

# Long term survival after treatment of disseminated T cell lymphoma presenting with tracheal obstruction in a patient with coeliac disease

D J SEDDON, K F CHUNG, F J PARADINAS, E S NEWLANDS, P D SNASHALL

*From the Departments of Medicine, Pathology, and Medical Oncology, Charing Cross Hospital, London*

**ABSTRACT** A 46 year old man with coeliac disease and upper airway obstruction was thought to have undifferentiated carcinoma of the trachea. Subsequent immunohistological examination showed that the neoplasm was a T cell lymphoma. He had combination chemotherapy and is alive and well seven years after diagnosis.

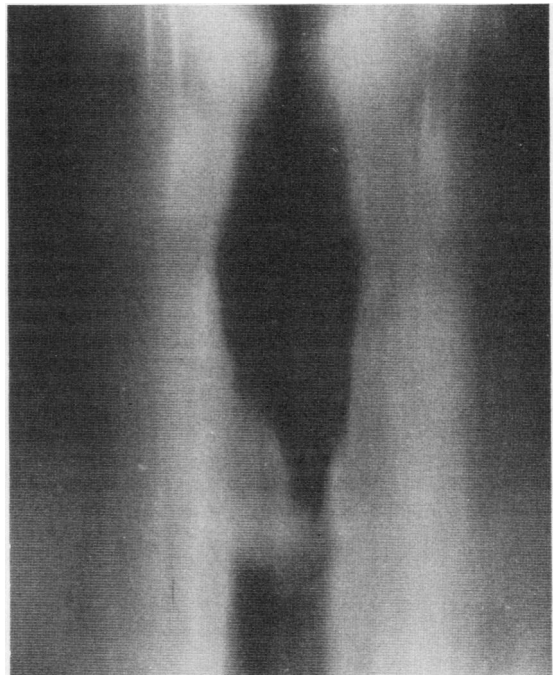
Primary neoplasms of the trachea are rare, presenting usually with obstruction of the upper airway. Both carcinoma and lymphoma may occur at this site. With modern treatment disseminated lymphoma has a more favourable prognosis than disseminated carcinoma and is therefore an important histological diagnosis.<sup>1</sup>

In patients with coeliac disease the most usual histological type of lymphoma is of T cell lineage. This presents most commonly with tumour masses in the small bowel, deposits elsewhere being a late and inconstant finding. Cure occurs in some cases.<sup>2,3</sup> We present a case of T cell lymphoma causing tracheal obstruction in a patient with coeliac disease and dermatitis herpetiformis.

## Case report

A 46 year old man was referred with a diagnosis of asthma not responding to treatment. He gave a history of shortness of breath for one month, which he described as difficulty in "getting the air into my chest." This symptom was present at rest and was worse during exercise. He had a one week history of unproductive cough and hoarseness without haemoptysis. There was no disturbance of sleep pattern. Although his symptoms were unaffected by posture, the patient had noticed stridor on extreme neck flexion. He had never smoked. When he was a young adult coeliac disease (from jejunal biopsy) and dermatitis herpetiformis (from skin biopsy) had been diagnosed. His treatment consisted of a gluten free diet and dapsone 100 mg daily. Physical examination indicated normal breath sounds over both lung fields, with harsh inspiratory and expiratory wheezing over the central airways. There was no cervical lymphadenopathy.

A chest radiograph was normal and direct laryngoscopy showed no supraglottic tumour. Barium studies of the stomach and small bowel showed no abnormality; jejunal biopsy was not repeated. Lung function testing showed FEV<sub>1</sub>



*Tomogram showing tumour arising from the right lateral tracheal wall.*

to be 3.65 litres (BTPS) and FVC 5.6 l (both within the predicted range of normal values). Peak flow (PEF) was reduced to 330 (normal 590) l/min. Total lung capacity, residual volume, and functional residual capacity were normal. Flow-volume loops showed a plateau in both expiration and inspiration. Laryngeal tomograms showed the presence of a tracheal tumour (figure) and this was confirmed at rigid bronchoscopy, which showed an irregular neoplasm 4 cm below the vocal cords. The tumour mass arose from the right tracheal wall and extended posteriorly. The length of the stricture was about 4 cm with a 5 mm lumen. A histological diagnosis of undifferentiated carcinoma was made from the tumour biopsy specimen.

After a course of radiotherapy to the trachea the obstruction of the upper airway improved considerably. Five months later the patient presented with a painful left elbow and shoulder. Radiographs of the left ulna showed a lytic lesion at its proximal end and an isotopic bone scan showed increased radionuclide uptake at this site and in the left acromion,

Address for reprint requests: Dr D J Seddon, Department of Medicine, Charing Cross Hospital, London W6 8RF

Accepted 22 March 1989

cervical spine, and skull. Bone marrow aspiration and trephine of the left iliac crest were performed.

The bone marrow smear was unremarkable, but biopsy of the left ulna showed marrow replaced by tumour. Abundant plasma cells, eosinophils, and non-neoplastic histiocytes were also present. Immunohistochemical examination of both the tracheal tumour and the marrow tumour showed  $\alpha_1$  antitrypsin,  $\alpha_1$  chymotrypsin, and muramidase in the tumour cell cytoplasm. Stains for leucocyte common antigen and the T cell markers MT-1 and UCHL-1 were positive. The B cell markers MB-1 and MB-2 were not detected. Kappa and lambda light chain stains showed the presence of polyclonal plasma cells. These findings indicated that the neoplasm was in fact a T cell lymphoma similar cytologically to the small bowel lymphoma seen in association with coeliac disease.

The patient was treated with M-BACOP (intravenous bleomycin 10 mg/m<sup>2</sup>, doxorubicin 45 mg/m<sup>2</sup>, and cyclophosphamide 400 mg/m<sup>2</sup> on day 1 with prednisolone 100 mg orally for five days) followed by intravenous methotrexate 200 mg/m<sup>2</sup> on day 8 with folinic acid rescue. Courses were repeated in a four week cycle, with a total of eight courses. He tolerated the chemotherapy well and repeat biopsy of the left ulna at the end of treatment showed no evidence of tumour. He remains well seven years later.

### Discussion

Primary lymphoma of the trachea is an exceedingly rare tumour,<sup>4</sup> and so far as we are aware this is the first known instance of coeliac disease associated T cell lymphoma presenting at this site.

This case underlines two important points. The first is the importance of considering upper airway obstruction in the diagnosis of "asthma," particularly if the airways obstruc-

tion is of recent onset in a middle aged or elderly patient, and if it is not reversed by administration of bronchodilating drugs. Examination of flow-volume loops will usually distinguish between small airways obstruction and the characteristic flattening of expiratory and inspiratory limbs of the loop in large extrathoracic airways.

Secondly, this case illustrates the importance of the diagnosis of lymphoma as opposed to carcinoma. Modern immunohistochemical techniques help considerably in establishing such a diagnosis. In this case the presence of T cell antigens on the surface of the tumour cells confirmed the diagnosis of T cell lymphoma. In a series of 120 cases of malignant tumours causing diagnostic difficulties and examined with immunohistological techniques lymphoma was found to be the most common diagnosis (66%) and accounted for 29 of the 43 cases thought initially to be anaplastic carcinoma.<sup>5</sup> Such techniques should be applied whenever there is difficulty in establishing the precise histological diagnosis of a malignant tumour.

### References

- 1 Mead GM, Whitehouse JMA. Modern management of non-Hodgkin's lymphoma. *Br Med J* 1986;293:577-80.
- 2 Isaacson PG, O'Connor NTJ, Spencer J, *et al.* Malignant histiocytosis of the intestine: a T-cell lymphoma. *Lancet* 1985; 01ii:688-91.
- 3 Tseng A, Coleman N, Cox RS. The treatment of malignant histiocytosis. *Blood* 1984;64:48-53.
- 4 Wiggins J, Sheffield E, Green M. Primary B-cell malignant lymphoma of the trachea. *Thorax* 1988;43:497-8.
- 5 Gatter KC, Alcock C, Heryet Y, Mason DY. Clinical importance of analysing malignant tumours of uncertain origin with immunohistological techniques. *Lancet* 1985;ii:1302-5.