The Journal of Laryngology and Otology
August 1998, Vol. **112,** pp. 788–789

Have we got the full picture?

R. DEMAJUMDAR, F.R.C.S., F.R.C.S.(OTO.)*, P. B. RAJESH, F.R.C.S. (C.THORACIC)†

Abstract
A 59-year-old man with long-standing chronic obstructive airways disease (COPD), became progressively dyspnoeic, and repeatedly blacked-out during forced expiration. Spirometry suggested the possibility of large airways obstruction, and failing to respond to aggressive bronchodilator and steroid therapy, the patient was labelled as being non-compliant. Finally, he was assessed by an otolaryngologist and a cause for upper airway obstruction was suspected. Bronchoscopy and computed tomography (CT) scanning demonstrated tracheomalacia and the patient underwent resection of this segment of abnormal trachea.

Tracheomalacia, although rare, results from the substitution of cartilage with fibrous tissue, leading to severe airway compromise.

This case emphasizes the fact that although many conditions are uncommon, the total incidence of rare conditions is surprisingly high, and that care needs to be taken at all times in the management of 'labelled' patients with chronic illness, in order not to overlook such life-threatening diagnoses.

Key words: Lung diseases, obstructive; Respiratory sounds; Tracheomalacia

Case report

A 59-year-old Caucasian male with a 10-year history of chronic obstructive airways disease (COPD) and angina, was under the care of a respiratory physician and a cardiologist. He was repeatedly admitted to hospital due to exacerbations of dyspnoea, and during the last four years his condition had become severe. The patient blacked-out whilst attempting forced expiration, and also complained of a choking sensation whilst lying in the right lateral position. Investigations had excluded congestive cardiac failure although spirometry on three separate occasions had suggested large airway obstruction, and the patient was treated with steroids, nebulizers and domiciliary oxygen. Unfortunately, his condition did not respond to the treatment, and this was attributed to poor compliance. During his numerous visits to the outpatient department, these atypical clinical features were never probed, and the patient was left dyspnoeic at rest and unable to carry out his normal activities.

Finally, one episode of severe dyspnoea resulted in the patient's transfer to the Accident and Emergency (A&E) department. The A&E doctor who examined him became suspicious of an upper airway problem, although stridor was absent, and referred the patient urgently for assessment by an otolaryngologist on-call. The otolaryngology registrar (RD) performed a fibre-optic flexible endoscopy on the patient under local anaesthesia and found the ENT system to be normal, with a normal mobile larynx. However, after further local anaesthesia, he was able to pass the flexible scope between the vocal folds to find an obstruction in the trachea. A soft-tissue neck radiograph was normal, and a plain chest radiograph showed changes consistent with COPD.

After advice from a senior colleague, the patient was referred to a consultant cardiothoracic surgeon (PBR) for bronchoscopy. A rigid bronchoscopy was performed with the patient under general anaesthetic, and surprisingly, the respiratory tract appeared to be normal down to the carina. A CT scan was performed (Figure 1) which showed a significant obstruction of the trachea, but no cause for extrinsic compression was visible. The bronchoscopy was repeated, with the patient breathing spontaneously, and this demonstrated collapse of the left tracheal wall leading to a near-total obstruction of the main airway, which varied with respiration.

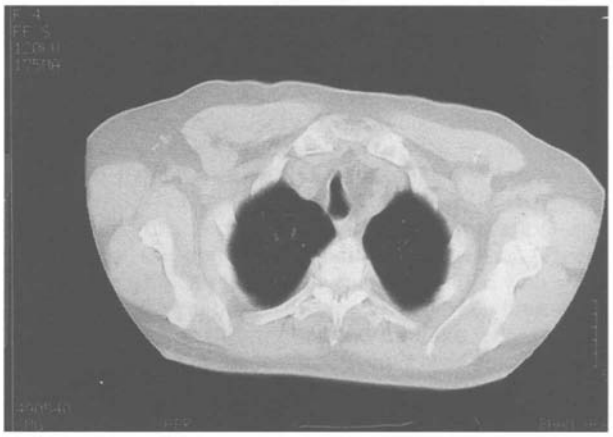

FIG. 1
Axial CT scan of neck demonstrating significant tracheal narrowing.

From the Department of Otolaryngology* and Cardiothoracic Surgery†, Heartlands Hospital, Birmingham, UK.
Accepted for publication: 24 May 1998.

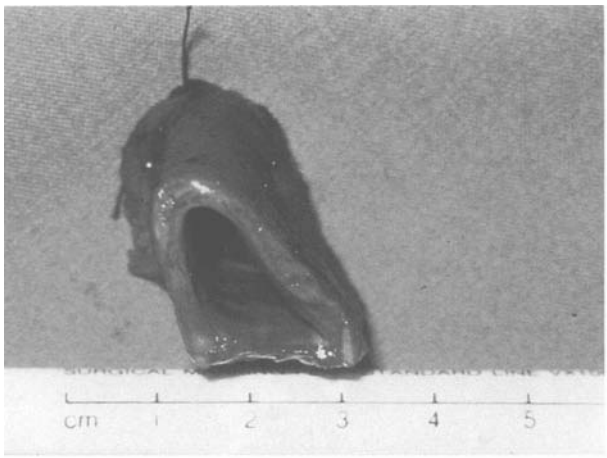

FIG. 2
Tracheal resection demonstrating obvious deformity, and abnormal cartilage.

This segment of soft, collapsible and narrowed trachea was only 2 cm in length, and a tracheal (Figure 2) resection with end-to-end anastomosis was carried out successfully. Histology of the specimen demonstrated that part of the cartilage had been replaced by hypocellular fibrous tissue which is typical of 'tracheomalacia'.

A significant improvement occurred in the patient's condition and his quality of life, leaving him with manageable COPD, and able to perform everyday tasks.

Discussion

Tracheomalacia is a serious condition, which can be either primary (rare) or secondary, which usually occurs due to endotracheal intubation, radiotherapy, trauma, or tumours (Hoskins *et al.*, 1991). There is weakness and fibrosis of the tracheal cartilage, making its walls collapse (Kang *et al.*, 1996) during expiration due to a rise in intrathoracic pressure (Stern *et al.*, 1993). Not only is there diminished ventilation, but also a Valsalva effect resulting in a reduction in venous return and thus precipitating a vaso-vagal syncope. None of the common causes of tracheomalacia were found in this patient, although there is an increased incidence amongst patients with chronic lung disease (Johnson *et al.*, 1973) such as COPD. This history from our patient suggested a far more complicated picture than severe COPD alone, and a large airway problem should have been suspected, particularly as this was suggested during spirometry.

Generally, stridor can be a key feature in tracheomalacia, although several cases have been described where stridor was either absent or occurred late (Duncan and Eid, 1991). Fluoroscopic examination can be useful in diagnosing tracheomalacia, but dynamic CT scanning is by far the investigation of choice (Feist *et al.*, 1975).

A major pitfall in the management of patients with chronic disease is the application of 'labels', and the follow-up of these patients is often delegated to junior staff. It is very easy during a busy clinic to disregard new symptoms, signs and investigation results that do not conform to the patient's diagnosis. We need to be vigilant at all times with chronically ill patients, and 'take a step back' to re-evaluate the situation at every consultation. As we are aware, rare conditions are rare but the overall prevalence of all rare conditions is surprisingly high. Every time a patient fails to respond to treatment we need to ask ourselves 'Is there something here that doesn't add up?' 'Have we got the full picture?' - We have now.

References

Duncan, S., Eid, N. (1991) Tracheomalacia and bronchopulmonary dysplasia. *Annals of Otology, Rhinology and Laryngology* **100(10):** 856–858.

Feist, J. H., Johnson, T. H., Wilson, R. J. (1975) Acquired tracheomalacia: aetiology and differential diagnosis. *Chest* **68(3):** 340–345.

Hoskins, M. C., Evans, R. A., King, S. J., Gishen, P. (1991) 'Sabre sheath' trachea with mediastinal lipomatosis mimicking a mediastinal tumour. *Clinical Radiology* **44(6):** 417–418.

Johnson, T. H., Mikita, J. J., Wilson, R. J., Feist, J. H. (1973) Acquired tracheomalacia. *Radiology* **109:** 577–580.

Kang, F. C., Tsai, Y. C., Jiang, C. Y., Chan, H. P., Chang, C. L. (1996) Acquired tracheomalacia - a case report. *Acta Anaesthesiologica Sinica* **34(4):** 239–242.

Stern, E. J., Graham, C. M., Webb, W. R., Gamsu, G. (1993) Normal trachea during forced expiration: Dynamic CT measurements. *Radiology* **187:** 27–31.

Address for correspondence:
R. Demajumdar,
9 The Green,
Oldbury,
Warley,
West Midlands B68 8DU.

https://doi.org/10.1017/S0022215100141714 Published online by Cambridge University Press