

## Neurofibroma of Trachea: A Case Report

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**Abstract:** Neurofibroma of the trachea is a rare tumour. We present a case of a 55-year-old male with a solitary neurofibroma in the upper part of the trachea without any evidence of neurofibromatosis. The patient presented with dyspnoea on exertion of 6 months and intermittent coughing lasting for 2 years. The preoperative flexible bronchoscopy and computed tomographic (CT) scan neck revealed an intraluminal polypoidal mass with attachment on the right posterolateral tracheal wall with 2/3<sup>rd</sup> occlusion of the tracheal lumen. The mass was resected through a vertical tracheostomy approach using bipolar cautery under local anaesthesia. To our knowledge this is the first case reported in which tracheal neurofibroma is excised under local anaesthesia through vertical tracheostomy.

**Key words:** Solitary neurofibroma, trachea, vertical tracheostomy.

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### I. Introduction

Intratracheal tumours are relatively rare, and benign neurogenous tumours arising in the trachea are among the rarest[1]. Neurofibroma is a benign proliferation of Schwann cells, perineural cells, and fibroblasts[2]. The incidence of neurofibromas involving the trachea or bronchus is lower than that of neurilemmomas. In a search of the world literature, there were only 23 cases of tracheobronchial neurofibromas reported[3]. We report the surgical removal of an intratracheal tumour under local anaesthesia through a vertical tracheostomy approach.

### II. Case History

A 55-year-old male, a chronic smoker, was referred to our hospital with history of dyspnoea on exertion of 6 months and chronic cough on and off for about 2 years. There was no history of dysphagia, fever, loss of appetite, loss of weight, haemoptysis, and lymphadenopathy or aspiration symptoms. There was no significant past medical or surgical history. On physical examination, he was well and not in stridor. His head and neck, chest and cardiovascular examinations were unremarkable. His routine investigations were normal. Flexible bronchoscopic examination showed a well-defined, glistening, polypoidal mass, arising from the right posterolateral wall of upper trachea partially obliterating the tracheal airway (Fig 1). Computed tomography (CT) scan of neck revealed an intraluminal enhancing polypoidal mass (15 mm x 13 mm) arising from the posterolateral wall of the trachea at the level of second tracheal cartilage (Fig. 2).

In the operation theatre, patient was placed in supine position and standard monitoring including pulse oximetry, electrocardiography and non-invasive blood pressure was connected. An intravenous access was obtained followed by commencement of intravenous fluid. After local infiltration with about 8ml of 2% xylocaine with 1:2,00,000 adrenaline, a vertical skin incision was given in midline from the lower border of cricoid cartilage to the suprasternal notch. Strap muscles were lateralised from midline and isthmus of thyroid identified and retracted upwards. Tracheal rings were identified. Then 1ml of 4% xylocaine was introduced intratracheally. Tracheal rings from second to fourth were split in midline and retracted laterally with stay sutures. A well demarcated, pedunculated intratracheal mass was found attached onto the right posterolateral tracheal wall at the level of second tracheal cartilage. Cuffed endotracheal tube was inserted below the mass after applying 2% xylocaine jelly. Cuff was inflated and tube pulled downwards away from the tumour (Fig 3). The mass was excised using bipolar cautery. Seepage of blood and tissue distally into the tracheo-bronchial tree was prevented by using judicious suctioning above the cuff. Removal of intratracheal mass was completed within 5 minutes of tracheal incision. Endotracheal tube was removed and replaced with a cuffed tracheostomy tube before shifting the patient to the ward. The whole procedure was completed within 30 minutes during which time patient maintained oxygen saturation within normal range and did not require any general anaesthesia. Gross examination of the excised tumour appeared as a single grey, well-demarcated solid,

polypoidal mass, (2 cm × 1.5 cm) in size, covered by intact mucosa with no sign of hemorrhage or necrosis. On microscopic examination, the mass was lined by pseudostratified ciliated columnar epithelium. The subepithelium showed tumour cells with wavy buckled nuclei admixed with fibroblasts arranged predominantly in sheets, focal storiform and haphazard pattern. Interspersed in between were many blood vessels lined by flattened endothelium. Extensive areas of myxoid change of the stroma were also seen. Peripheral areas showed a few benign mucinous glands (Fig.4). Overall, these findings were suggestive of neurofibroma involving upper trachea.

Postoperative, he was well and nursed in normal ward. Decannulation of the tracheostomy tube was done on first post-operative day and wound closed by suturing with 3-0 ethilon. He was given intravenous antibiotics and oral analgesics for five days. The postoperative course was uneventful, and the patient was asymptomatic at the time of discharge. Follow up fiberoptic laryngoscopy at 2 months after surgery revealed no abnormality.

### III. Discussion

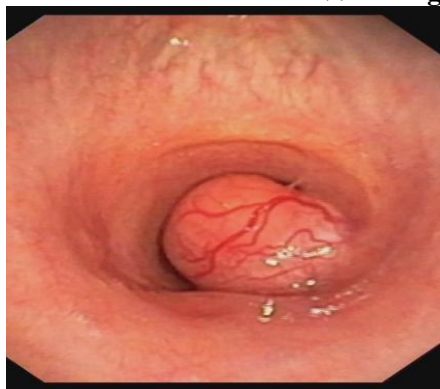
Papilloma, polyps, fibroma and hamartoma account for most of the benign tumours arising in the tracheobronchial tree, followed by leiomyoma and neurogenic tumours. Neurogenic tumours include neurilemoma (Schwannoma), neurofibroma and neuroma, of which neurofibromas of trachea are the rarest[3]. Neurofibromas are peripheral nerve sheath tumours that are typically characterised as benign, non encapsulated proliferations of schwann cells, perineural cells and fibroblasts. Depending on their patterns of growth, neurofibromas are classified as solitary, plexiform, diffuse, or a combination in which the diffuse and plexiform subtypes are closely associated with NF1.

Solitary neurofibromas as reported in our case, are sporadic, localized lesions arising from within the endoneurium. Histologically they are circumscribed lesions that mainly consists of elongated spindle cells with pale eosinophilic cytoplasm and wavy nuclei occasionally found in a fibromyxoid matrix[4].

The intratracheal tumours initially present as cough, wheezing, and dyspnoea, which are mostly related to airway obstruction as seen in our patient. Treatment depends on size and location of tumours. Small tumours can be removed endoscopically with or without laser, however if tumours invade adjacent tissue open surgery should be carried out [3]. In our case intraluminal polypoidal mass (15 mm x 13 mm) with attachment on the right posterolateral tracheal wall was resected through vertical tracheostomy under local anaesthesia with the general anaesthesia team on standby for any eventuality.

Tracheal neoplasm will often be misdiagnosed until at least 75% luminal obstruction occurs. A high index of suspicion for rare pathology, careful imaging techniques and adequate biopsy specimen are necessary for diagnosis when the presentation has unusual features. Complete resection improves chances for cure without recurrence[5].

### IV. Figures And Table



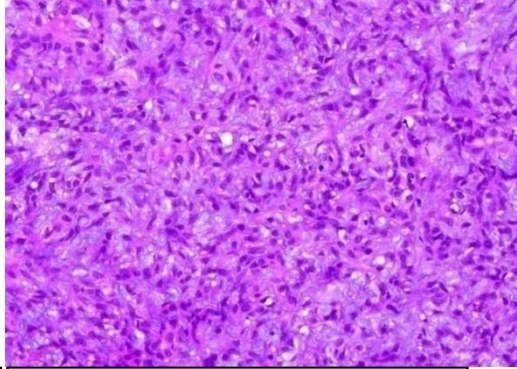
**Fig.1:** Fibreoptic bronchoscopy picture shows an intratracheal polypoidal mass with smooth and glistening surface



**Fig2.** Sagittal CT scan shows smooth intratracheal soft tissue mass based on the right posterolateral wall of the trachea



**Fig3.**Intraoperative picture showing intratracheal mass and endotracheal tube below the mass.



**Fig 4.**Tumour cells with wavy buckled nuclei admixed with fibroblast arranged in sheets.

## V. Conclusion

Tracheal tumours should be kept in mind for differential diagnosis during work-up of adult-onset dyspnoea and wheeze[6]. Appropriate surgical resection depending on the location and extent of the intratracheal tumour is the treatment of choice. High location of the intratracheal tumour, difficult intubation from above the mass, anticipated difficulty in endoscopic resection without a tracheostomy and possibility of resection through the same vertical tracheostomy incision without general anaesthesia should be considered before the taking up the surgery under local anaesthesia.

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