Tracheal Schwannoma Presenting as Subcutaneous Emphysema and Pneumomediastinum

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SUMMARY
Primary tracheal schwannomas are rare benign tumours. This is a case report, and therefore, no specific methods or results are applicable. We here report a case of a tracheal schwannoma in an early adolescent girl presenting with subcutaneous emphysema and symptoms of airway obstruction. Tracheal resection and reconstruction by primary anastomosis were performed. Pathology confirmed the diagnosis of tracheal schwannoma. This is an unusual life-threatening presentation of a benign rare tracheal tumour with a challenging approach to management.

BACKGROUND
Primary tracheal tumours are rare tumours with a significantly lower incidence rate compared with other tumours of the lungs and bronchial tree. They account for 1% of all tumours with the majority being malignant in nature. Squamous cell carcinoma and adenoid cystic carcinoma are the most common pathological subtypes.1

Benign tracheal tumours are even more rare, and they include papillomas, fibromas and haemangiomas with papillomas being the most common histological subtype.2 Little information is available with respect to their natural history and behavior.2

We hereby report a case of a benign tracheal schwannoma, in an early adolescent girl presenting with subcutaneous emphysema and pneumomediastinum.

Case presentation
A previously healthy girl in her early adolescence presented to our emergency department with hoarseness, choking sensation, and shortness of breath. She reports that her symptoms started immediately after eating crackers and ingesting alcohol at a local party. During this episode, she attempted forced coughing for relief without significant improvement. On further questioning, she reports that her dyspnoea has been progressively worsening over the past few months. This was evaluated by a cardiologist who attributed her symptoms to anxiety. She also has a non-intentional weight loss of 4 kg within 2 month period. She denies nausea, vomiting, or retching. There is no history of trauma, fall, or previous surgeries. Her only medication is isotretinoin acid for acne.

Investigations
The initial workup included a complete blood count, electrolytes, and a C reactive protein, which were normal except for magnesium of 1.1 mg/dL.

Flexible fibreoptic laryngoscopy through the left nasal cavity showed mobile vocal cords with normal motion and complete closure and was otherwise unremarkable.

A chest X-ray was then done, and it showed pneumomediastinum and emphysema in the neck and bilateral axillary areas (figure 1). Further evaluation by a non-enhanced chest CT scan revealed a moderate-sized pneumomediastinum, bilateral subcutaneous emphysema involving the upper anterior chest wall, bilateral axillary region and the base of neck in addition to a 1.7×1.4 cm well-circumscribed endotracheal soft tissue polypoid lesion at 5.4 cm above the carina almost completely obstructing the tracheal lumen (figure 2).

Treatment
After this critical finding, securing a definitive airway was the primary concern. A tracheostomy was not possible given that the tumour is at the level of the manubrium sterni. She underwent surgical debulking of the tracheal mass using a rigid bronchoscope by the ear, nose and throat surgeons. To secure her oxygenation during the procedure, she was placed on extracorporeal membrane oxygenation (ECMO) with peripheral cannulation.

During rigid bronchoscopy, a hard, immobile, submucosal midline tracheal mass was identified. The mass was debulked using the optical forceps and samples were sent for pathology. An examination of the oesophagus was also done. No masses were evident. An endotracheal tube was then inserted by the anaesthesiology team bypassing the mass, ECMO was discontinued, and the patient was transferred to the intensive care unit.
Case report

The pathology result was consistent with schwannoma. The tumour cells were positive for S-100, SOX-10, and vimentin (figure 3).

The case was discussed in the thoracic tumour board and surgery was preferred over endoscopic resection and radiotherapy given the submucosal extension of the tumour and the increased risk of recurrence with such procedures. She underwent tracheal resection and reconstruction by primary anastomosis. She was extubated in the operating room and transferred to the floor where she had a smooth recovery until discharge.

Outcome and follow-up

The patient was followed up until writing this manuscript. She reported doing well without the recurrence of her symptoms. A follow-up rigid bronchoscopy was done 3 months later for the removal of tracheal sutures showed no evidence of recurrence.

DISCUSSION

Pneumomediastinum, the presence of air in the mediastinum, occurs when there is leakage of air from any site along the respiratory tract or from an intrathoracic or intraabdominal hollow organ. While it was recently observed with increasing frequency in COVID-19 patients during the pandemic; pneumomediastinum can have a range of underlying causes.

Pneumomediastinum can occur spontaneously due to a sudden increase in intrathoracic pressure, interalveolar pressure and the subsequent rupture of alveoli without an apparent clinical cause, it is referred to as ‘Hamman’s syndrome’. However, the majority of cases are secondary pneumomediastinum associated with underlying conditions affecting the tracheobronchial tree, alveoli or oesophagus. These secondary causes include trauma, alveolar rupture due to conditions that cause a sudden increase in intra-alveolar pressure such as asthma and COPD, medical procedures (such as positive pressure ventilation, bronchoscopy, or endoscopy), oesophageal rupture (as seen in Boerhaave’s syndrome), and specific pulmonary infections such as pneumocystis are also known to cause pneumomediastinum.

While there have been limited reports of pneumomediastinum linked to endobronchial tumours (such as angiofibromas, Hodgkin’s lymphoma and carcinoid tumours), there is currently no existing literature documenting endotracheal tumours as a cause of secondary pneumomediastinum.

We hypothesize that this rare occurrence of secondary pneumomediastinum in association with an endotracheal tumour can be attributed to multiple factors.

1. The large calibre of the trachea as compared with the main and segmental bronchi prevets the sudden excessive rise of interalveolar pressure.

Figure 1 Chest X-ray showing pneumomediastinum (arrow) and subcutaneous emphysema (circle) in the lower neck and bilateral axillary areas (circle).

Figure 2 From left to right, CT scan of the chest showing: Pneumomediastinum and subcutaneous emphysema in the neck, bilateral axillary areas and anterior chest wall (A); A well-circumscribed soft tissue polypoid lesion 1.7×1.4 cm obstructing around 90% of the tracheal lumen located 5.4 cm above the carina (B); Coronal axis showing the tracheal tumour (C); Sagittal view of the same scan (D).
2. The integrity and patency of the tracheal wall are preserved by the existence of the cartilaginous rings, a feature not found in the segmental bronchi.

3. The appearance of earlier upper airway symptoms (dyspnoea, stridor, cough...) would bring attention to an underlying pathology before near-total obstruction of the tracheal lumen and subsequent pneumomediastinum. Thus, secondary pneumomediastinum depicts a late presentation of endotracheal schwannoma in our patient.

Benign tracheal tumours account for a quarter of primary tracheal tumours. Neurogenic tumours of the tracheobronchial tree are extremely uncommon, and they include neurofibromas and schwannomas. Schwannomas are uncommon benign nerve sheath tumours often arising in the posterior mediastinum and costovertebral angle. Tracheal schwannomas are extremely rare, lung and bronchial schwannomas being more common. It is rarely associated with neurofibromatosis type I and is typically well-circumscribed and encapsulated tumours.

The first tracheal schwannoma was reported by Straus in 1951. A literature review by Ge et al, conducted between 1950 and 2013, identified only 51 cases of tracheal schwannomas, 2 of which were malignant. In this review, a female predilection was noticed, with the distal third of the trachea being the most commonly affected site, followed by the proximal than the mid-third of the trachea. The size of the tumour ranged from 1 to 4 cm, with the majority being less than 3 cm.

A classification of pulmonary schwannomas was proposed by Kasahara et al. Tracheal lesions and lesions visualized during bronchoscopy in the proximal bronchi were classified as central. Central lesions are also divided into two subtypes: (1) tumours that exist only in the intraluminal space and (2) tumours that occur in both intraluminal and extraluminal space.

Schwannomas are slowly growing tumours, usually presenting with symptoms of airway obstruction (wheezes, stridor, dyspnoea). Other symptoms may include hoarseness and hemoptysis. Affected patients are usually misdiagnosed with asthma, with a 17-month average delay in diagnosis, and are usually diagnosed when the patient fails to respond to standard treatment of obstructive lung disease.

The presentation with subcutaneous emphysema and pneumomediastinum (our patient above) is unusual and was not reported previously in the literature.
The diagnosis of this benign tumour is usually done by direct visualization of the trachea through bronchoscopy, and pathological confirmation by a subsequent biopsy which accounts for the potential risk of upper airway obstruction with endoscopic procedures. CT is potentially helpful in the identification of the site, size and extent of the tumour. When spirometry is done, tracheal schwannomas show a fixed upper airway obstruction on flow volume loops with flattening of both the inspiratory and expiratory limbs.

Different modalities have been described in the management of tracheal schwannomas, including primary tracheal resection or endoscopic treatment such as laser with or without a CO2, electrocautery snaring, argon plasma coagulation, cryotherapy, endoscopic excision and microdebridement. In our case, and in view of the submucosal involvement, primary surgical correction was felt to be the best for this young patient.

Patients with a pedunculated tumour can be treated by endoscopic resection, followed by bronchoscopic surveillance with possible recurrence in one-quarter of the cases. The online supplemental table summarises the characteristics and treatment modalities used for the few rare previously reported cases of benign tracheal schwannomas.

### Learning points

- In conclusion, we report an unusual life-threatening clinical presentation of a benign tracheal schwannoma lesion.
- Even though a tracheal schwanna is a benign tumour, sometimes it can have a life-threatening presentation.
- Different modalities were described for the management of tracheal schwannomas.
- The best approach is an individualised strategy based on patient and tumour characteristics.

### REFERENCES