Case Report

Tracheal Tumour: An Unusual Cause of Severe Dyspnoea Treated as Severe Asthma

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Abstract

Inflammatory myofibroblastic tumours (IMTs) are rare neoplasms with benign clinical course. Although the aetiology is unclear, it is believed that IMTs are true neoplasms rather than a reactive or inflammatory lesion. We present the case of a 43-year-old female who presented to us with respiratory symptoms. She was being treated for bronchial asthma for the preceding nine months. Diagnostic testing revealed pedunculated tracheal tumour on computed tomography of the chest. Tracheostomy and tumour coblation was performed under general anaesthesia and histopathology revealed IMT.
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Key words: Inflammatory myofibroblastic tumours, Benign, Pedunculated, Coblation.

Introduction

Primary tracheal tumours are relatively rare and are usually malignant (80%–90%) in adults[1] and benign (60%–70%) in children.[2] These make up only about 2% of all tumours that arise from the upper airways. Most of the tracheal tumours are squamous cell carcinomas or adenoid cystic carcinomas. Inflammatory myofibroblastic tumour (IMT) is rare constituting 0.04%–0.07% of all respiratory tract tumours and usually presents in children under 16 years of age.[3] As patients with tracheal tumours initially present with non-specific respiratory symptoms, definitive diagnosis is often delayed. Chest computed tomography is an important imaging modality to diagnose and stage patients with suspected tracheal neoplasms. Bronchoscopy is essential to make histopathological diagnosis. Persistent or progressive local disease can cause complications, like haemorrhage, tracheal stenosis, or oesophageal-tracheal fistula.

Case Report

A 43-year-old female with no known co-morbidities, presented with dyspnoea and episodes of haemoptysis since preceding 8-9 months. She was being treated symptomatically before reporting to us. At presentation, stridor was the only abnormal finding on physical examination. She was unable to perform pulmonary function testing because of respiratory distress. Laboratory investigations were normal. Chest radiograph was normal. Thereafter, computed tomography (CT) of the chest was done which revealed endotracheal mass obliterating the lumen of the trachea (Figures 1A & B).

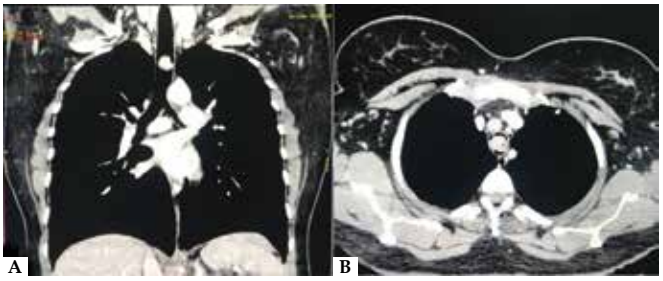

Figure 1. Computed tomography of the chest (coronal and axial view) showing tracheal tumour obliterating >95% of its lumen.

Flexible fibreoptic bronchoscopy showed a smooth-surfaced, pedunculated mass arising from the posterior tracheal wall in the lower third of the trachea (Figure 2A). The bronchoscope could not be negotiated beyond the tumour. Tracheostomy followed by tumour excision with coblation was done under general anaesthesia. On histopathological examination (Figure 2B), diagnosis of IMT was made. Immunohistochemistry demonstrated positivity for smooth muscle actin (SMA), caldesmon and anaplastic lymphoma kinase-1 (ALK-1) which confirmed IMT. The patient's symptoms resolved completely and subsequently tracheostomy tube was removed on third post-operative day. Check bronchoscopy was done before discharge which was essentially normal.

Discussion

Primary tracheal tumours account for 0.1%-0.4% of all malignant diseases.[4] IMTs, a rare variant constitute 20% of all primary lung tumours and 57% of all benign lung tumours.

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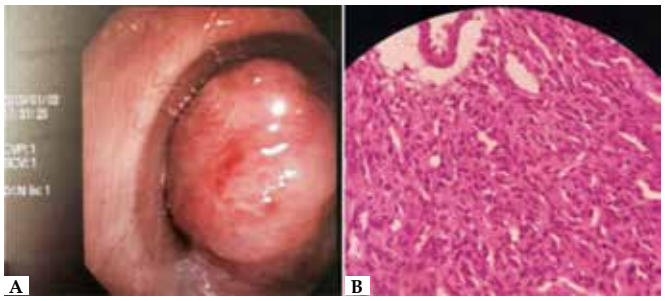

Figure 2. (A) Bronchoscopic view of the tracheal tumour and (B) photomicrograph of the excised tracheal tumour showing inflammatory myoblastic tumour (Haematoxylin and eosin, X 400).

The various other name of this entitiy include inflammatory pseudotumour, histiocytoma, fibroushistiocytoma, xanthoma, xanthofibroma, xantogranuloma and plasma cell granuloma.[5] Although the lung is the most common site for these tumours, IMTs may also develop in other sites, such as stomach, orbit, mesentery, heart, gastrointestinal tract, adrenal gland and central nervous system.

The symptoms of IMTs are usually non-specific and depends on the site and size of the lesion. Patients usually present with respiratory symptoms, such as dyspnoea, stridor, chronic cough, haemoptysis and pleuritic chest pain. In the small subset of patients with endobronchial lesions, the presentation may be acute due to post-obstructive pneumonia or symptoms associated with airway obstruction. Some cases have been mis-diagnosed and treated as asthma[6], as in our case.

Radiographic investigations with chest radiograph, CT and endoscopy are the used in the diagnostic work-up of obstructive tracheal lesions.[7] Average diameter of tumour ranges from 5cm to 10cm, although tumours up to 20cm have also been reported.[8,9] Macroscopically, these appear lobular, multinodular and hard masses. In our case, the tumour was pedunculated, round, smooth surfaced, about 1.5 centimetres and above the carina. Histologically, the tumour is composed of myofibroblastic spindle cells infiltrated by plasma cells, lymphocytes and eosinophils. IMTs are locally invasive tumours with a recurrence rate of 18% to 40% and have metastatic potential.

Diagnosis is usually delayed as symptoms are non-specific. Management includes interventional endoscopy, surgery, radiotherapy and endoluminal brachytherapy. However, complete surgical resection is the gold standard.[10] This is preferable modality to endoscopic resection particularly when the tumour is not pedunculated or seems to grow deep into the wall. Tumour resection can be done either bronchoscopically with biopsy forceps, carbon dioxide laser or open surgical intervention with segmental tracheal resection. Adjuvant radiotherapy and chemotherapy are reserved for the aggressive variants. The prognosis after radical resection is excellent. Radiotherapy alone is a possible treatment option in inoperable cases. Endobronchial brachytherapy may be used for tracheal tumours[11], especially if the tumour is small in size. The overall prognosis remains good but patient needs regular follow-up to detect early recurrence.

In conclusion, Inflammatory myofibroblastic tumour are a rare variant involving the airways and should be considered as differential diagnosis in cases presenting with respiratory distress due to upper airway obstruction. Complete surgical resection is the gold standard. Radiotherapy and chemotherapy should be considered after surgery in case of metastasis or recurrence or in cases where tumour is not amendable to resection. It is only through vigilant observation, knowledge and clinical skills, a physician can diagnose and treat timely with certainty, as our patient was mis-diagnosed initially requiring frequent hospital visits and compromising her quality of life.

References

1. Grillo HC. Primary tracheal tumours. In Grillo HC, editor *Surgery of the Trachea and Bronchi,* 4th edition. London: BC Decker; 2004:pp208–47.
2. Gilbert JB, Mazzarella LA, Feit LJ. Primary tracheal tumours in infants and children. *J Pediatr* 1949;35:63–69.
3. Wenig BM, Devaney K, Bisceglia M. Inflammatory myofibroblastic tumor of the larynx: a clinicopathologic study of eight cases simulating a malignant spindle cell neoplasm. *Cancer* 1995;76:2217–29.
4. Cerfolio RJ, Allen MS, Nascimento AG, Deschamps C, Trastek VF, Miller DL, *et al.* Inflammatory pseudotumors of the lung. *Ann Thorac Surg* 1999;67:933–6.
5. De Palma A, Loizzi D, Sollitto F, Loizzi M. Surgical treatment of a rare case of tracheal inflammatory pseudotumor in pediatric age. *Interact Cardiovasc Thorac Surg* 2009;9:1035–37.
6. Andrade FM, Abou-Mourad OM, Judice LF, Bento CA, Schau B, Carvalho AC, *et al.* Endotracheal inflammatory pseudotumor: the role of interventional bronchoscopy. *Ann Thorac Surg* 2010;90:e36–7.
7. Bumber Z, Jurlina M, Manojlovic S, Jakić-Razumović J. Inflammatory pseudotumor of the trachea. *J Pediatr Surg* 2001;36:631–4.
8. Fletcher CDM. Tumors of soft tissue. In: *Diagnostic Histopathology of Tumors;* 4th edition. USA: Elsevier Saunders; 2013:pp 1823–4.
9. Weiss SW, Goldblum JR. Fibrous tumors of infancy and childhood. In: Weiss SW and Goldblum JR, editors *Enzinger and Weiss's Soft Tissue Tumors*; 5th edition. USA: Mosby Elsevier; 2008:pp 284–9.
10. Fabre D, Fadel E, Singhal S, Montpreville VD, Mussot S, Mercier O, *et al.* Complete resection of pulmonary inflammatory pseudotumors has excellent long term prognosis. *J Thorac Cardiovasc Surg* 2009;137:435–40.
11. Abbate G, Lancella A, Contini R, Scotti A. A primary squamous cell carcinoma of the trachea: case report and review of the literature. *Acta Otorhinologica Italica* 2010;30:209–12.