Tracheal schwannoma: management by endoscopic laser resection

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Abstract
The case history is presented of a patient with a tracheal schwannoma who remains free of disease five and a half years after laser resection. This illustrates the potential of this approach in the management of tracheal schwannoma.

Tracheal schwannomas are among the rarest of tracheal tumours, and there is no unanimity of opinion regarding their treatment. We report the successful long term management of one such tumour by bronchoscopic laser resection.

Case report
A 45 year old man was referred for laser bronchoscopy because of progressive dyspnoea and wheezing. He was only recognised to have stridor after six months of unsuccessful treatment with bronchodilators and corticosteroids. On examination he had stridor and severe dyspnoea in the supine position, but no signs and symptoms when he sat upright. Tracheal tomographic scans showed a well circumscribed 2-0 cm mass filling the lumen of the cervical trachea (fig 1).

At rigid bronchoscopy, performed under general anaesthesia, there was a 2-0 cm pedunculated polypoid mass arising from the posterior wall of the trachea producing near total obstruction of the airway. The base of the tumour stalk was located less than 2-0 cm from the vocal cords, far more proximal than would have been expected from the preoperative radiographs. Because the tumour was pedunculated it was mobile within the tracheal lumen, accounting for the positional nature of his signs and symptoms. The tumour was resected by NdYAG laser and the patient experienced dramatic resolution of his symptoms.

Pathological examination showed a spindle cell neoplasm with features characteristic of a schwannoma. The tumour cells were arranged in short intersecting fascicles and displayed some tendency to surround collagen fibrils. Focal nuclear palisading was present (fig 2). Immunocytochemically the tumour cells were positive for $100$ protein, laminin, nerve growth factor receptor, and vimentin, but were negative for neurofilaments, muscle specific actins, cytokeratins, and melanoma related antigen (HMB45). Ultrastructurally the cells were elongated and invested with multiple layers of basal lamina. They had long interdigitating cell processes with occasional junctions. Mesaxon formation was present but no nerve axons were seen. Flow cytometry showed a low proliferative rate and no aneuploidy.

Because of the proximity of the tumour to the vocal cords, and because the endoscopic resection appeared to have been complete, the patient has been followed by bronchoscopy every 6–12 months. There has been no evidence of tumour regrowth during the five and a half years since his initial laser bronchoscopy.

Discussion
Neurogenic tumours are among the rarest tracheal neoplasms. A series of 44 tracheal tumours treated at the University of Toronto over 20 years included one neurofibroma,1 and a series of 50 tracheobronchial tumours seen at the Peking Union Medical College over 24 years included two neurogenic tumours.2 In the largest series, reported by Grillo from the Massachusetts General Hospital, there were two neurogenic tumours out of a total of 198

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Figure 1  Tracheal tomographic scan showing a smooth, well circumscribed 2-0 cm mass (arrows) filling the proximal tracheal lumen.
primary tracheal tumours seen over a 26 year period.¹

Neurogenic tumours are classified as neurofibromas or nerve sheath tumours on the basis of their histological appearance and the presence of nerve axons.ª Neurofibromas show proliferation of all elements of the nerve, including schwann cells, perineural cells, and nerve axons. Nerve sheath tumours are composed solely of nerve sheath cells²³ and, on the basis of S100 staining, are subdivided into true schwannomas (neurilemmomas) and non-schwannian nerve sheath tumours. Our patient’s tumour had the appearance of a true schwanna and lacked any histological features of malignancy. Tracheobronchial neurofibromas can be multiple, are sometimes associated with von Recklinghausen’s disease, and, rarely, may undergo malignant degeneration. Schwannomas are solitary, encapsulated, and benign. Tracheal schwannomas are rare: our patient is the twentieth reported case.

The most common site for both types of neurogenic tumours is the distal trachea, followed by the proximal trachea and the middle third of the trachea. The signs and symptoms are similar to those associated with other tracheal tumours, and include shortness of breath, cough, wheezing, and occasionally, haemoptysis. Because the clinical presentation is non-specific and insidious, diagnosis is often delayed for up to a year after the onset of symptoms. Endoscopically and radiographically these tumours usually present as a discrete, pedunculated intratracheal mass, but are sometimes sessile and can have an extratracheal component.¹²⁶ Our patient therefore had a typical clinical presentation.

Tracheal schwannomas have been treated both by formal tracheal resection and by endoscopic excision. Laser resection has not been reported previously. It is difficult to determine what is the optimal treatment because schwannomas are so rare, and because patient follow up has often been short. Local recurrence has been reported 12 years after initial endoscopic excision, suggesting that patients treated in this manner should be kept under bronchoscopic surveillance. The recurrent tumour was a sessile mass with a large extraluminal component. The choice of treatment should probably be influenced by the clinical presentation of the tumour (that is, pedunculated v sessile), the risk of tracheal resection, and the presence or absence of an extratracheal component. Our experience emphasises the benign nature of tracheal schwannomas and illustrates that laser resection and bronchoscopic surveillance is an acceptable approach to the management of this tumour when it is pedunculated and has no demonstrable extratracheal component.

References

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Notes

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