Fibrohistiocytoma: a rare tumour of the trachea

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Learning Point for Clinicians

Fibrohistiocytomas of the trachea are rare tumours of the trachea. It may be difficult on histology to distinguish benign and malignant disease and expert opinion is required. These tumours can be successfully excised through interventional rigid bronchoscopy.

Case report

A 54-year-old Caucasian male was referred with haemoptysis. There was no associated dyspnoea, noisy breathing or loss of weight. His medication consisted of montelukast and loratidine for allergic rhinitis. Family history included his father having a pituitary tumour and grandfather having a liver tumour. He had no known allergies, and he had never smoked. He was a retired electronics engineer and may have been exposed to asbestos and beryllium in the past. Examination of the pharynx, cardiovascular, respiratory and gastrointestinal systems was normal.

Investigations revealed a normal full blood count, coagulation profile, inflammatory markers and biochemistry. A postero-anterior chest radiograph was unremarkable, and the patient had flexible bronchoscopy to identify a cause of his haemoptysis. At bronchoscopy, an exophytic lesion of the trachea just below the vocal cords was seen. Computed tomography (CT) demonstrated a small polyp in the posterior wall of the trachea just below the vocal cords, and subsequent positron emission tomography-CT (PET-CT) demonstrated that the lesion was not 18-fluodeoxyglucose avid.

The tracheal lesion was excised using a carbon dioxide laser delivered through a micromanipulator attached to an operating microscope, using a line-of-sight technique during suspension laryngoscopy, with supraglottic high-frequency jet ventilation1 under general anaesthesia (Figure 1A). Macroscopic appearance was a 10 x 8 x 6 mm firm exophytic lesion. Microscopically, the lesion was composed of spindle cells with indistinct margins and large vesicular oval nuclei, described as epithelioid and histiocytoid in appearance. There was focal necrosis but no granulomas were seen, and Ziehl Neelson and diastase resistant periodic acid-Schiff histochemical stains for acid-fast bacilli and fungi were negative. Mitotic figures were identified (2 per 10 high-power fields counted) (Figure 1B). Immunohistochemistry was not diagnostic. It showed patchy staining for S100 protein and smooth muscle actin and weak focal staining for epithelial membrane antigen. A striking presence of CD1a positive cells and a smaller proportion of CD68 cells were noted. The lesion was negative for skeletal muscle markers and epithelial markers. No neoplastic lymphoid process was demonstrated on
an extensive panel of immunostains. Molecular genetics (interphase fluorescence in situ hybridization) for the EWSR1 and FUS gene rearrangement associated with rhabdomyosarcoma was negative by interphase fluorescence in situ hybridization.

The slides were reviewed by a number of pathologists with specialist interest in head and neck pathology and soft tissue pathology. All agreed that this was a spindle tumour with fibrohistiocytic morphology but there was concern as to whether the features represented an entirely benign lesion. The pathology review taken together with the clinical and radiological findings in the multidisciplinary team setting supports a diagnosis of benign fibrous histiocytoma occurring at a highly unusual site.

At review (6 months), the patient was asymptomatic and on endoscopy, no residual lesion was visualized.

Discussion
Primary tumours of the trachea are rare, and primary sarcomas of the trachea are exceedingly rare with the majority being reported in patients <18 years of age.2 Common symptoms on presentation include dyspnoea, cough, stridor and haemoptysis. Most are proximal in the trachea as in this case. Chest radiography may not always demonstrate the presence of the lesion in the trachea, especially if only the postero-anterior view is obtained but CT in most cases is helpful at identifying the endotracheal lesion. The biological behaviour of fibrous histiocytomas of the trachea is unclear and the histological appearances can present diagnostic difficulty as in our case. Histologically similar cases have been interpreted as benign and malignant.3,4 In all but two of the adult cases reported the histology was that of a malignant tumour.

The management of these tumours poses a clinical dilemma due to the lack of understanding of their natural history. Some have been managed with radical surgery3,5, whereas others have been treated more conservatively with endoscopic resection.4,6 In this case, the histology was thought to favour a benign tumour, and the patient was managed endoscopically but will remain under long-term surveillance so as to facilitate early salvage treatment in the event of recurrent disease.

This case highlights the need to establish an international registry for such rare tumours with data on long-term clinical outcomes. The clinical information accumulated from such a registry will greatly facilitate appropriate management decisions in the future.

Conclusion
Fibrohistiocytomas are extremely rare tumours of the trachea especially in adults. The diagnosis can be difficult to ascertain histologically and expert opinion is required. These tumours can be successfully excised through interventional rigid bronchoscopy.

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References