FIBROMA OF THE SUPRAGLOTTIC LARYNX

A Case Report

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INTRODUCTION

Solitary fibrous tumors are relatively rare mesenchymal neoplasms that were originally described as pleural- or peritoneal- based lesions. Although they were considered a form of mesothelioma, subsequent investigation failed to reveal mesothelial differentiation. Characterization of their histologic and immunohistochemical features, as well as identification in a multitude of nonmesothelial-based locations has further served to distinguish these lesions from the more diffuse and aggressive mesothelioma. Reports of solitary fibrous tumour in the larynx are extremely rare[1]

Mesenchymal tumors of the head and neck, primarily those originating in the larynx, are rare and include several histological types. One of these neoplasms is the solitary fibrous tumor (SFT). SFT is a neoplasm considered part of the solitary fibrous tumor - hemangiopericytoma spectrum. This tumor was first described in the pleura. Approximately 50% of SFT cases are described in the thorax, while only 6% of the cases develop in the head and neck[2]. At that location, the oral cavity is the most frequent subsite affected, although other cases have been reported in orbits, nose, paranasal sinuses, nasopharynx, parapharynx, larynx, major salivary glands, and thyroid[3]. No more than 11 cases have been reported in the larynx until this moment. We present two new cases that were surgically treated and a literature review of the larynx SFT reports.

We report a case of solitary fibrous tumor of the larynx in a 30 year old man.

CASE REPORT

A 30 year old male presented with change in voice since 3 months, gradually progressive. It was followed by gradually progressive difficulty in breathing since 15 days. Patient presented with stridor in the ENT OPD. Family and medical histories were non-contributory. Patient was a chronic alcoholic since 10 years.

Physical examination revealed respiratory distress with stridor. There were no palpable cervical masses, and the rest of the head and neck examination was unremarkable. On indirect laryngoscopic examination, a grayish smooth spherical mass was seen in the supraglottic region. Rest of the structures like arytenoids, aryepiglottic folds and vocal cords could not be visualized because of the mass. The glottic chink could not be visualized. As patient was in respiratory distress and stridor, an *emergency tracheostomy* was done immediately.

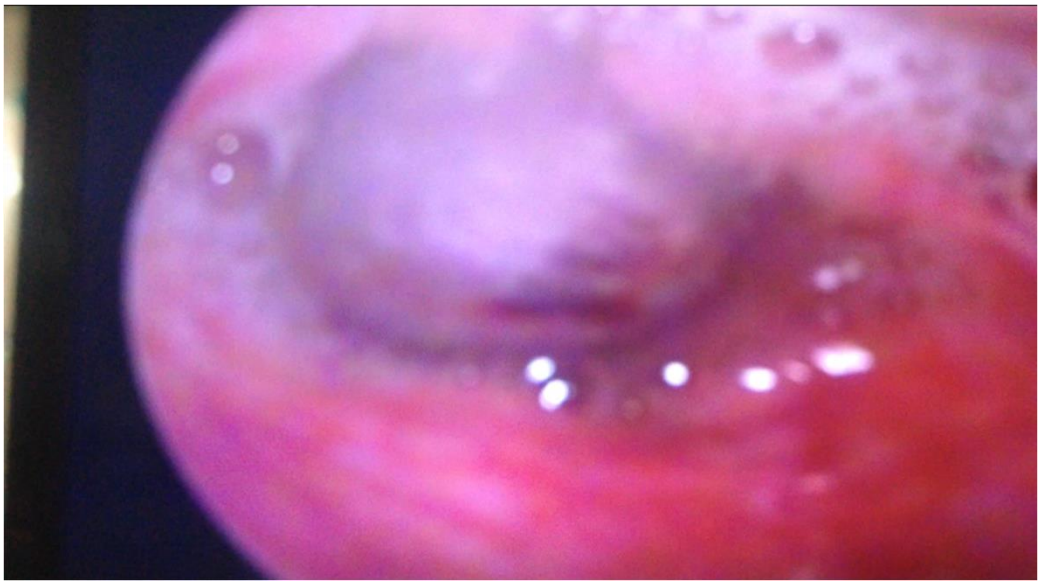

FIGURE 1 – PREOPERATIVE 70 DEGREE ENDOSCOPIC PICTURE SHOWING THE LARYNGEAL MASS.

Routine investigations were done which revealed that the patient was *HCV positive*. Rest of the haemogram was within normal limits.

A contrast enhanced CT scan (coronal cuts) revealed possibility of benign laryngeal lesion – Papilloma/Polyp. A 35 X 21 X 16 mm well defined lesion showing intense heterogenous enhancement is noted involving the supraglottis and laryngeal vestibule. True and false vocal cords were not separately seen. The lesion was causing effacement of paralaryngeal spaces bilaterally. And the lesion was seen to cause complete obstruction of the airway. Tracheostomy tube was seen in situ.

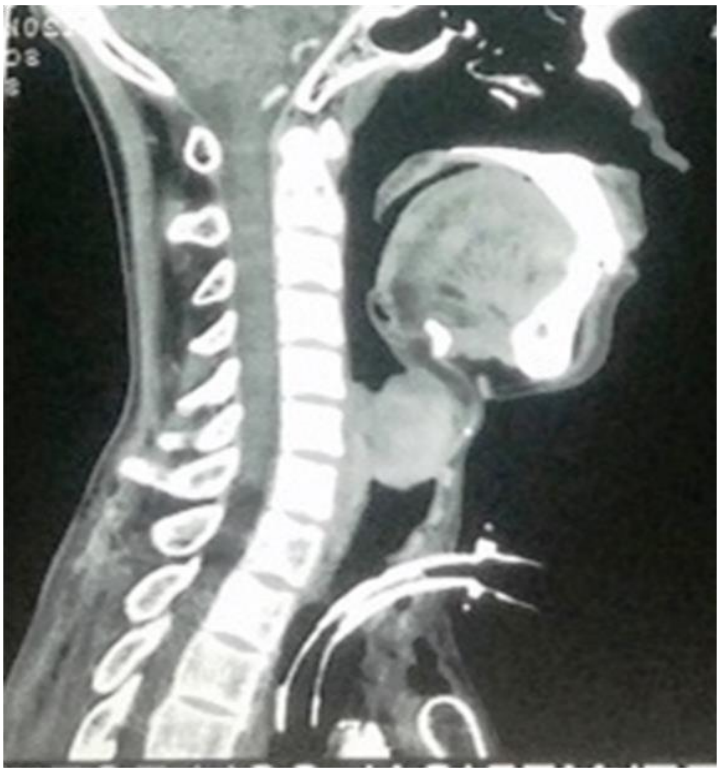

FIGURE 2 – CONTRAST ENHANCED CT SCAN RADIOGRAPH (CORONAL CUT) SHOWING HETEROGENOUS CONTRAST ENHANCEMENT IN THE SUPRAGLOTTIC REGION.

The patient was planned for surgical removal of the suspicious supraglottic laryngeal mass. On direct examination, the size of the mass appeared to be 2.5 X 2.5 cm in size, the margins and edges of the mass were not properly visualized. It was firm to hard in consistency, resting over the vocal cords. The vocal cords and glottis chink were not visualized. Using micro laryngeal instruments, the excision of this mass was done in fragments, some part of it was seen in the subglottis and it was removed as well. The fragments were sent for histopathological examination.

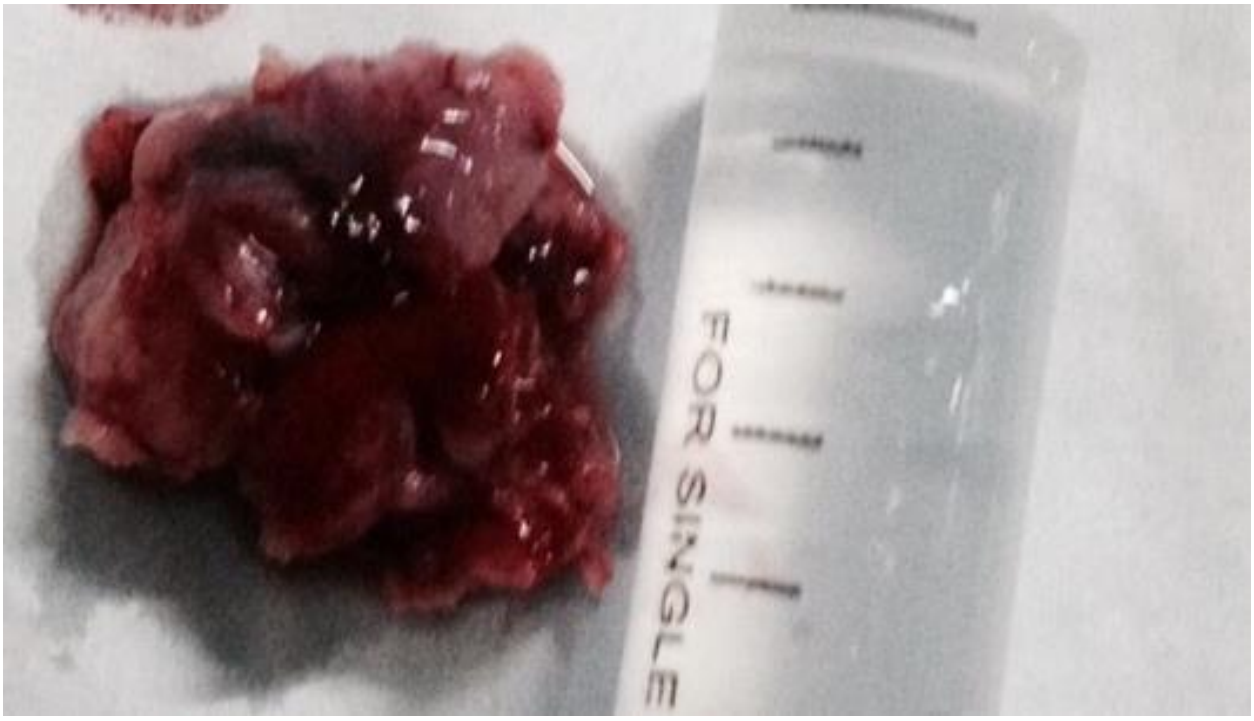

FIGURE 3 – POSTOPERATIVE EXCISED SPECIMEN

Pathological Findings

Gross examination revealed multiple greyish brown, soft to firm tissue pieces with specks of haemorrhage. Three tissue pieces were larger ranging in size from 1.5 X 1.0 X 0.3 cm to 1 X 0.8 X 0.2 cm and rest smaller tissue pieces altogether measuring 2 X 1.5 X 0.3 cm. Outer surface of 2 tissue pieces show whitish areas. Cut surface of the largest tissue piece was greyish white in colour with a few specks of haemorrhage.

Cassette 1-3: both bisected halves of larger tissue pieces.

Cassette 4: rest of the smaller tissue pieces.

Microscopic examination revealed a tumourtissur focally covered with flattened epithelium. Tumour was composed of interlacing bundles, fascicles and sheets of spindle cells. These cells have bipolar eosinophilic cytoplasm and oval to spindle vesicular nuclei. Stroma showed variable amount of collagenized tissue with scattered thin walled blood vessels. There was no evidence of mitotic activity or nuclear atypia in tumour cells.

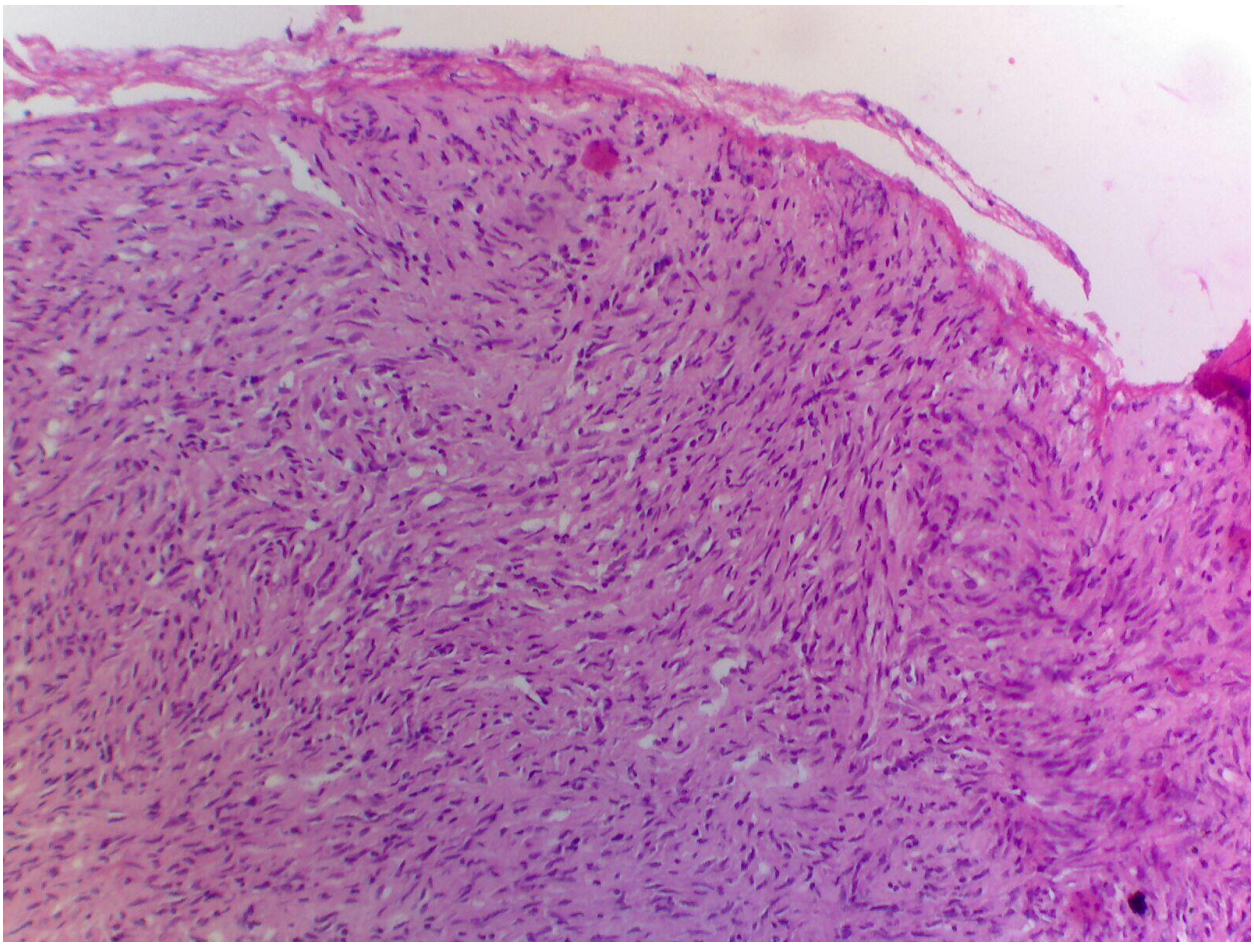

FIGURE 4 – MICROSCOPIC PICTURE OF THE SPECIMEN SHOWING THE TUMOUR COMPOSED OF INTERLACING BUNDLES, FASCICLES AND SHEETS OF SPINDLE CELLS.

Biopsy from growth supraglottic larynx showed features of a benign mesenchymaltumour morphologically consistent with fibroma. Immunohistochemistry was recommended (but the patient refused this investigation).

Two weeks following surgery, 70 degree endoscopy laryngoscopic examination was done and the vocal cords were seen to be mobile. The patient was decannulated after two weeks, following which the patient had a good speech with no respiratory distress or stidor.

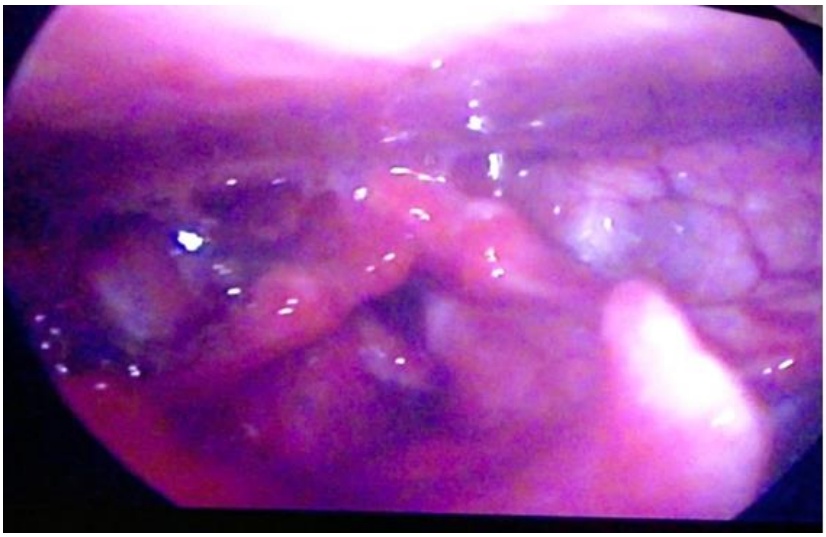

FIGURE 4 – POSTOPERATIVE 70 DEGREE ENDOSCOPIC PICTURE SHOWING THE NORMAL VOCAL CORDS.

DISCUSSION

Wagner[4] detailed the microscopic features and suggested a pleural lymphatic endothelial origin of primary pleural tumours in 1870. In 1931, Klemperer and Rabin[5]were the first to describe what we now consider the SFT as a lesion, which arose from subpleural alveolar tissues. These lesions grew slowly and frequently attained a massive size before causing clinical symptoms. They further distinguished these lesions from the more diffuse “mesotheliomas”, which were derived from the mesothelium proper. This neoplasm has been described in a host of sites and is typically benign with morphological indicators of a potentially more aggressive course, including cytological atypia, tumour necrosis, and mitotic activity[4-7]. Recurrence has also been noted when incomplete excision has been performed.

SFT must be included in the differential diagnosis of lesions with fibrous component, including fibromatosis, fibrous histocytoma, fibrosarcoma, synovial sarcoma, and metastatic scirrhous carcinoma. All these entities exhibit a dense fibrous collagen matrix that can reveal low T2 signal intensity in Magnetic Resonance Imaging. At CT, the tumors were highly vascularized, polypoid-to spherical-shaped, arising from the submucosa. Usually they show isoattenuation relative to the adjacent muscles. Low-attenuation areas inside the tumor may be related to cystic or myxoid degeneration or stroma areas with low contrast uptake[8]. Its clinical behavior is variable, although most of them are slow-growing, with benign course. It is difficult to differentiate them from other slow-growing mesenchymal diseases, since the diagnosis depends on architectural, cytomorphological, and immunohistochemical aspects[3]. The histopathological characteristics does not differ among the anatomical sites; they are characterized by the lack of an architectural pattern, varied cellularity, with areas of stromal hyalinization, and often associated to a hemangiopericytoma-like vascular pattern. These tumors show immunoreactivity for CD34, CD99, bcl-2, and vimentin; and are negative for cytokeratin, S-100, and smooth muscle actin[9]. When tumors develop in larynx, it usually affect middle-aged adults, like other extrapleural disease cases. Perhaps due to early presentation, most of lesions are < 5 cm in diameter[10] .It generally presents with progressive dysphonia, foreign body sensation, voice quality change, dyspnea or cough, with slow evolution of symptoms, depending on which part of the larynx is involved.

TABLE Summary of literature review of larynx solitary fibrous tumor cases*

Variable	**n = 12 (100%)**
Gender	
Male	8 (66.6)
Female	4 (33.3)
Symptoms	
Dyspnea	6 (50.0)
Dysphagia	2 (16.6)
Foreign body sensation	7 (58.3)
AW obstruction	2 (16.6)
Dysphonia	7 (58.3)
Location	
Supraglottis	11 (91.6)
Subglottis	1 (8.3)
Treatment	
Partial laryngectomy	4 (33.3)
CO2 laser resection	3 (25.0)
Total laryngectomy	1 (8.3)
Enucleation by pharyngotomy	4 (33.3)
Min-max	
Age	13-77
Diameter	1.0-5.1
Average (SD)	
Age	52.08 (19.96)

Variable	n = 12 (100%)
Diameter	2.99 (1.19)

*1 case not included (data not available) and included cases described in this paper

n (%): relative and absolute frequency, respectively; AW: airway; min-max: minimum and maximum values; SD: standard deviation; age in years, diameter in centimeters.

Solitary fibrous tumor of the larynx presents in men at a mean age of 42 years (29–60 years). Patients are usually not smokers and clinically develop progressive hoarseness, foreign body sensation, cough, or even acute upper airway distress. Cervical adenopathies have not been present in the cases reported. Clinical suspicion for a primary squamous cell carcinoma of the larynx has been the rule, and imaging studies follow to investigate this suspicion. Fiberoptic laryngoscopy of the reported cases revealed a smooth, round, well-circumscribed submucosal mass arising from the false vocal cord. Two occurred in the right side and the other on the left. Head and neck computed tomographic scan confirmed the endoscopic impression and is essential to rule out adjacent bony or cartilaginous involvement. Treatment of choice is transoral carbon dioxide laser surgery[11,12] .After undergoing this procedure, 1 patient developed laryngeal dyspnea after having a few months of clinical remission and subsequently underwent a modified left ventricular laryngectomy[11] .Cervical lymph node dissection is not indicated. The disadvantage of the carbon dioxide laser surgical resection is that the lesion is cauterized and, therefore, evaluation of surgical margins is difficult. This is significant in that recurrence has been reported when complete excision was not accomplished. In our institute, we used microlaryngeal instruments for excising the mass, following which the patient responded well.Most SFTs are benign; however, Benlyazidet al[11] reported the concomitant occurrence of SFT and adenocarcinoma, the latter arising in the lungs and the former of unknown origin. Briselli et al[13] reported that 12% of SFTs of the pleura are responsible for the patient's death because of their extensive intrathoracic growth, and Witkin and Rosai[14] also described a more aggressive behavior of those arising in the mediastinum.Cartilaginous tumors, such as chondrosarcomas and chondromas are the most common mesenchymal neoplasms of the larynx. Owing to the morphologic findings in this case, the differential diagnosis included fibroblastic or myofibroblastic neoplasms, such as leiomyoma/leiomyosarcoma, rhabdomyoma/ rhabdomyosarcoma, myofibromatosis, inflammatory myofibroblastic tumor (inflammatory pseudotumor), synovial sarcoma, and SFT.Sarcomatoid carcinoma, which is an epithelial tumor with ''mesenchymal'' differentiation, should also be considered in the differential diagnosis; however, no epithelial component was identified in this case. Finally, metastatic or primary malignant melanoma was also considered due to its capacity to mimic a multitude of other lesions[15].

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