Inflammatory myofibroblastic tumour: a rare cause of central airway obstruction

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DESCRIPTION

A 70-year-old woman with no comorbidities presented with cough, wheezing, exertional dyspnoea and a sensation of throat fullness. She never smoked and was recently diagnosed with asthma based on a normal chest radiograph and pulmonary function testing but failed to improve with bronchodilators and steroids. Her initial chest radiograph was unremarkable, and spirometry suggested a fixed intrathoracic obstruction. CT of the chest obtained after 2 months showed an irregular density in the trachea, 1.4×1.7 cm (figure 1). Flexible bronchoscopy showed a polypoid mass 2–3 cm above the carina adherent to the right wall of the trachea (figure 2). Rigid bronchoscopy with neodymium-doped yttrium aluminium garnet (Nd:YAG) laser photoresection was performed. Given the vascular nature of the tumour, removal was complicated by significant bleeding, which required endobronchial epinephrine and laser coagulation. The patient recovered without further complications. Immunohistochemical staining on the excised tumour was positive for vimentin and Anaplastic-Lymphoma Kinase-1 (ALK-1). The diagnosis was inflammatory myofibroblastic tumour (IMT). Her symptoms resolved completely after resection. Multiple CTs and surveillance bronchoscopies showed no recurrence.

IMTs (or inflammatory pseudotumors) represent less than 1% of all lung tumours and the pathogenesis is unknown.[1] They are often composed of smooth muscle cells with a lymphocytic or plasmacytic infiltrate. They are classified as tumours of intermediate malignant potential given invasion of local tissue.[2] Complete removal, often via surgical resection, remains the treatment of choice. Recurrence is rare, generally seen in cases of incomplete resection.[2] ALK gene rearrangements are common, and treatment with newer ALK inhibitors including crizotinib has been proposed for patients not amenable to surgical or bronchoscopic removal.[3]

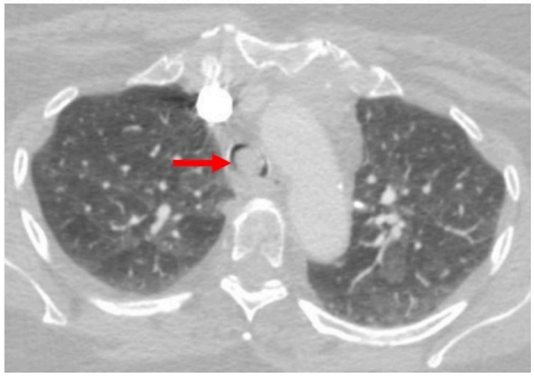

Figure 1 Axial image from chest CT with intravenous contrast revealing tumour in the trachea proximal to the carina (red arrow) with near complete occlusion of the airway.

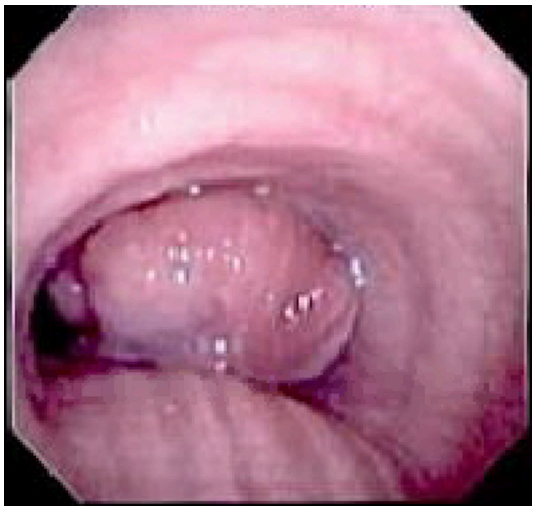

Figure 2 Image seen on flexible bronchoscopy of polypoid tracheal mass, corresponding to lesion identified on chest CT.

Learning points

- Tracheal tumours can present with wheezing and an obstructive pattern on pulmonary function testing, mimicking other causes of obstructive lung disease.
- Inflammatory myofibroblastic tumour is a rare tumour of intermediate malignant potential composed of a mixed inflammatory cell infiltrate.
- Tumour removal is the treatment of choice, with an excellent prognosis after resection in the majority of patients.

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REFERENCES

1 Icmeli OS, Alpay LA, Gundogus B, *et al*. Inflammatory myofibroblastic tumor: a rare tumor of the lung. *Eur Clin Resp J* 2004;1:25390.
2 Thistlethwaite PA, Renner J, Duhamel D, *et al*. Surgical management of endobronchial inflammatory myofibroblastic tumors. *Ann Thorac Surg* 2011;91:367–72.
3 Jindal A, Bal A, Agarwal R. Inflammatory myofibroblastic tumor of the trachea in the pediatric age group: case report and systematic review of the literature. *J Bronchology Interv Pulmonol* 2015;22:58–65.