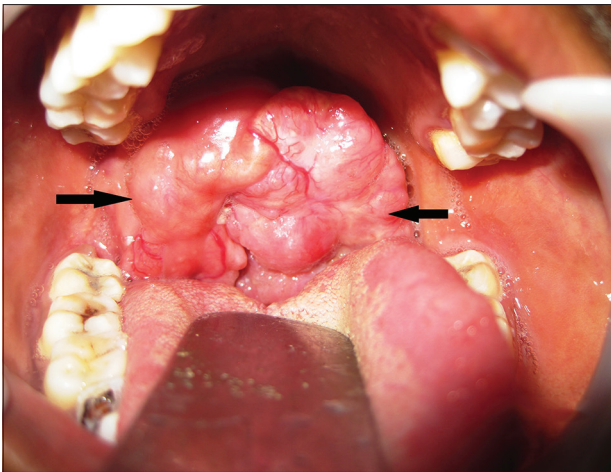


Figure 1: Intraoral clinical photograph showing the swelling in the soft palate region thus obliterating the oropharynx

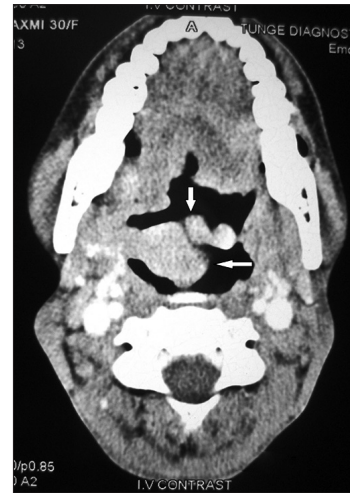


Figure 2: Computed tomography axial view showing soft tissue mass (marked by arrow) arising from soft palate reaching up to the posterior pharyngeal wall causing obliteration of nasopharyngeal and oropharyngeal airway

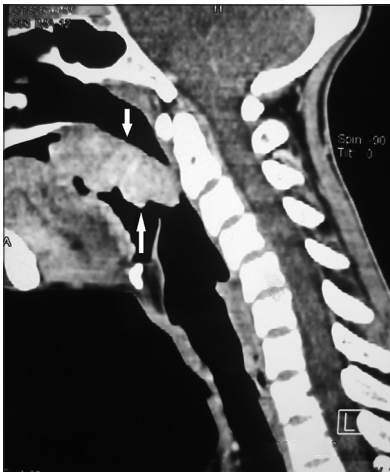


Figure 3: Computed tomography sagittal view showing soft tissue mass (marked by arrow) arising from soft palate reaching up to the posterior pharyngeal wall causing obliteration of nasopharyngeal and oropharyngeal airway

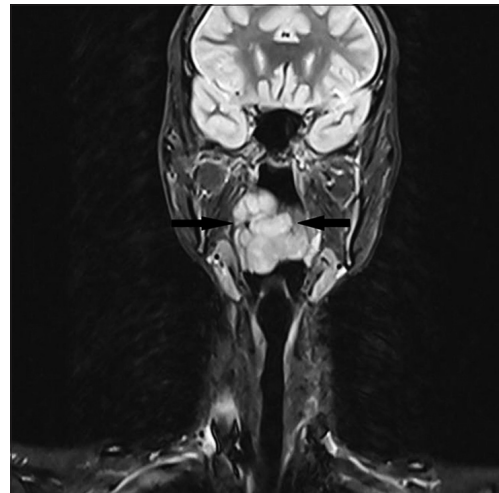


Figure 4: Magnetic resonance imaging coronal section showing soft tissue mass (marked by arrow) arising from soft palate reaching up to the posterior pharyngeal wall causing obliteration of nasopharyngeal and oropharyngeal airway

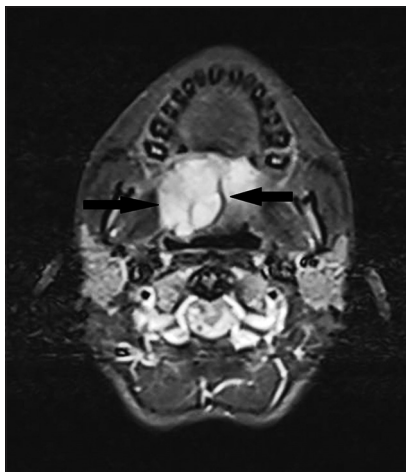


Figure 5: Magnetic resonance imaging axial section showing soft tissue mass (marked by arrow) arising from soft palate reaching up to the posterior pharyngeal wall causing obliteration of nasopharyngeal and oropharyngeal airway

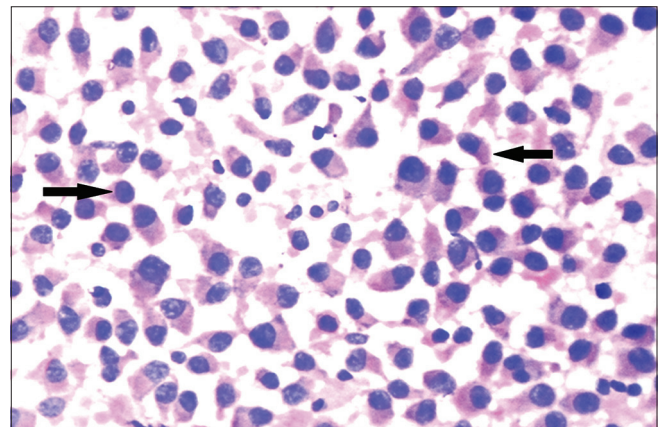


Figure 6: Fine needle aspiration cytology smear showing epithelial cells (marked by arrow) with eccentric nuclei (H and E, $\times 400$)

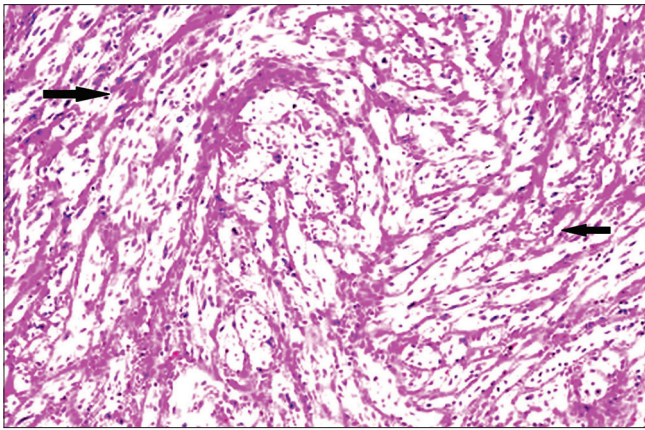


Figure 7: Histopathological section showing islands of plasmacytoid myoepithelial cells (marked by arrow) with round eccentric nuclei distributed over myxoid connective tissue matrix (H and E, $\times 100$)

of myoepithelioma [Figures 6 and 7]. Based on the clinical, radiological, and histopathological findings, a diagnosis of myoepithelioma of the soft palate was made. The treatment plan was made for surgical excision with wide margins to avoid recurrence and subsequent long-term follow-up.

Discussion

Myoepithelioma is a benign neoplasm of salivary glands that represents 1.5% of all salivary glands neoplasms derived from myoepithelial cells. Their most frequent location is parotid glands. No gender predilection has yet been reported in the literature, and the highest incidence is observed in the third decade of life.^[5]

Myoepithelioma is a well circumscribed or encapsulated tumor. Histopathologically, these tumors are classified into five variants: Spindle cell type, plasmacytoid cell type, reticular type, clear cell type, and a combination of these types depending on the shape and cellular content. Spindle cell subtype is the most common variant and has a predilection for occurrence in parotid glands of older patients.^[5,6] The plasmacytoid type has a predilection for the palate of younger individuals and may be arranged in sheets, however, it is often less cellular and the cells may be arranged in groups separated by an abundant, loose myxoid matrix that is predominantly composed of hyaluronic acid. The plasmacytoid cells are rounded, oval or polyhedral with eosinophilic cytoplasm, and eccentric nuclei.^[6,7] The most important diagnostic criteria are to differentiate myoepithelioma from its malignant counterpart. Malignant myoepithelioma is more aggressive and show recurrence even after adequate treatment.^[7,8] Clinically, malignant myoepithelioma is rapidly growing, ulcerated mass often invading into adjacent tissues causing bony erosion with metastasis into other parts of the body. Histopathologically presence of cellular atypia, cellular pleomorphism, cellular necrosis, increased mitotic figures, invasive growth pattern or a combination of all these favors the diagnosis

of malignant myoepithelioma.^[8,9] Differential diagnosis of myoepitheliomas of soft tissue include pleomorphic adenoma, neurinomas, hemangiomas, malignant tumors, metastatic tumors, lymphoma, solitary fibrous tumor, nerve sheath tumors, fibrous histiocytoma, paraganglioma, leiomyoma, leiomyosarcoma, hemangiopericytoma, and other inflammatory diseases. Most of these lesions share common clinical and radiological features, so biopsy plays a significant role in confirmation of the diagnosis of myoepithelioma, as it is difficult to differentiate myoepithelioma from other salivary gland tumors like pleomorphic adenoma. CT scan of myoepitheliomas shows varying CT enhancement patterns, which include: Faint enhancement, no significant enhancement or marked enhancement. Factors, which affect enhancement pattern of myoepithelioma include histopathological component, stroma, vascularity, and histological cell type.^[7-9] The cellular myoepithelioma with fibrous stroma being more vascular shows more significant enhancement than those myoepithelioma being rich in connective tissue myxoid stromal component. The treatment of choice for myoepitheliomas is mainly surgical with complete surgical excision of the lesion including wide margins of the nonlesional area, without recurrence risk even after 10 years of surgery.^[9,10] Other factors affecting treatment plan are the size of the lesion, age, and importantly anatomic location. Radiation therapy is the treatment of choice only in cases where a surgical operation is not feasible due to age and anatomic location of the tumor.^[10]

Conclusion

Myoepithelioma should be differentiated from the other soft tissue tumors, especially those arising from minor salivary glands, such as pleomorphic adenoma and adenoid cystic carcinoma. Nonetheless, the main success factor associated with the treatment is the early diagnosis and the first efficient treatment from specialized services, which include a team of radiologist, pathologist, anesthetist, otolaryngologist, and oral surgeon. Resection with some safe margin is the best primary method to avoid recurrence and subsequent long-term follow-up is a must to rule out possible malignant changes if any.

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