# Tracheal Tumors And Stridor: Case Report And Literature Review

Y Ramakrishnan, P Yates

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**Abstract**

Tracheal tumours, although rare, are treatable if diagnosed early. Presenting symptoms vary widely from exertional dyspnoea (most common), to hoarseness and stridor. These non-specific symptoms may lead to delayed diagnosis. If hoarseness or stridor are the dominant symptoms, then the patient is likely to be referred to the Ear Nose and Throat department. Diagnostic delay can occur due to failure to consider the diagnosis, or the tracheal lesion too small or distal to be directly visualised. This case report highlights the above pitfalls and looks into current management of primary malignant tracheal tumours.

## CASE REPORT

A 54 year old gentleman with chronic obstructive pulmonary disease (COPD) presented with 1 year history of progressive stridor, 2 months history hoarse voice and 2 weeks history of food sticking at the back of his throat. His stridor and hoarseness were intermittently worsened by exacerbation of his COPD, but were unresponsive to inhalers, steroids and antibiotics. He was initially refereed for assessment of his hoarseness. At that time flexible nasolaryngoscopy and chest xray did not reveal any abnormality. Past medical history of hypertension and perforated gastric ulcer was noted. His medication included inhalers, lansoprazole and antihypertensives. He previously smoked 50 rolls of tobacco per week and drank 4 cans of lager weekly.

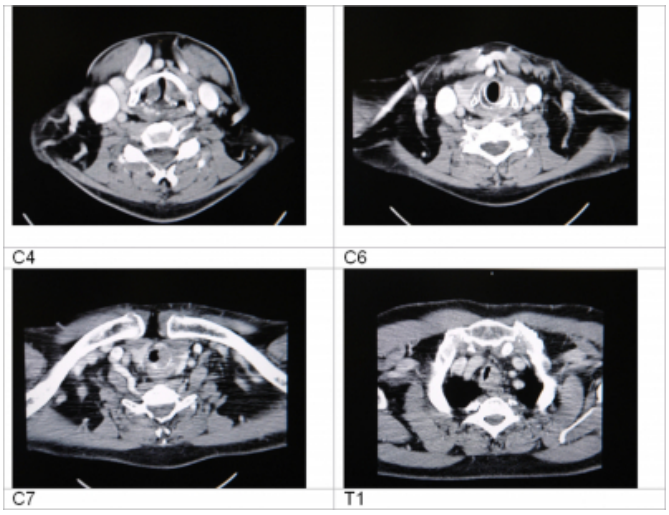
On examination, he had stridor at rest but was completing full sentences. He had a respiratory rate of 16 and normal oxygen saturation on pulse oximetry. His trachea was central with scattered wheeze throughout his chest. Systemic examination was unremarkable. Nasoendoscopy revealed a lesion in the cervical trachea and reduced mobility of left vocal cord. His CT showed a tracheal mass extending from the subglottis to the thoracic inlet; the airway being reduced to 3mm at its narrowest point. No direct invasion of the surrounding tissues and no nodal or pulmonary metastasis were noted.

In order to give symptomatic relief and obtain tissue diagnosis, he underwent urgent bronchoscopy and laser debulking of tracheal lesion. Intraoperative examination revealed a 95% stenosis in the cervical trachea but the thoracic trachea was normal. Biopsies revealed moderately differentiated squamous cell carcinoma (SCC) of trachea.

Treatment options were presented to the patient and he opted for primary chemoradiotherapy rather than surgical resection. He received 63Gy in 30 fractions with mitomycin. During the course of his radiotherapy, he developed further episodes of stridor and the tumour required further debulking on two occasions. Five months after completing radiotherapy, he underwent stenting for dyspnoea and stridor. One year after the original diagnosis, he appears to be progressing well with no further complications and no signs of recurrent or metastatic disease.

**Figure 1**

Figures 1-4: Tracheal lesions at different levels




## DISCUSSION

Primary tracheal tumours are very rare with an incidence of 0.2 per 100,000 per year and account for < 1% of all cancer deaths. In 1990, 44 deaths in England and Wales were caused by tracheal tumors, compared with 803 for carcinoma larynx and 34331 from lung carcinoma.[1] The largest published experience at Massachusetts General Hospital saw 198 cases treated over a 26 year period.[2]

90% of primary tracheal tumors are malignant. There are two main histologic types: squamous cell carcinoma (SCC) and adenoid cystic carcinoma (ACC) both presenting with equal frequency. The next most common primary malignant tumors were carcinoid tumors and mucoepidermoid carcinoma. The remaining 10% benign primary tumors are composed of squamous papilloma, pleomorphic adenoma, granular cell tumor, and benign cartilaginous tumors.

Mean age of presentation for SCC is 60 years whilst ACC occurs in younger individuals usually less than 50 years. Tracheal tumours are rare in children.[3] Male to female ratio is 4:1 and 1:1 in SCC and ACC respectively. No consistent aetiology has been found for tracheal primaries apart from benign squamous papillomas which is associated with viral infection. Most SCC tumors occur sporadically and probably follow a similar pattern of carcinogenesis as lung cancers.

Both tumours have different natural histories. Adenoid cystic carcinoma tends to be more insidious with slower growth pattern. Tumors may involve a large length of trachea by submucosal spread, often displacing mediastinal structures. Hematogenous metastases particularly to the lung are not uncommon. In comparison, SCC may be ulcerative or exophytic in appearance. It often directly invades mediastinal structures and tends to metastasize to regional lymph nodes.

The presentation of primary tumors of the trachea is variable. In a series of 329 patients with primary tracheal malignancies, exertional dyspnea was found to be the most frequent symptom (71%), followed by cough (40%), hemoptysis (34%), wheeze(19.5%), and stridor (17.5%). Symptoms related to involvement of adjacent structures, such as hoarseness and dysphagia, were less common. Repeated episodes of pneumonia that respond to antibiotics and physiotherapy can also occur. In the absence of hemoptysis, a diagnosis of adult-onset asthma is often made, thus delaying definitive treatment.

Tracheal lesions are best diagnosed by bronchoscopy or radiographic imaging. Tracheal evaluation using spiral CT is currently the imaging of choice because it allows accurate volumetric acquisition, unlike conventional CT which may miss small lesions or underestimate the longitudinal dimension of the tumor.

In addition, spiral CT may allow differentiation of mucosal from submucosal masses as well as displaying submucosal spread. Bronchoscopy has the advantage of allowing direct visualization and biopsy of the lesion in addition to treating it by debulking. Standard chest radiographs are insensitive in the detection of tracheal neoplasms. Spirometry can also play an important role in the diagnosis of symptomatic tracheal lesions, but characteristic abnormalities of the flow-volume loop may not appear until the airway is reduced to 8 mm. Moreover, this may be masked in the setting of small airways disease such as COPD.

Because symptoms are non-specific and CXR is often normal, diagnosis can be delayed anywhere from 2 months to 2 years. In one series, delayed diagnosis of more than 6 months after symptoms onset occurred in one third of patients. Patients are often misdiagnosed with asthma or emphysema. In this case report, this patient was treated for repeated exacerbation of COPD and had no obvious tracheal lesion on nasoendoscopy until a few months later. At this point, more than 90% of the airway was obstructed at initial presentation to the ENT department.

Primary surgery with adjuvant radical radiotherapy, when indicated, appears to provide optimal results, achieving 5 year survival rates of up to 72% in some studies.[4] If the lesion is too extensive, radiotherapy, bronchoscopic

debulking of the tumor, laser treatment, or internal stenting may provide palliation.[5] If the airway is critically obstructed, maintenance of airway via laser or other modalities should be carried out before full evaluation on the suitability of resection is carried out.

Curative tracheal resection is possible when tumour length is limited and extensive regional disease and distant metastasis is absent. This usually followed by adjuvant radiotherapy to limit locoregional recurrence.

In unresectable tumours, the aim is to maintain adequate airway and slow progression of disease. Primary radiotherapy is commonly used with doses greater than 60Gy shown to be effective.[6] Local destruction of the tumor can be accomplished by a various methods including bronchoscopic debulking, laser, cryosurgery, brachytherapy, photodynamic therapy or argon beam coagulation. Malignant strictures can be stented using T-tubes or a trachesotomy tube.

A recent study by Gaissert et al 2004 [7]compared survival rates after surgical resection or palliative treatment to compare prognostic factors. Of 191 patients who underwent resection followed by adjuvant radiotherapy, overall operative mortality was 7.3% and improved each decade from 21% to 3%. Overall 5- and 10-year survival in resected ACC was 52% and 29% (unresectable 33% and 10%) and in resected SCC 39% and 18% (unresectable 7.3% and 4.9%). They also found that in patients who had undergone complete resection, negative airway margins and ACC were associated with long-term survival superior to palliative therapy. Tumour length, lymph node status or type of resection did not appear to affect prognosis. This importance of complete resection and poor correlation of lymph node status is confirmed by Regnard et al.[8]

The outcome of patients receiving primary radiotherapy is poor; ACC having a better prognosis than SCC. The median survival of SCC receiving primary DXT varies from 5 to 8 months [9,10]; in one study 13% of patients lived more than 2 years[9]. Factors favourable for survival were the achievement of complete remission, the absence of mediastinal lymphnode involvement, and the use of additional brachytherapy.[11] Mean survival of adenocystic treated with radiotherapy alone was 6.2 years.[12]

## CONCLUSION

The rarity of tracheal primary malignancies poses diagnostic and therapeutic challenges to the clinician. An awareness of this problem, especially if the patient is refractory to treatment, should make one reconsider the original diagnosis and arrange the appropriate investigation before the malignancy reaches an advanced stage.

## ADDRESS FOR CORRESPONDENCE

Mr Y Ramakrishnan Senior House Officer ENT (Lewisham University Hospital) 10 Hospital Way Lewisham, London SE13 6UF Email: yujayr@doctors.org.uk

### References

1. Gelder CM, Hetzel MR. Primary tracheal tumours: a national survery. Thorax 1993 48:688-92
2. Grillo HC, Mathisen DJ Primary tracheal tumors: treatment and results. Ann Thorac Surg Jan 1990; 49(1): 69-77
3. Gaissert HA, Mathisen DJ, Grillo HC, Vacanti JP, Wain JC.Tracheobronchial sleeve resection in children and adolescents. J Pediatr Surg.1994 29(2):192-7; discussion 197-8.
4. Hazama K, Miyoshi S, Akashi A, Yasumitsu T, Maeda H, Nakamura K, Tada H, Matsuda H Clinicopathological investigation of 20 cases of primary tracheal cancer. Eur J Cardiothorac Surg. 2003 23(1):1-5
5. Allen MS alignant tracheal tumors. Mayo Clin Proc. Jul 1993 ;68(7):680-4
6. Mornex F, Coquard R, Danhier S, Maingon P, El Husseini G, Van Houtte P. Role of radiation therapy in the treatment of primary tracheal carcinoma. Int J Radiat Oncol Biol Phys. 1998 41(2):299-305
7. Gaissert HA, Grillo HC, Shadmehr MB, Wright CD, Gokhale M, Wain JC, Mathisen DJ. Long-term survival after resection of primary adenoid cystic and squamous cell carcinoma of the trachea and carina. Ann Thorac Surg. 2004 78(6):1889-97
8. Regnard JF, Fourquier P, Levasseur P. Results and prognostic factors in resections of primary tracheal tumors: a multicenter retrospective study. The French Society of Cardiovascular Surgery. J Thorac Cardiovasc Surg. 1996 111(4):808-13; discussion 813-4.
9. Chao MW, Smith JG, Laidlaw C, Joon DL, Ball D. Results of treating primary tumors of the trachea with radiotherapy. Int J Radiat Oncol Biol Phys. 1998 41(4):779-85
10. Manninen MP, Pukander JS, Flander MK, Laippala PJ, Huhtala HS, Karma PH. Treatment of primary tracheal carcinoma in Finland in 1967-1985 Acta Oncol. 1993 32(3):277-82
11. Schraube P, Latz D, Wannenmacher M. Treatment of primary squamous cell carcinoma of the trachea: the role of radiation therapy. Radiother Oncol. 1994 33(3):254-8
12. Maziak DE, Todd TR, Keshavjee SH, Winton TL, Van Nostrand P, Pearson FG Adenoid cystic carcinoma of the airway: thirty-two-year experience. J Thorac Cardiovasc Surg. 1996 112(6):1522-31; discussion 1531-2.
13. Henning A, Gaisser Primary tracheal tumor. Chest Surgery Clinics of North America. 2003 13: p247-256
14. Velez Jo ET, Morehead RS Hemoptysis and dyspnea in a 67-year-old man with a normal chest radiograph. Chest 1999 116(3): 803-7
15. Tracheal tumours Brian James Daley, MD, MBA, FACS, Mudiwa Munyikwa http://www.emedicine.com/med/topic2975.htm

## Author Information

**Y. Ramakrishnan, MRCS Ed**
Freeman Hospital

**P. Yates, FRCS (ED)**
Freeman Hospital