

Myxoma : Life-threatening Benign Nonepithelial Tumor of the Larynx

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Myxoma is a rare nonepithelial neoplasm of the larynx frequently misdiagnosed as a large vocal polyp due to its slow-growing nature. Myxoma is a benign but often infiltrating neoplasm of uncertain mesenchymal cell origin, characterized by irregular round, spindle or stellate cells within a matrix containing abundant mucoid material, scant vascularity and a variable meshwork of reticulum and collagen. We report one case of myxoma with life-threatening dyspnea requiring tracheotomy.

Key Words: Larynx, myxoma

Myxoma is a tumor of mesenchymal origin and consists mainly of cells phenotypically resembling spindle or stellate cells. The term myxoma was introduced by Virchow in 1871. Myxoma is mainly found in extremities such as the thigh. In the head and neck region, bone and soft tissues are affected. The most common site of myxoma in the head and neck is the mandible (Batsakis, 1979). Myxoma of the larynx is extremely rare. A review of the literature in English for the past 30 years revealed only three reports of myxoma in the larynx. We report a case of laryngeal myxoma accompanying dyspnea.

A CASE REPORT

A 62-year-old man came to the emergency room

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complaining of dyspnea for the previous three days. A year earlier, he was diagnosed as having Reinke's edema in the right true vocal cord but refused the operation to remove the diffuse polyp. He had valvular heart disease and had a 60-pack-a-year smoking habit and a *Soju* (Korean distilled alcohol) 1 bottle (360 cc)-a-day drinking habit for the previous 30 years. Indirect laryngoscopy revealed a pedunculated mass at the entire free margin of the right true vocal cord, obstructing most of the laryngeal inlet and moving to and fro during respiration. Emergency tracheotomy was performed in the emergency room. A Portex-James tube #7.5 was inserted through the second tracheal ring. Echocardiography showed aortic regurgitation, grade II. Patient history revealed no diabetes mellitus, hypertension or hepatitis. There was no lymphadenopathy.

In the operating room, an endotracheal tube was placed through tracheostoma and a direct laryngoscope was inserted. A large polypoid mass was noted at the right true vocal cord with a broad base. The mass was grasped with a right-curved cup-forcep and resected with upward scissors. The remnants of the mass were further resected. Grossly,

the specimen was measured at $2.5 \times 2.5 \times 1.5$ cm with a glistening surface (Fig. 1). Microscopically, spindle or stellate cells were noted in the myxoid substrate (Fig. 2). Pathologic diagnosis of the specimen was myxoma. The tracheostoma was sealed two

days after the operation. The patient was discharged without dyspnea.

DISCUSSION

Myxoma is a neoplasm of mesenchymal origin. Soft-tissue myxomas may occur anytime from birth to old age. According to our review of the literature in English, only three cases of laryngeal myxoma have been reported over the past 30 years (Chen and Ballecer, 1986; Senna *et al.* 1991; Hadley *et al.* 1994)(Table 1).

Clinical presentation of laryngeal myxoma includes hoarseness, dysphonia, dysphagia or airway obstruction and especially dyspnea, which is related to the location and size of the tumor. A large myxoma may require tracheotomy due to dyspnea. The chief complaints of most patients, however, are limited to hoarseness and dysphonia.

Although laryngoscopy and imaging studies may aid in the diagnosis, actual diagnosis is made through tissue biopsy. Laryngeal examinations may reveal a

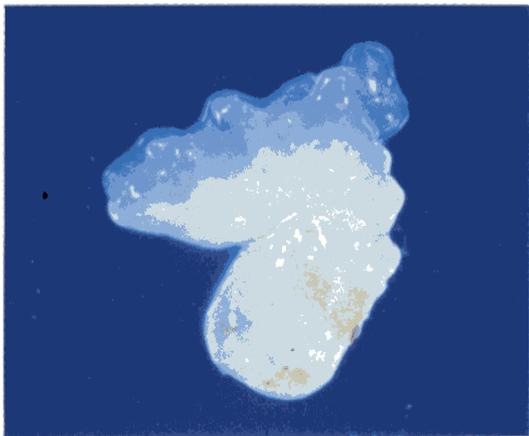


Fig. 1. A white glistening gelatinous mass measuring $2.5 \times 2.5 \times 1.5$ cm.

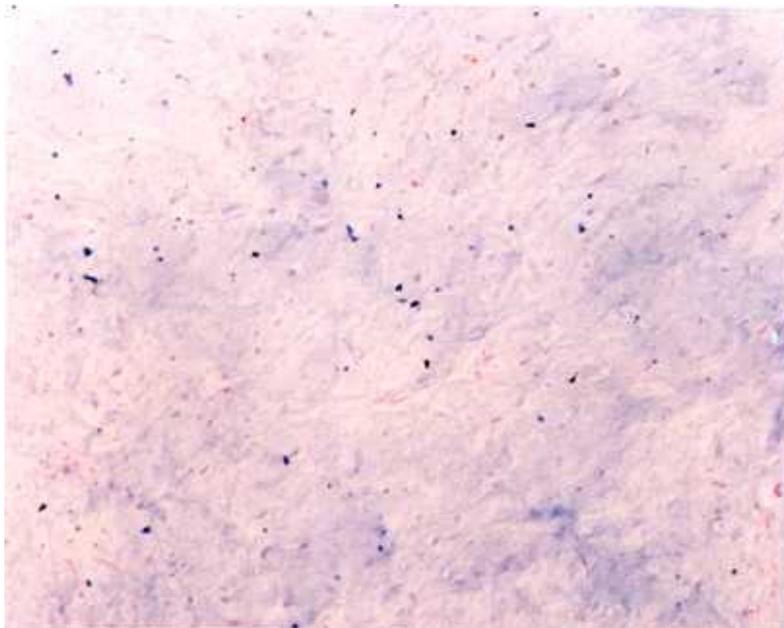


Fig. 2. Scanty vascularity is shown with spindle-shaped cells. Scattered spindle cells are noted in a myxoid stroma (Alcian blue; $\times 100$).

Table 1. Reported cases of laryngeal myxoma in the literature

No.	Author	Age	Sex	Clinical features	Treatment	Follow-up	Location
1	Chen et al(1986)	37	M	Dysphonia, dysphagia	Laryngoscopic excision	NED 1 y	Lingual surface, epiglottis
2	Sena et al(1991)	70	M	Hoarseness	Explo-neck	NED 12 m	Left supraglottis
3	Hadley et al(1994)	64	M	Dysphonia	Laryngoscopic excision	NED 18 m	Left vocal fold
4	Authors (1997)	62	M	Dyspnea	Laryngoscopic excision	NED 2 m	Right vocal fold

NED : no evidence of disease

simple large polyp and CT may suggest a benign mass of larynx. The most prevalent site of laryngeal myxoma, according to the literature review, is the vocal fold (Table 1).

Histopathologic diagnosis of laryngeal myxoma is made from its unusual histological features: spindle or stellate cells within a matrix containing a scanty vascularity; a variable meshwork of reticulum and collagen; and abundant mucoid material (Tucker, 1993).

Differential diagnosis between laryngeal myxoma and myxoid change of vocal polyp is important. The key for the pathologic differentiation is the scanty vascularity, no fibrin and no hemorrhage in the laryngeal myxoma, as opposed to a plentiful vascularity and hemorrhage with hemosiderin-laden macrophages in the myxoid change of vocal polyp (Hadley *et al.* 1994).

Myxoma is differentiated from the mesenchymal neoplasms --leiomyosarcoma, malignant fibrous histiocytoma, myxoid liposarcoma, neurofibroma and metastatic mucinous carcinoma-- through the immunohistochemical staining method. The spindle cells of myxoma are reactive to vimentin but lack desmin, cytokeratin, actin, α_1 -antitrypsin, α_1 -antichymotrypsin, and S-100 protein (Gnepp, 1988).

Complete excision is the treatment of choice, although the treatment modality may vary according to the size of the mass. The tumor may be slow-

growing but it is infiltrative and destructive. Since the tumor is poorly circumscribed and tends to infiltrate, the tumor must be removed with surrounding normal tissues in order to prevent recurrence. Radiation therapy is generally ineffective. Prognosis is good in cases of complete excision. Regular follow-up is required to rule out recurrence.

Laryngeal myxoma is a rare tumor with hoarseness as a main symptom. Complete surgical excision, including the surrounding normal tissue, is mandatory to prevent recurrence.

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