

the lower lobe. An operative diagnosis of bronchopulmonary sequestration was made and lobectomy completed.

Macroscopically the lobe measured  $9 \times 7 \times 7$  cm and on section showed a consolidated posterobasal segment with overlying pleural thickening. Multiple cysts measuring up to 2.5 cm in diameter were present in the consolidated segment (fig 2) and an aberrant vessel passed through the diaphragmatic surface of the abnormal area. Histological examination revealed cystic structures lined by cuboidal to pseudostratified columnar epithelium with smooth muscle fibres in the walls, and amorphous eosinophilic debris with foamy macrophages in their lumina. The interstitium was infiltrated by chronic inflammatory cells with lymphoid follicle formation. The pathological appearances were those of type II cystic adenomatoid malformation with numerous cysts arising in an intralobar bronchopulmonary sequestration.<sup>2-4</sup>

On retrospective review of the antenatal ultrasound examination no evidence was seen of the abnormal systemic arterial supply to this sequestered segment of lung.

## Discussion

To our knowledge intralobar bronchopulmonary sequestration in which an unexpanded portion of lung with no tracheobronchial connection and a systemic arterial supply has not hitherto been reported to be diagnosed prenatally.<sup>5,6</sup> This condition should now be included in the differential diagnosis of abnormal intrathoracic lesions detectable on antenatal ultrasound scan when a careful search should be made for an aberrant systemic blood supply.

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## Progressive tracheal and superior vena caval compression caused by benign neurofibromatosis

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### Abstract

The case history presented is of a patient with progressive tracheal and superior vena caval compression caused by a benign neurofibroma, a previously unrecognised feature of neurofibromatosis. The patient was successfully treated by surgical decompression.

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### Case report

A 21 year old woman, a known asthmatic for 11 years, was admitted to hospital with a four week history of progressive shortness of breath associated with dry cough and considerable stridor. She was initially treated with increas-

ing doses of bronchodilators and oral steroids, but continued to deteriorate. The patient's father was known to suffer from Von Recklinghausen's disease and no other member of the family was known to be affected.

On examination she looked unwell and was short of breath at rest. She had multiple cafe au lait spots and subcutaneous nodules, her face was puffy, and she had prominent subcutaneous veins on the top half of her body. Inspiratory stridor of moderate degree was noted.

A chest MRI scan (figure) showed a large



MRI scan of the chest showing a lobulated mediastinal mass (empty arrow) causing lower tracheal compression (solid arrow). © University Department of Medical Illustration.

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multilobulated mediastinal mass causing extrinsic tracheal compression and deviation to the left. Biopsies of the subcutaneous nodules confirmed the diagnosis of neurofibromatosis. Bronchoscopy showed evidence of extrinsic compression of the trachea with intraluminal bulging of the posterior wall; the endobronchial tree was otherwise normal. In view of the severity of superior vena caval obstruction mediastinoscopy was not performed.

At this stage palliative radiotherapy was contemplated, but surgical decompression was considered more appropriate and had the advantage of providing a tissue diagnosis. A right thoracotomy was therefore performed. The right lung looked healthy, a large mediastinal tumour was mobilised and excised, and the tumour extension into the neck was divided at the level of the dome of the pleura. The chest was closed in layers. On day 1 after surgery the patient's blood gases deteriorated and she had to be mechanically ventilated for 48 hours. Her postoperative recovery was otherwise uneventful.

Microscopic examination showed that the tumour was composed of spindle cells in an abundant fibrous stroma and showed focal myxoid changes; the cells were positive for S-100 protein. In areas there was a moderate degree of nuclear pleomorphism and occasional multinucleated tumour cells were seen. No mitotic figures were identified in multiple blocks. The overall impression was that the tumour was hypercellular and suggested a neurofibroma in an active growth phase and no features of malignancy.

Since mediastinal neurofibromas occurring in patients with diffuse neurofibromatosis are at an increased risk of neoplastic changes, and since tumour resection was incomplete, the mediastinum was irradiated with 3000 cGy in 16 fractions over 21 days. The patient remained well and asymptomatic 12 months postoperatively.

### Discussion

Progressive (over a few weeks) tracheal and superior vena caval obstruction in a previously

healthy individual is usually caused by a malignant neoplasm.<sup>1</sup> Empirical palliative radiotherapy and chemotherapy is occasionally given to patients without obtaining a histological diagnosis.<sup>2</sup> Attempts to prove the clinical suspicion of neoplasia after such treatment may be impossible.<sup>3</sup> Furthermore, in certain benign conditions such as neurofibromatosis, which affect up to one million individuals worldwide, the risk of malignant change increases by exposure to high doses of radiation.<sup>4</sup> The review by Ahmann<sup>5</sup> of data from over 90 publications, including 1980 reported cases of superior vena caval syndrome, showed that bronchogenic carcinoma was responsible for 85% of the cases. The commonest benign causes for superior vena caval obstruction were fibrosing mediastinitis and granulomatous disease caused by fungal infections (histoplasmosis). Intravenous catheters and pacing electrodes are becoming more frequent causes of superior vena caval obstructions. It is therefore vital to strive for a histological diagnosis in such cases unless there is a very compelling reason why this should not be done.

Modern anaesthetics and intensive care facilities have made thoracic procedures safer in these critically ill patients.<sup>6,7</sup> If mediastinoscopy is considered unsafe,<sup>8</sup> thoracotomy may prove to be helpful from both diagnostic and therapeutic points of view.

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