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Myoepithelioma of the Vocal Fold - A Case Report

ABSTRACT

Background: Myoepithelioma of the vocal fold is an extremely rare, benign tumor with non-specific symptoms. To the best of our knowledge, we report the sixth case of myoepithelioma in the larynx and the third benign larynx localization of myoepithelioma. **Summary**: We report a case of 16-year-old male presenting with hoarseness of voice. He came to our hospital with a history of previous three surgeries; the histopathological examination reported as "Vocal Cord Polyp" and "Inflammatory Myofibroblastic Tumor". Stroboscopy revealed a multilobulated vascular lesion attached to the left vocal process. CT scan showed a well-demarcated focal nodular lesion arising from left true vocal cord, projecting into laryngeal lumen. Major laser surgery was performed under general anesthesia in our hospital. The final histopathologic diagnosis was myoepithelioma (spindle cell type) of left vocal fold. **Conclusion**: The larynx is a very rare site for this type of tumor. Hence, it is crucial to make the correct diagnosis, as it can be confused with other spindle cell tumors which occur in the larynx.

Key words: Larynx, Myoepithelioma, Vocal fold

INTRODUCTION

Myoepithelioma is a rare, benign salivary gland tumor arising from the proliferation of myoepithelial cells, belonging to a distinct category of neoplasms according to the World Health Organization.

Myoepitheliomas of major salivary glands were described by Sheldon and Heyman for the first time in 1943 as a variant of pleomorphic adenoma,[1] then first in 1991 and later in 2005, the WHO recognized myoepithelioma as a distinct entity.[2,3]

Myoepitheliomas account for 2.2% and 5.7% of all benign major and minor salivary gland neoplasms, respectively. The parotid gland is affected in approximately 40% of cases.[4] The remaining are in glandula submandibularis and seromucous glands of the nose and larynx.[5,6] Other sites reported are the skin, chest, lung, and pancreas.[5-7]

Myoepithelioma of parotid occurs in older people and is composed more often of spindle and epithelial cells. Myoepithelioma of minor glands occurs in slightly younger individuals and is composed of plasmacytoid cells.[8]

According to our PubMed research for keywords "Larynx, Myoepithelioma," previously five case reports on the similar topic have been published as follows: myoepithelioma of the larynx by Chatziavramidis *et al*. published in 2009;[5] plasmacytoid myoepithelioma of the laryngeal region by Martínez-Madrigal *et al*. published in 1995;[9] primary myoepithelial carcinoma of the larynx by Yu *et al*. published in 2011;[10] calponin expression in laryngeal myoepithelial carcinoma and its prognostic implications by Mao *et al*. published in 2010;[11] and malignant myoepithelioma of the larynx with massive metastatic spread to the liver by Ibrahim *et al*. published in 1991.[12]

To the best of our knowledge, we report the sixth case of myoepithelioma in larynx and the third benign larynx localization of myoepithelioma.

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CASE REPORT

A 16-year-old male presented with hoarseness of voice for 9 months. There was no associated pain, dysphagia, or difficulty in talking. He underwent 3 surgeries at different institutions with intermittent mild improvement in a span of 3–4 months. The post-operative histopathological diagnosis was given as "Vocal Cord Polyp," "Recurrent Vocal Cord Polyp," and "Inflammatory Myofibroblastic Tumor," respectively.

The paraffin blocks submitted for review in our hospital were reported as recurrent spindle cell tumor, left vocal fold - the possibilities including myofibroblastic tumor and myoepithelial tumor. Features of sarcomatous squamous cell carcinoma were absent.

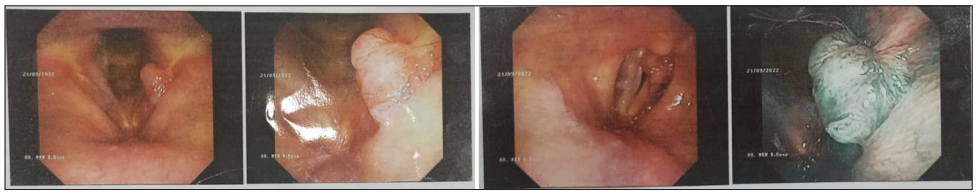

Figure 1: Stroboscopy findings

Stroboscopy revealed a left posterior multilobulated vascular lesion, attached to the left vocal process [Figure 1]. There was marked decrease in amplitude of the left mucosal wave.

CT scan of the neck showed a well-demarcated focal nodular lesion arising from the left true vocal cord and projecting into the laryngeal lumen. It measures about 5.3 × 5 × 5.3 mm (AP × TR × CC) and shows mild homogeneous post-contrast enhancement.

Major laser surgery was performed under general anesthesia at our hospital in view of left vocal fold posterior growth in September [Figure 2]. The procedure was uneventful and given tablet azithromycin for 1-month post-operative. In our case, the patient has not yet reported for follow-up.

Histopathological examination

Whitish fragment measuring 1 × 0.7 × 0.5 cm was received. Histology revealed a tumor arranged in lobules and composed of short spindle cells loosely arranged in fascicles within a myxoid stroma. The cells possess scanty cytoplasm and contain uniform bland appearing oval or elongated nuclei. Mitoses are rare. The overlying squamous epithelium is flattened but otherwise unremarkable [Figure 3]. The diagnosis was given as recurrent spindle cell tumor of left vocal fold.

Immunohistochemical studies showed positivity for Pan CK, EMA, SMA in the spindle cells. Calponin, S100 protein, CD34, CK5/6, P40 were negative; Ki67 was about 10% in areas of high proliferation [Figure 4].

Final diagnosis

Myoepithelioma, left vocal fold.

DISCUSSION

Myoepithelioma of the larynx is a very rare tumor, originating from the myoepithelial cells of mucosal glands of larynx. Myoepithelial cells are contractile cells originating from the ectoderm and are found in normal tissues with secretory function that aids in excreting glandular secretions.[13] Myoepithelial cells contain myofilaments in their cytoplasm and show contractility. These cells support the parenchyma and contribute to the production of laminin, collagen type-IV, and fibronectin to maintain the "basal lamina."[9] The hypothesized cell of origin is a common stem cell with a bidirectional differentiation into epithelial or myoepithelial cell; the

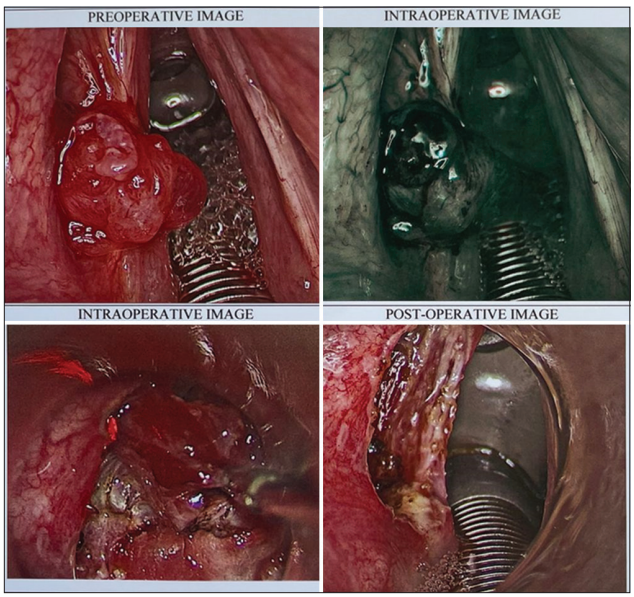

Figure 2: Pre-operative, intraoperative, and post-operative images of major laser surgery

varied histological types (spindle, plasmacytoid, epithelioid, clear, and oncocytic) exhibited by myoepitheliomas can be attributed to the various stages in the differentiation from a cell that has the potential to differentiate into epithelial cells.[14] The most frequent types of myoepithelioma are the spindle-cell form and the plasmacytoid form.[5] The spindle cell type tumors have a stroma-like appearance, often confused with mesenchymal tumors (lesions of fibroblasts, Schwann cells, or smooth muscle cells).[15] Myoepitheliomas are slow-growing, painless, and circumscribed masses.

Majority of the myoepitheliomas are benign. Malignant transformation to myoepithelial carcinoma has been reported but is extremely rare.[16] Myoepithelial carcinomas have moderate-to-severe atypia with vesicular nuclei and prominent nucleoli, high mitotic count, and necrosis.[16,17]

In our case, tumor was of spindle cell type. The common differentials considered were as follows: (1) Sarcomatoid squamous cell carcinoma: However, the absence of atypia and high mitotic activity in the tumor cells and dysplasia/carcinoma - *in situ* in the overlying squamous epithelium helped in ruling out this diagnosis; (2) myofibroblastic tumor: Positive epithelial markers (pan CK and EMA) on immunohistochemistry ruled out this diagnosis; (3) other mesenchymal tumors such as Schwannoma would be positive

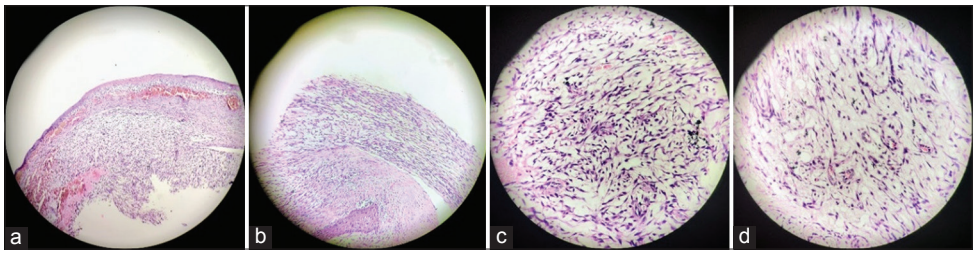

Figure 3: Histological sections of tumor showed short spindle cells loosely arranged in fascicles within a myxoid stroma. (a) and (b) H and E stain at ×100 magnification; (c) and (d) H and E stain at ×400 magnification

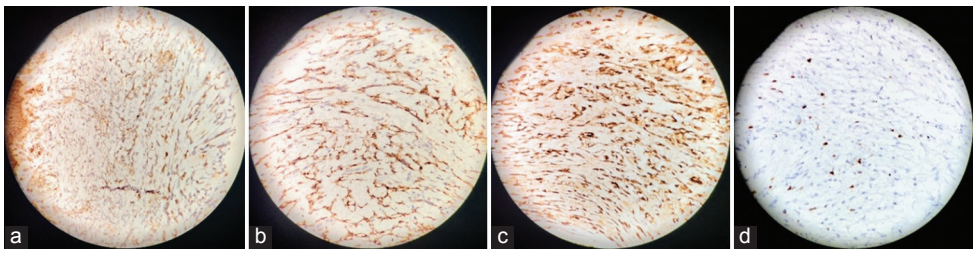

Figure 4: Immunohistochemistry showed positivity for (a) pan cytokeratin (pan CK), (b) epithelial membrane antigen (EMA), (c) smooth muscle actin (SMA), (d) Ki67 proliferation index. (a-d) in 400× magnification

for S-100 protein on immunohistochemistry. However, in our case, S-100 staining was negative; (4) pleomorphic adenoma (mixed tumor): No ductal elements or cartilage was identified in this tumor; (5) amelanotic melanoma: It was ruled out on the basis of morphology as well as immunohistochemistry for S-100 protein, which was negative.

The treatment of choice for benign lesions is complete surgical excision with free margins. For malignant tumors, tumor-free margins, neck dissection, and radiotherapy are recommended.[7] Regular follow-up is necessary to evaluate for local recurrence.

CONCLUSION

The larynx is a very rare site for myoepithelioma, this being the sixth published case of myoepithelioma of the larynx. Although rare, it has to be considered in the differential diagnosis of other spindle cell tumors in the larynx.

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