

Case report

Intramural neurofibroma of the trachea treated by multiple stents

J H Cranshaw, C Morgan, G Knowles, A G Nicholson, P Goldstraw

Abstract

The case history is presented of a patient in whom an intramural tracheal neurofibroma developed, causing severe airway stenosis. The patient was treated with multiple stents over a period of 5 years because of progression of the disease and associated airflow limitation. Clinicians should be aware of this rare complication of neurofibromatosis.

(*Thorax* 2001;56:583-584)

Keywords: neurofibromatosis; airway obstruction; stent

Neurofibromatosis may result in neurogenic tumours arising from any of the nerves in the mediastinum, chest wall, or lung parenchyma. However, clinical presentation resulting from involvement of the upper airways is exceptionally rare. We present a case of tracheal neurofibromatosis causing severe airway stenosis.

Case report

A 48 year old woman with Von Recklinghausen's neurofibromatosis was referred for investigation of upper tracheal narrowing. She was a smoker of five pack years with a two year history of deteriorating breathlessness on exercise culminating in dyspnoea at rest. There had been no significant improvement after inhaled bronchodilators or a course of prednisolone. Maximum flow volume loops and respiratory function tests indicated upper airway obstruction (forced expiratory volume in one second (FEV₁) 2.24 l (predicted 3.26 l); forced vital capacity (FVC) 4.57 l (predicted 3.79 l); FEV₁/FVC 0.49; peak expiratory flow rate (PEFR) 3.29 l/s (predicted 7.91 l/s); peak inspiratory flow rate (PIFR) 3.40 l/s (predicted 4.82 l/s); PIFR/PEFR 0.42). Gas transfer was also reduced, consistent with smoking and neurofibromatosis related lung pathology, and a computed tomographic (CT) scan of the lungs showed patchy emphysematous changes, apical cystic disease, and basal fibrosis (carbon monoxide transfer factor (TLCO) 7.91 mmol/min/kPa (predicted 10.2 mmol/min/kPa); alveolar volume (VA) 6.63 l (predicted 5.47 l); carbon monoxide transfer coefficient (KCO) 1.19 mmol/min/kPa/l (predicted 2.03 mmol/min/kPa/l)). A CT scan of the superior mediastinum showed tracheal intramural thickening but there was no other localised

lesion in the mediastinum or lung. Fiberoptic bronchoscopy revealed tracheal narrowing without endobronchial abnormality and no malignant cells were found in bronchial aspirates.

Rigid bronchoscopy was performed. The extrathoracic and intrathoracic trachea was narrowed from 2.5 cm below the larynx to the carina with luminal narrowing from both lateral aspects. The lumen narrowed to 20% of normal in the intrathoracic region and here there was evidence of widespread intramural thickening. The overlying mucosa appeared thick and baggy. Normal posterior wall movement contributed to airway obstruction on expiration. Biopsy samples were taken. The carina, main bronchi, lobar and segmental anatomy was normal.

Microscopic examination showed normal epithelium but diffuse replacement of the mucosa by cytologically benign wavy spindle cells which enveloped but did not destroy the adnexal structures. These cells were S-100 positive, confirming the presence of a neurofibroma, consistent with neurofibromatosis.

The extent of the airway involvement precluded resectional treatment and so it was decided to stent the tracheal obstruction. An expandable covered metal wallstent was deployed in the trachea under radiographic and endoscopic control. Subsequent migration meant that this stent had to be replaced and an uncovered stent was inserted with a good clinical result.

Dyspnoea recurred at 5 months. Rigid bronchoscopy at this time revealed the appearance of the stent to be satisfactory but there was severe proximal tracheal narrowing. A second expandable uncovered metal wallstent was therefore positioned in this region with improvement of symptoms, exercise tolerance, and spirometric values. The patient was well for 2 years although routine bronchoscopies showed progressive tracheal narrowing both proximal and distal to the two stents. Eventually the patient returned with persistent wheezing. Rigid bronchoscopy allowed the insertion of two more short expandable uncovered metal wallstents (fig 1), one just below the larynx and one at the distal end of the trachea providing immediate clinical improvement. Further biopsy specimens showed persistence of neurofibromatosis in the tracheal wall.

Departments of Anaesthesia, Surgery and Histopathology, Royal Brompton & Harefield NHS Trust, London SW3 6NP, UK
J H Cranshaw
C Morgan
A G Nicholson
P Goldstraw

Department of Respiratory Medicine, Kingston Hospital, London SW3 6NP, UK
G Knowles

Correspondence to:
Mr P Goldstraw
p.goldstraw@rbh.nthames.nhs.uk

Received 17 February 2000
Returned to authors
17 May 2000
Revised version received
23 January 2001
Accepted for publication
14 February 2001

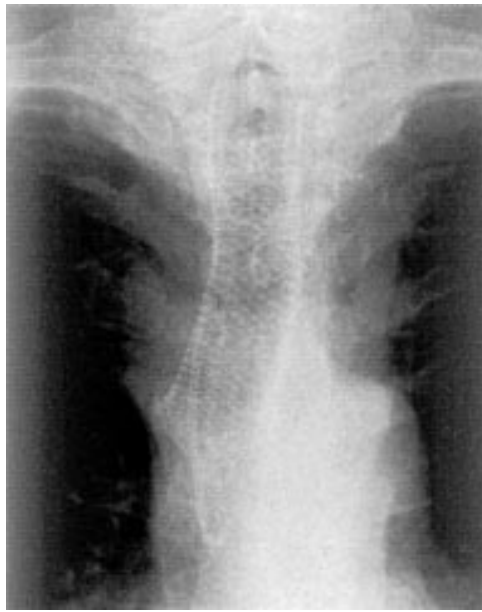


Figure 1 Chest radiograph after insertion of a total of four intratracheal stents.

The patient has now been followed up since her first stent insertion in 1995. At the present time she remains well and is reviewed three monthly with check bronchoscopic examinations.

Discussion

Von Recklinghausen's neurofibromatosis involves the thorax in 10–20% of cases. In patients over 35 years of age pulmonary manifestations include bilateral basal interstitial fibrosis and apical bullae. Why these interstitial changes occur is unknown. The lungs may also be sites of primary or metastatic neurogenic tumours. Primary neurogenic tumours involving the upper

airways in neurofibromatosis are rare and the actual incidence remains unrecorded.¹ Upper airway obstruction by a neurofibroma of the tongue has been reported,² as has external tracheal compression by a mediastinal mass.³ Chalmers and Armstrong⁴ reported a paediatric case of tracheal compression by an extratracheal plexiform neurofibroma that resulted in stridor. Reports of intramural lesions are rarer. Endobronchial nodules possibly responsible for obstructive pneumonitis have been reported^{5,6} and diffuse submucosal neurofibromatous infiltration of the bronchus has been found in a patient with HIV related immunosuppression.⁷ However, we believe that this is the first English language report of an intramural neurofibroma necessitating surgical stenting to maintain airway patency.

Despite concern that expandable metal stents may fracture or erode the airway wall with time, our experience over a maximum of 10 years suggests that tumour or inflammatory tissue ingrowth is the only major complication of these stents in the long term. In this case, given the extensive nature of the disease and its unremitting and progressive nature, the use of what is normally a short term tool in the long term setting is justifiable.

- 1 Unger PD, Geller SA, Anderson PJ. Pulmonary lesions in a patient with neurofibromatosis. *Arch Pathol Lab Med* 1984;108:654–7.
- 2 Crozier WC. Upper airway obstruction in neurofibromatosis. *Anaesthesia* 1987;42:1209–11.
- 3 Aughenbaugh GL. Thoracic manifestations of neurocutaneous diseases. *Radiol Clin North Am* 1984;22:741–55.
- 4 Chalmers AH, Armstrong P. Plexiform mediastinal neurofibromas. A report of two cases. *Br J Radiol* 1977;50:215–7.
- 5 Bartlett JP, Adams WE. Solitary primary neurogenic tumor of the lung. *J Thorac Surg* 1946;15:251–60.
- 6 Madewell JE, Feigin DS. Benign tumors of the lung. *Roentgenology* 1977;12:175–86.
- 7 Gillissen A, Kotterba S, Rasche K, et al. A rare manifestation of von Recklinghausen neurofibromatosis: advanced neurofibromatous infiltration in lung of a HIV-positive patient. *Respiration* 1994;61:292–4.