CASE REPORT

LIFE-THREATENING ANGIOMYXOMA OF THE LARYNX

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ABSTRACT

BACKGROUND: Angiomyxoma is a benign proliferative mesenchymal tumor and a very rare mass in the larynx. There is not enough information about the etiology, clinical finding, treatment and prognosis of laryngeal angiomyxoma.
CASE DETAILS: A 52 years old man presented with respiratory distress. Also, he had suffered from dysphagia, dysphonia, cough, and obstructive sleep apnea in the supine position for 6 months. He was operated on via transoral approach under general anesthesia with orotracheal intubation. The mass was encapsulated and completely removed. The histopathologic diagnosis was reported as angiomyxoma.
CONCLUSION: Angiomyxoma should be considered in the differential diagnosis of the larynx masses. The treatment of angiomyxomas of the larynx is surgical. The mass can be usually excised intraorally or endoscopically.
KEYWORDS: Angiomyxoma, larynx, dysphonia, respiratory distress

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INTRODUCTION

Angiomyxoma is a benign proliferative mesenchymal tumor. The mass appears very extremely rare in the larynx (1-3). Therefore, there is not enough information about the etiology, clinical finding, treatment and prognosis of laryngeal angiomyxoma. To the best of our knowledge, only three cases regarding angiomyxoma of the larynx have been reported in the literature (1-3). Herein, we described a case with angiomyxoma located at the right aryepiglottic fold of the larynx.

CASE REPORT

A 52 years old man presented with respiratory distress in our outpatient clinic in 20 April 2012. Also, he had suffered from dysphagia, dysphonia, cough, and obstructive sleep apnea in the supine position for 6 months. He had no history of a systemic disease, and was not and alcohol user. He had been smoking a packet of cigarette for 35 years. There was no history of trauma. Endoscopic examination revealed a large smooth surface polypoid, well-circumscribed mass based in the right supraglottic area of the larynx that obstructed most of the view of the rima glottidis and moving during respiration and phonation (Figure 1). The remainder of the clinical examination was unremarkable. Computed tomography scan showed a well-defined, enhancing solid mass, measuring 45×30 mm and narrowing the airway space and originating from the supraglottic area of the larynx (Figure 2).

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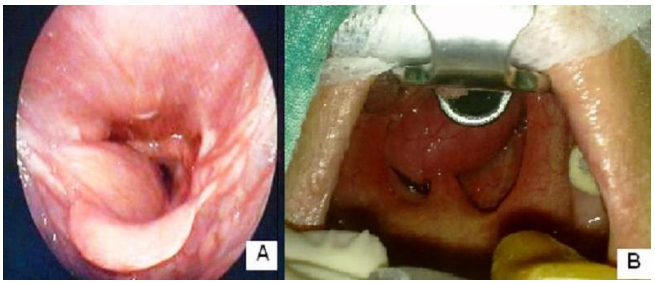

Figure 1: *(A) Endoscopic appearance of the angiomyxoma in the right supraglottic area of the larynx. (B) Intraoperative the view of the angiomyxoma in the supine position intraorally*

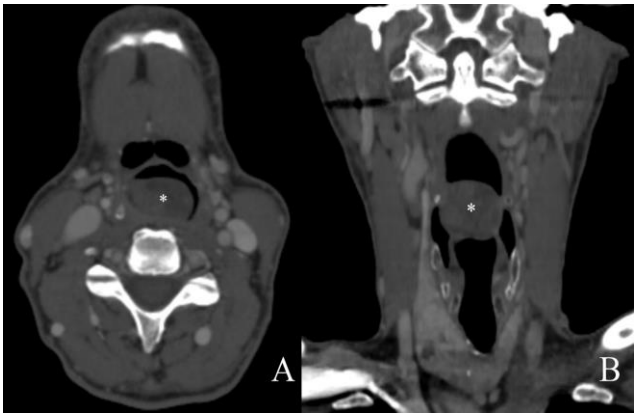

Figure 2: *(A) Axial and (B) coronal contrast-enhanced CT scan demonstrated solid mass narrowing the airway space and originating from the supraglottic area of the larynx*

Pre-operative informed consent was obtained from the patient. He was operated on via transoral approach under general anesthesia with orotracheal intubation. The polypoid mass was at the right aryepiglottic fold with a broad base. It was encapsulated and completely removed in 25 minutes (Figure 3). After surgery, the true vocal cords were normal. On macroscopic examination, the specimen measured 4.5x3.0x2.5 cm and was well circumscribed and encapsulated with a glistening surface. Histopathologic examination of the mass revealed that the tumor was characterized by a spindle-shaped proliferation or satellite cells widely separated by loose, and also myxoid stroma including a prominent vascular component. The spindle cells showed no nuclear atypia or mitoses. Immunostaining for S-100 protein and muscle-specific antigen was negative. Vascular component was found positive with cluster of differentiation (CD) 34 (Figure 4). The histopathologic diagnosis was reported as angiomyxoma. Postoperatively, no complication was encountered and no recurrence was seen during the 29-month follow-up period.

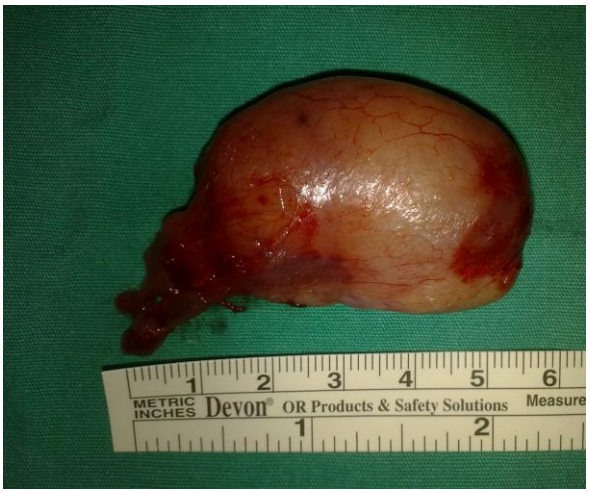

Figure 3: *Postoperatively, the view of a well-circumscribed and encapsulated tumoral specimen*

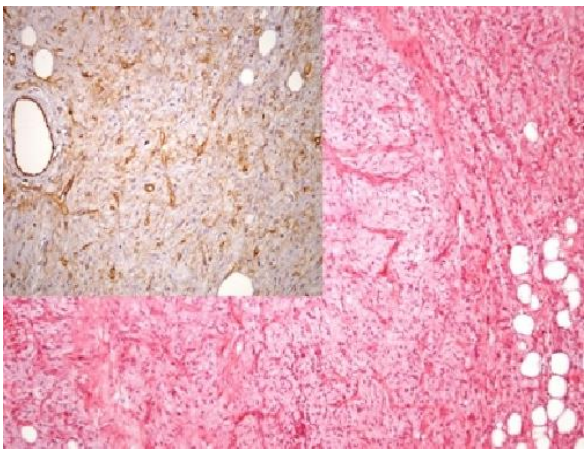

Figure 4: *Spindle-shaped or stellate cells widely separated by loose, myxoid stroma (Hematoxylin-Eosin X40)(small figure: vascular component stained with CD34 (HEX40)*

DISCUSSION

Steeper and Rosai (4) described aggressive angiomyxomas of the female pelvis and perineum arising from mesenchymal cells. Angiomyxoma of the larynx is understood to be counterpart of these tumors (1-3). The presenting symptoms of larengeal angiomyxoma are similar to many of the larengeal masses, and these include dysphagia, dysphonia, dyspnea, cough, respiratory distress

and obstructive sleep apnea (1, 2). It may be caused by airway obstruction requiring emergency tracheotomy. Endoscopic examination of the larenx usually reveals a well circumscribed, smooth and polypoidal mass (1, 2). Radiological imaging methods can be useful in determining the origin and extent of the lesion.

Histologically, angiomyxomas consist of spindle-shaped or stellate cells widely separated by loose, myxoid stroma, and a prominent vascular component. Immunohistochemically, aggressive angiomyxomas of pelvi-perineal regions have been shown positive majority of tumors for desmin, vimentin, CD44, CD34, oestrogen receptors, and smooth muscle actin. They are negative for S-100 protein and muscle-specific antigen (2, 5, 6). In our case, the only similarity with the pelvi-perineal form was negative for S-100 protein, muscle-specific antigen, and positive for CD 34.

The differential diagnosis of larengeal angiomyxoma includes many benign and malignant masses in the larenx. Internal laryngocele, fibroepithelial polyps, myxomas, myxoid neurofibromas, schwannomas, rhabdomyosarcomas, myxoid chondrosarcomas, myxoid liposarcomas, myxoid variant of malignant fibrous histiocytoma, nerve sheath myxoma low-grade myxofibrosarcomas, and other soft tissue tumors, all should be considered differential diagnosis (1,4).

The treatment of angiomyxomas of the larynx is surgical. The mass can usually be excised intraorally or endoscopically, but an external approach may occasionally be required (1, 2). Izadi et al. (1) removed the mass with a CO_2 laser. Sylvester et al. (2) operated via laryngofissure approach and covering tracheostomy. Their patient was decannulated one week postoperatively and later discharged from the hospital. We excised in the mass intraorally and did not apply tracheostomy. The patient was discharged from the hospital on the second day after surgery. The use and effect of radiotherapy and chemotherapy in treatment for angiomyxoma of the larynx is unknown, but chemotherapy and radiotherapy are not effective for aggressive angiomyxomas of the pelvis (2).

In conclusion, angiomyxoma is a benign and a very extremely rare tumor of the larynx and should be considered in the differential diagnosis of the larynx masses. The treatment of angiomyxomas of the larynx is surgical and the mass can usually be excised intraorally or endoscopically. Although angiomyxoma is a benign tumor, it occasionally behaves aggressively. Since angiomyxoma has a potential for local recurrence or metastasis, we recommend periodic control examinations and long-term close follow-up postoperatively.

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