



## A Rare Case of Laryngeal Schwannoma

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### Abstract

**Introduction:** Schwannoma within larynx is uncommon and arises from perineural schwann cells. These tumors are a potential threat to airway and usually cause a challenge to otorhinolaryngologists in diagnosis and management. A definite diagnosis can only be made histologically. Very few benign lesions due to location, size and site of origin need total or partial laryngectomy. Laryngeal Schwannoma is one of them. We present a case report of this rare laryngeal entity wherein diagnosis was tortuous and tormenting and various investigations gave conflicting information adding to the diagnostic dilemma.

**Methods:** A case report of laryngeal schwannoma in a young patient in a tertiary care rural hospital in September 2015 is discussed

**Result/Discussion:** The clinical course, quest of diagnosis, conflicting inputs given by various investigations, management is discussed.

**Conclusions:** Our case is unique as laryngeal schwannoma is a rare entity with diagnostic crisis. Decision making for management was also challenging due to benign nature and extent of lesion. In light of final histopathology report agreeing with USG guided report, we feel later should be considered as a reliable implement for diagnosis of this rare tumour

**Keywords:** Laryngeal schwannoma; Stridor; Neurilemoma

### Introduction

Schwannoma within larynx is uncommon and consists of only 0.1% to 1.5% of all benign laryngeal tumours [1,2]. As few as 250 cases are reported in literature till date [3]. Schwannoma arises from perineural schwann cells and are well encapsulated, slow growing, submucosal abutting the parent nerve but extrinsic to nerve fascicles [1,4]. Around 80% are located in the region of aryepiglottic folds and 20% are located in region of false and true vocal cords [4] and known to arise usually from internal branch of superior laryngeal nerve [2,4].

Laryngeal schwannomas usually occur in females in 4<sup>th</sup> and 5<sup>th</sup> decades. These tumors are a potential threat to airway and usually cause a challenge to otorhinolaryngologists in diagnosis and management [5]. A definite diagnosis can only be made histologically [6]. The most successful curative option is complete surgical resection [7]. Scores of articles with detailed literature review of Laryngeal Schwannoma have been published. Practising Otorhinolaryngologists should always be alive to the possibility of this uncommon laryngeal lesion. Very few benign lesions due to location, size and site of origin need total or partial laryngectomy. Laryngeal Schwannoma is one of them. We present a case report of this rare laryngeal entity.

### Case Presentation

A 27 years old female came to ENT OPD with history of change in voice since a year. Onset of hoarseness of voice was sudden without any precipitating factors like upper respiratory tract infection, trauma, surgical procedure and it worsened gradually for the past one year. Symptom was refractory to all the medications that she took from general practitioners. Around same time she also developed mucoid expectoration and breathlessness on exertion. Peculiar symptom of dull pain on the left side of neck was characteristic. There was no fever, dysphagia, odynophagia, neck swelling or stridor. Patient had no addiction and no known systemic illness.

On Indirect laryngoscopy, there was submucosal swelling involving left aryepiglottic fold, arytenoids, left vocal cords and extending to left pyriform fossa. Patient was admitted, investigated and a direct laryngoscopy under general anaesthesia with biopsy was planned. Direct laryngoscopy using Chevalier Jackson type of direct laryngoscope was performed. During direct laryngoscopy, a smooth, firm, submucosal mass arising from the left hemilarynx was seen and biopsy was

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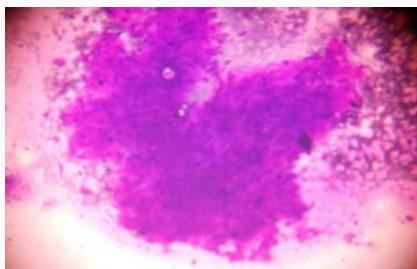
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**Figure 1:** Chest radiograph postero-anterior view showing pneumothorax.



**Figure 2:** Radiogram after insertion of Intercostal Drain.

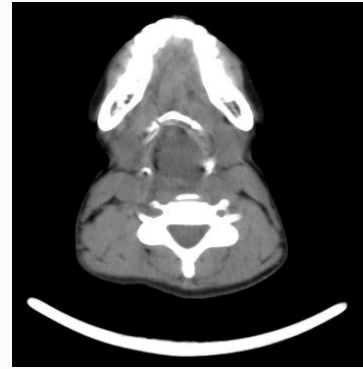


**Figure 3:** Ultrasound guided Fine Needle Aspiration Cytology suggestive possibility of schwannoma or leiomyoma.

performed. At the end of the procedure patient suddenly developed stridor for which emergency tracheostomy was done and a cuffed 7.5 portex tracheostomy tube inserted. Air blast was good but following tracheostomy patient had developed emphysema over face, neck, chest. An emergency chest radiograph posterior anterior view revealed bilateral pneumothorax (Figure 1) which was taken care by putting Intercostal Drain no 28 in 4<sup>th</sup> intercostal space in midclavicular line for 3 days (Figure 2). Patient responded positively and was stable.

The biopsy report was inconclusive showing only squamous cell hyperplasia. Patient was further investigated. Ultrasonography was suggestive of possible Schwannoma or leiomyoma whereas Computed Tomogram scan uncovered heterogeneously enhancing soft tissue density mass lesion in hypopharynx on left side involving left side of cricoid extending upto the right half of cricoid cartilage with near obliteration of tracheal lumen (Figure 4 and 5) most likely to be neoplastic.

Patient was re-biopsied but report was far from conclusive and showed chronic lymphoplasmacytic inflammatory infiltrate with



**Figure 4:** Computed tomogram showing the extent of tumour.



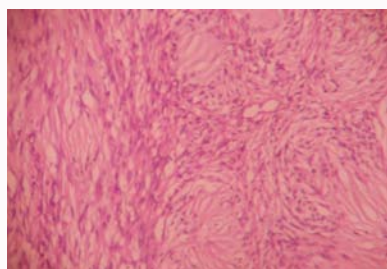
**Figure 5:** Total laryngectomy specimen showing tumour.

no malignant cells. With no diagnosis forthcoming from above investigations Ultrasound guided Fine Needle Aspiration Cytology was performed which was reported as Schwannoma and a deeper tissue biopsy was advised for confirmation (Figure 3). Accordingly patient was subjected to third biopsy and this time reported as Enchondroma.

Though the lesion was benign its location, site, extent and possible origin from cricoids with histopathology report as enchondroma prompted us perform a total laryngectomy. Tracheal stoma was created by taking bevelled shaped incision at 2<sup>nd</sup> tracheal ring. Neopharynx was created in a T shaped manner after preserving maximum right pyriform fossa mucosa. A 7.5 no portex cuffed tracheostomy tube and Ryle's tube number 16 was inserted at the time of creation of neopharynx.

The mass was firm globular to ovoid of about 5 cm x 5 cm x 4 cm in dimensions arising from the left Areepiglottic folds extending to and involving the left vocal cord, anterior commissure and cricoids causing bulge into pyriform fossa on the left side pushing epiglottis to the right side. No involvement of trachea was observed (Figure 6).

Patient was kept on broad spectrum antibiotics. Nebulisation with budacort, salbutamol and mucomix was given. Meticulous stomal care and wound dressing was done. Drain was removed on 3<sup>rd</sup> postoperative day. Ryle's feeding was started on 3<sup>rd</sup> postoperative day and continued upto 21 days. On 14<sup>th</sup> day oral feeds were started after ruling out pharyngocutaneous fistula. There were no perioperative complications. All in all patients made a remarkable recovery and discharged on 23<sup>rd</sup> postoperative day with a permanent healthy stoma. Patient was followed up after 15 days. Tissue was fixed in formalin



**Figure 6:** Postoperative histopathology showing schwannoma.



**Figure 7:** Stoma at the time of discharge.

for 48 hrs and sectioned in 3 mm slices. Final histopathology report was schwannoma of left recurrent laryngeal nerve showing spindle nuclear cells placed in palisades appearing like a verocay body (Figure 7) Patient is under follow up and doing well. We are planning for a secondary q234 Tracheosophageal Prosthesis for voice rehabilitation in coming few months.

## Discussion

Schwannoma is a slow growing, benign, encapsulated tumour arising from schwann cells. Laryngeal schwannoma is extremely rare and most commonly found in females in 4<sup>th</sup> to 5<sup>th</sup> decade of life [8]. However no age is immune to this entity. The case we are reporting is also a young female in her 3<sup>rd</sup> decade. Most commonly involved is the internal branch of superior laryngeal nerve [9,10]. Often however, this is not noticeable intraoperatively and likely to originate from the smaller distal nerve fibres in the laryngeal submucosa [10]. In this case too, parent nerve was not discernible. Schwannomas classically affect nerve sheaths rather than nerve fibres and hence symptoms are determined by site and mass effect [11,12].

Laryngeal schwannomas may approach a large size, causing upper airway obstruction, dysphonia and even vocal cord fixation, depending on their location, however most of them have insidious course [13-16]. Two distinct feature in our case were sudden hoarseness of voice and dull pain. It is said that dull pain is very typical of schwannoma [9].

Most common site is at areyepiglottic folds (80%), areytenoids, ventricular folds and true and false vocal cords (20%) [17,18]. The diagnostic work-up includes indirect laryngoscopy which usually reveals a submucosal mass. CT findings for benign laryngeal schwannoma usually include heterogenous enhancement of the lesion, no cartilage erosion and absence of infiltrative pattern [16]. The lesion is usually round to oval is attenuated with muscle and sharply demarcated [18]. In this case site of origin was AE folds. In

the quest of diagnosis, CONFLICTING inputs GIVEN BY VARIOUS investigations led to diagnostic impasse. Initially two biopsy were inconclusive while third biopsy was reported as enchondroma. Neoplastic etiology was suspected on Computes Tomography scan. Ultrasound and Ultrasound guided Fine Needle Aspiration Cytology revealed a diagnosis of Schwannoma. Variable opinions of various investigations compounded the diagnostic process. There are many cases in literature suggesting that Fine Needle Aspiration Cytology has low accuracy in the diagnosis of neural tumours [19-21]. But it was Ultrasound & Ultrasound guided Fine Needle Aspiration Cytology report which concurred with final histopathology report in our case. The preoperative diagnosis may be difficult. Intraoperative findings help us to finalize our diagnosis [12]. Enger and Weiss established three histological criteria for the diagnosis of schwannoma: encapsulation, presence of Antoni A and/or Antoni B stroma, and S-100 protein positivity [22].

Malignant transformation is rare in Schwannoma and is highly radioresistant [23]. Complete surgical resection is the treatment of choice. Depending upon the site size and location, a median or lateral thyrotomy or a median or lateral pharyngotomy are the various surgical approaches especially for tumours more than 5cm. For smaller tumor, endoscopic endolaryngeal CO<sub>2</sub> laser assisted excision can also be done [1,4,24,25]. Above procedures should be done without disturbing the laryngeal framework.

In our case the mass was involving left areyepiglottic fold, left areytenoid extending to left side cricoid cartilage and also crossing midline anteriorly. As any partial procedure would have made the laryngeal framework highly unstable, decision in favour of total laryngectomy was made. Our case is unique as laryngeal schwannoma is a rare entity, diagnosis was tortuous and tormenting and various investigations gave conflicting information adding to the diagnostic dilemma. Decision making for management was also challenging due to benign nature and extent of lesion. In light of final histopathology report agreeing with USG guided report, we feel later should be considered as a reliable implement while sailing through such a diagnostic crisis.

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