Lethal tracheal dissolution during treatment for thyroid lymphoma

Figure 2 Portable chest radiograph after intubation showing extensive subcutaneous emphysema of the upper chest and neck. The cervical trachea is markedly distended (arrows).

mon: hoarseness (9–67%), dysphagia (9–60%), and dyspnoea or stridor (9–35%). Although the optimal treatment for thyroid lymphoma is uncertain, patients with stage IIE or more advanced disease should probably receive chemotherapy in addition to local treatment (surgery and/or radiotherapy).6 Perforation of the gastrointestinal tract at the site of disease in patients with gastrointestinal lymphoma who are receiving chemotherapy has been well described;1 a similar perforation has been reported in a patient with thyroid lymphoma metastatic to the small bowel.7 Lethal tracheal perforation during chemotherapy has not previously been described in a patient with primary thyroid lymphoma. Most series of patients with thyroid lymphoma fail to mention whether the tumour invaded the trachea or larynx.8 In one report two patients with needle biopsy performed before surgery and fine needle biopsy performed after surgery were missed unless a careful examination of the trachea was performed.9

Computed tomographic scanning is the recommended method of assessment of extrathyroid extension of lymphoma.9 Early awareness of tracheal involvement by lymphoma should alert the clinician to the remote possibility of tracheal dissolution during treatment of this extremely difficult clinical problem.

Commentary: lymphoma involving the mediastinum – challenges in diagnosis and management

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Lymphomas are the seventh most common malignancy and the incidence of non-Hodgkin's lymphoma is increasing. Correct diagnosis is important as they are treatable and, in some cases, curable. Lymphomas can involve any organ system and, as thoracic involvement is common, they often present to respiratory physicians. The diagnosis is not always straightforward and treatment is complex. The case reports in this issue of Thorax illustrate some of the diagnostic and therapeutic problems associated with this disease. Mediastinal tumours in adults are divided fairly equally throughout the mediastinal compartments. Approximately 40% of tumours are found in the anterosuperior compartment. Thymomas are the most common tumour in this region and lymphomas and carcinoids may involve the thymus. Mediastinal germ cell tumours are located in the anterosuperior compartment and thyroid tumours may involve the upper part of the superior mediastinum. Secondary tumours, particularly bronchogenic carcinoma, should also always be considered. Thymomas are often suspected by their radiological appearance and they may have associated systemic features, most notably myasthenia gravis. They are usually treated by surgical extirpation which allows further detailed study of the pathology and avoids the increased incidence of local recurrence seen if needle biopsy is performed before surgery. Fine needle aspiration is simple and may be sufficient to diagnose a carcinoma. However, as Robinson et al point out, the diagnosis of lymphoma may be missed unless adequate tissue is removed. This is required for study of the morphology of the tumour which provides prognostic information and assists management. A comprehensive immunohistological examination should be performed in case misleading information is obtained from one immuno-
histochemical method. Some “anaplastic carcinomas” turn out to be high grade non-Hodgkin’s lymphomas which are potentially curable.1 Tumour-specific chromosomal abnormalities have been identified in several types of lymphoma, germ cell tumours, Ewing’s sarcoma, and peripheral neuroepithelioma. The diagnostic usefulness of this technique is likely to increase as further abnormalities are identified. Raised serum tumour markers such as α-fetoprotein and human chorionic gonadotrophin may help to support a histological diagnosis of a mediastinal germ cell tumour. Rarer tumours such as the “atypical teratoma syndrome” should be considered if a rapidly growing undifferentiated carcinoma is found in a predominantly midline distribution. This tumour, which occurs particularly in young men, often responds well to intensive cisplatin based chemotherapy.2

Most cases of Hodgkin’s disease that occur in the mediastinum are of the nodular sclerosing type. The choice between primary chemotherapy and radiation is usually made on the basis of tumour stage and bulk of the disease. A good clinical and radiological response to treatment occurs in most patients, but radiological abnormalities may persist with widening of the mediastinum and architectural distortion of the lymph node areas. It is often impossible to know whether there is persistent active Hodgkin’s disease. This creates a dilemma as consolidation treatment with radiotherapy may be required but it increases the chance of long term toxicity. An example of this problem is presented in the report by Thomas et al. BCNU (carmustine) is one of a group of chemically unrelated drugs (others include bleomycin, busulfan, methotrexate, cyclophosphamide in high doses, and mitomycin C) known for their damaging effects on lung. The probability of pulmonary injury increases when radiotherapy is given to the lung, either before or after cytotoxic chemotherapy.3 Dry cough and dyspnoea are the characteristic symptoms of acute pneumonitis due to cytotoxic drugs or radiation. Drug-related damage often causes changes in the basal respiratory segments with linear and nodular shadowing which may become confluent on the chest radiograph. Severe respiratory insufficiency does sometimes occur, but prompt treatment with high dose steroids often reverses the process. Steroids are less effective in chronic progressive drug-induced fibrosis. The decision to consolidate treatment with radiation therapy is usually made on clinical grounds. A biopsy sample of residual tissue after treatment is subject to sampling error; additional imaging by gallium or positron emission tomographic scanning is only sometimes helpful in distinguishing tumour from residual non-malignant tissue. The risk of local relapse is much greater when a lymphoma occupies more than one third of the width of the mediastinum although it can be reduced by radiotherapy. However, the addition of radiotherapy has not been shown to improve overall survival in this group of patients. About half the cases of Hodgkin’s disease have mediastinal involvement which may include the thymus. Cysts in the thymus such as those described by El-Sharkawi and Patel may be found when this tumour is part of the extranodal component with Hodgkin’s disease. Whether Hodgkin’s disease originates in the thymus in these cases is not clear, but management is similar to mediastinal Hodgkin’s disease.

A significant number of non-Hodgkin’s lymphomas arise in extranodal sites. Their clinical behaviour is related more to their biology than to their anatomical location. Thyroid lymphomas are rare and account for about 5% of thyroid malignancies. They are usually associated with Hashimoto’s thyroiditis and occur most commonly in women. Many are low grade tumours similar to MALT lymphoma found in the gastrointestinal tract.4 Others are intermediate or high grade B cell tumours. Although they are often confined to the thyroid and cervical lymph nodes at presentation, tumours with aggressive type histology may disseminate widely.5 T cell lymphomas of the thyroid are rare so that other sites of origin should be considered, particularly as the patient described by Melnyk et al had axillary lymphadenopathy and skin involvement. The tumour in the patient they described had many characteristics similar to those seen in peripheral T cell lymphomas of the angiocentric type. These are aggressive tumours, associated with necrosis, and can involve the mediastinum and lung. Tumours of the nasopharynx were formerly and appropriately called “lethal midline granuloma”. They behave as a locally destructive inflammatory tumour and evolve into a generalised T cell lung lymphoma. They are known to produce a coagulative necrosis and the case described by Melnyk et al probably falls into this category. The destructive lesion they described is dissimilar from the more widely recognised “tumour lysis syndrome”, occasionally seen shortly after the start of chemotherapy for leukaemia and lymphomas. Rapid dissociation of tumour cells may cause a gross metabolic disturbance such as lactic acidosis, hyperkalaemia, hyperphosphataemia, and hyperuricaemic renal failure. These complications can often be avoided by careful preparation of the patient before chemotherapy, ensuring that there is adequate hydration and inhibition of uric acid synthesis. In solid tumours, such as germ cell malignancies or gestational trophoblastic tumours, rapid cytolysis of extensive pre-existing pulmonary metastases may lead to respiratory decompensation. It may be possible to lessen this effect by delaying chemotherapy at a lower than usual dose.

Correct diagnosis of mediastinal tumours is important and their investigation is best performed by a multidisciplinary specialist team of surgeons, pathologists, oncologists, and radiologists. Lymphoma and Hodgkin’s disease are curable in some patients and treatment in others results in prolonged survival. However, the complications of treatment and emergence of drug resistance illustrate some of the frustrations of treatment.

LETTERS TO THE EDITOR

Guidelines for care during bronchoscopy

Guidelines for care during bronchoscopy were agreed by the British Thoracic Society in January 1993. A retrospective audit of adherence to these guidelines in Scotland was performed for 23 hospitals by means of a written questionnaire and the results were presented at a recent meeting of the Scottish Thoracic Society.

Of the 33 respiratory consultants who replied to our survey, 31 routinely applied pulse oximetry during bronchoscopy. 31 wore gloves, but only eight wore a gown, four a mask, and three gowns during the procedure, despite recommendations that mask, gown, gloves, and close fitting eye protection should be worn in all cases. Resuscitation equipment was available in the bronchoscopy suite in 31 cases, and in all but one case a nurse or second doctor was present during the procedure. Twenty-eight consultants reported that their bronchoscopy nurses adhered to the policy of the BTS Working Party on Infection Control. In 30 of the 33 replies ECG monitoring was available for patients with known cardiac problems, and in all cases antidotes were available for potentially respiratory depressant drugs. In general, adherence to the BTS guidelines on care during bronchoscopy is satisfactory, but in view of non-compliance with the wearing of masks, gown and gowns, these components need to be re-addressed.

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Apnoea in Duchenne muscular dystrophy

Drs Khan and Heckmann (February 1994;49:157-61) concluded that "sleep related breathing abnormality in Duchenne muscular dystrophy is initially obstructive...". We have recently performed a night time study of six patients with Duchenne muscular dystrophy and reached somewhat different conclusions.1 Like them, we also observed predominantly obstructive apnoeas in some but not all patients (two out of six), both were severe, frequent and had macroglossia. Four patients had central apnoeas only. This predominance of central events would agree with the data published several years ago by Smith et al. However, as correction and analysis were not performed in these patients, the smaller events at night were of central origin.1 We therefore believe that the conclusion of Khan and Heckmann, that most of the apnoeas in patients with Duchenne muscular dystrophy were obstructive, because it has been validated using appropriate methodology. Their conclusion is also weakened by the fact that the sleep studies were carried out at home in a non-researched environment in a semi-automatic way. This approach would be a valuable one in screening or epidemiological studies but it is probably not appropriate for pathophysiological research. To obtain accurate and reproducible data on such studies to be performed under supervision and to be manually scored.1 It is important to clarify the nature of the sleep respiratory events in patients with Duchenne muscular dystrophy because it may have therapeutic implications. If the most predominant respiratory event is obstructive apnoea, nasal continuous positive airway pressure (CPAP) should be effective. CPAP is cheaper and easier to use than intermittent positive ventilation, which nowadays constitutes the treatment of choice for patients with Duchenne muscular dystrophy and respiratory failure.

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Chest physiotherapy in cystic fibrosis

I am concerned that the paper by Miller et al (Thorax 1995;50:10:165) is not a valid comparison of autogenic drainage and the active cycle of breathing techniques (ACBT) as stated.

ACBT is a method of physiotherapy which includes thoracic expansion exercise, breathing control, and the forced expiration technique (FET). The FET is an important component combining forced expiration (huffing) and breathing control. Several huffs to low lung volume immediately after breathing control are often needed before secretions are mobilised from the smaller peripheral airways to the larger airways.1,2 It is only when the secretions have reached the larger proximal airways that the huff from high lung volume is required. The forced expiration technique described in the paper of Miller et al is similar to that above, but there is very good reason to believe that the method performed by the patients was "one huff from mid to low lung volume followed by another huff at a higher lung volume." Whilst this difference may appear subtle, to a physiotherapist and patient it is a major difference in technique and does not correspond with the technique previously described in the literature.3

The statement that the two treatment regimens used in this study were equally as good is not in dispute, but the claim that one of the regimens was ACBT is cause for concern. If my belief is correct, this study was not a true comparison between ACBT and autogenic drainage. The results are likely to mislead medical practitioners, physiotherapists, and patients and could appropriately influence the direction of future research. Further studies are required to provide a valid comparison between these two techniques.

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AUTHORS’ REPLY

In the first draft of the paper the description of ACBT differed from the published version. The forced expiratory technique was described as "one huff from mid to low lung volume, followed by another huff at a higher lung volume". We were encouraged to cough and expectorate only if