## Case Report

DOI: http://dx.doi.org/10.18203/issn.2454-5929.ijohns20182724

# Isolated paraglottic neurofibroma; lateral thyrotomy approach: a case report

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Received: 27 April 2018 Accepted: 30 May 2018

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#### **ABSTRACT**

Isolated neurofibromas are rare tumours of the paraglottic space. A 59 year old lady presented with progressive hoarseness for 5 years. Examination revealed a smooth submucosal globular mass in the glottosupraglottic region on the left side. After confirming findings on a contrast CT scan lateral thyrotomy approach was employed to excise the tumour in toto. Histopathology revealed a neurofibroma. Further workup to exclude neurofibromatosis (NF1) was done. Isolated laryngeal neurofibromas present a challenge to surgeons for providing complete excision of tumour while giving a good voice quality and lateral thyrotomy approach is the ideal approach for satisfying both these parameters.

**Keywords:** Neurofibroma, Lateral thyrotomy, Paraglottic

#### **INTRODUCTION**

Laryngeal neurofibromas are extremely rare tumors. They constitute 0.03 to 0.1% of benign tumors of larynx. Neurofibromas arise from aberrant proliferation of Schwann cells, fibroblasts and perineural cells found throughout the body. Most of these lesions are located in the supraglottis whereas glottis and subglottic occurrence is rare. In this case report we present a patient of supraglottic neurofibroma which was excised successfully by lateral thyrotomy approach.

### **CASE REPORT**

A 59 year old lady presented with progressive hoarseness over 5 years. She complained of inability to raise both pitch and intensity of her voice. Patient had no history of vocal misuse or dysphagia or symptoms suggestive of aspiration and had no addictions.

Indirect laryngoscopy and  $70^{\circ}$  telescopy revealed a smooth submucosal mass occupying left aryepiglottic fold and false cord region extending superiorly upto pharyngoepiglottic fold obliterating left pyriform fossa, inferiorly extending up to level of left true vocal cord; displacing it medially thus reducing glottic chink to 2 mm. Right vocal cord mobility was normal. There was no evidence of stridor.

Contrast enhanced CT scan revealed well defined rounded lesion measuring 2.7×2.3×3.1 cms [AP×TR×SI] predominantly solid with large cystic component in the left aryepiglottic fold and pyriform sinus filling the pyriform sinus extending caudally in paraglottic space displacing left false and true cord medially, contained within laryngeal cartilage, no extralaryngeal component or invasion of thyroid cartilage. On injection of contrast, solid component was seen to be enhancing. Differential diagnosis of minor salivary gland tumour vs. Schwanomma was described. Based on scan findings

patient was planned for excision of mass by lateral thyrotomy approach.

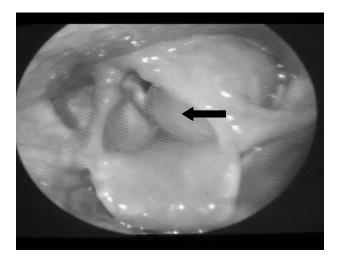


Figure 1: Preoperative 70 degree telescopy picture showing smooth submucosal mass occupying the left aryeepiglottic fold.

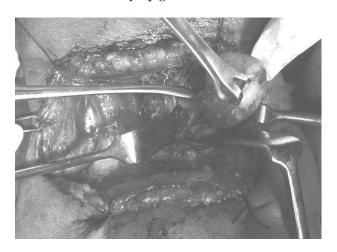


Figure 2: Intraoperative picture of tumour being dissected out by lateral thyrotomy approach.

In view of anticipated difficult airway elective tracheostomy was performed. Horizontal skin crease incision was given 1 cm below superior border of left thyroid lamina, subplatysmal flaps were raised, dissection done and left thyroid lamina exposed and a 2×2 cms window created in its upper third leaving a 5 mm margin from superior and inferior border. After carefully lifting the outer perichondrium inner perichondrium was incised to reveal a 3×2 cms smooth globular mass in left paraglottic space which was fairly easily dissected out and excised in toto without breaching laryngeal mucosa. Inner perichondrium was resutured, cartilage reposited and outer perichondrium closed.

Indirect laryngoscopic examination done on 3rd postoperative day revealed no laryngeal edema hence patient was decannulated on 5<sup>th</sup> post-operative day. Histopathologial examination revealed spindle shaped cells arranged in whorls and interlacing fashion suggestive of neurofibroma.

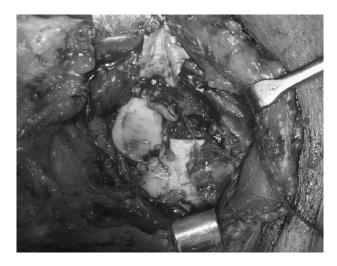


Figure 3: Reconstruction of the laryngeal framework.

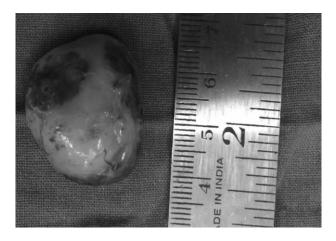


Figure 4: Specimen of paraglottic tumour dissected out in toto measuring 2.5×2.5 cms.

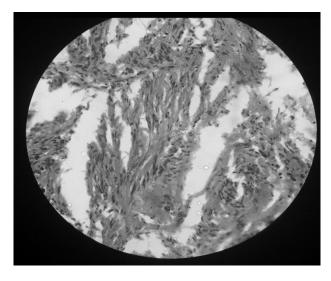


Figure 5: histopathological picture showing spindle shaped cells arranged in whorls and surrounding collagenous stroma.

Patient was regularly followed up with sessions of speech therapy with significant improvement in voice quality over 4 weeks. With diagnosis of neurofibromatosis in mind MRI Brain and pure tone audiometry was performed which were found to be normal. No other neurocutaneous stigmata were found.

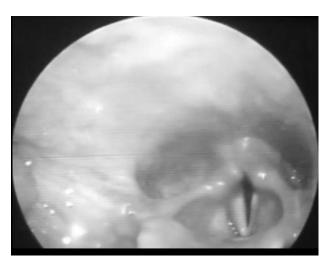


Figure 6: Postoperative 70 degree telescopy picture.

#### DISCUSSION

Neurogenic tumors of larynx are extremely rare.<sup>1</sup> Cases described are usually found in association with Von Recklinghausen disease.<sup>2</sup> Literature mentions less than 30 cases of endolaryngeal neurofibroma since its first description of Hollinger in 1950, majority of which have been described in paediatric population and in association with NF1.<sup>2</sup> Women are more commonly affected with male to female ratio of 2:3.<sup>3</sup>

In this case patient presented only with dysphonia however symptoms like dysphagia, stridor or foreign body sensation can occur. Laryngeal neurofibromas are most commonly found in the supraglottic region because of rich terminal nerve plexus.<sup>4</sup> Neurofibromas are divided into two subtypes plexiform and non plexiform. The plexiform variety is associated with neurofibromatosis type 1, is locally aggressive with surrounding infiltration making dissection difficult whereas non plexiform types are discrete, well defined and amenable to complete excision as in our case report. Plexiform variety is associated with high degree of recurrence and upto 10% cases are documented to have a malignant transformation. Recurrence and reresection are a rule with rate of malignant transformation being higher for plexiform than for non plexiform neurofibroma especially in the setting of neurofibromatosis 1.5-10

Main differential diagnosis of submucosal supraglottic swelling includes chondromas, adenomas, mucoceole, laryngocoele, lipomas, neurofibromas and Schwanommas. In this case given the prolonged history and CT scan findings a differential diagnosis of

neurogenic tumor vs. minor salivary gland tumor was considered. Malignant pathology was considered less likely due to prolonged duration of symptoms leading to rapid worsening. Intraoperative finding of tumor being easily dissectable from surrounding tissue was characteristic of non plexiform variety of neurofibroma as well as benign minor salivary gland tumor.

Most of the cases of endolaryngeal neurofibromas are diagnosed retrospectively.<sup>2</sup> Hence further workup of patient is warranted in the form of neurological and audiometric evaluation. MRI brain and PTA performed in this case were normal confirming this case to be of isolated neurofibroma.

The appropriate management of this tumor seems to be controversial while some advocate a more conservative approach in patients with minimal symptoms.<sup>6,7</sup> A surgical excision of such a tumor in the larynx is warranted given the concern for progressive airway obstruction. Various approaches have been described in literature for excision of paraglottic tumors which include transoral laser resection, lateral and medial thyrotomy approach. A transoral laser resection offers obvious advantage which includes a potential to avoid tracheostomy, shorter duration of surgery and shorter hospitalisation period. However it can be used only for smaller sized lesions and in centres where laser is available. Also plexiform variety if present will be difficult to dissect with minimal injury to surrounding tissue leading to poor voice quality post operatively. The traditional lateral thyrotomy approach which was employed by us provided a good surgical exposure enabling complete excision of tumor with good postoperative convalescence and voice quality as the laryngeal mucosa was not breached. The only drawback in our case was the requirement for tracheostomy due to unavailability of jet ventilation technique of anaesthesia at our centre.

#### **CONCLUSION**

We advocate lateral thyrotomy approach for excision of paraglottic tumors as a safe and effective approach providing adequate exposure and good and early voice rehabilitation.

Funding: No funding sources Conflict of interest: None declared Ethical approval: Not required

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Cite this article as: Naik AD, Lambor D, Shetgaunkar R. Isolated paraglottic neurofibroma; lateral thyrotomy approach: a case report. Int J Otorhinolaryngol Head Neck Surg 2018;4:1104-7.