improvement in his pulmonary physiology; FEV₁ 1.96 l and FVC 3.18 l recorded.

Discussion

The first descriptions of disease currently recognized as sarcoidosis were documented at the end of the 19th century and concerned skin eruptions (1). Since that time, it has been realized that it is a systemic disease characterized histologically by non-caseating granulomata. An aetiologi- cal agent eludes characterization. A number of recognizable patterns of disease at presentation are documented including Lofgren's syndrome (2). Lymph node enlargement is frequently found in sarcoidosis, occurring in up to 90% of patients during the course of their disease. The most common pattern of mediastinal adenopathy is bilateral involvement of the bronchopulmonary, right paratracheal and aortopulmonary nodes, but any combination can occur (3). Despite this, the incidence of obstruction of the intrathoracic vasculature is low. To the authors' knowledge, only four reports of superior vena cava (4-7) and one of innominate vena (8) obstruction due to sarcoid are described in the literature. No cases of subclavian vein obstruction have been reported previously. Obstruction of vessels can be caused by two mechanisms: involvement of the vessel wall with granulomata or extrinsic compression by enlarged lymph nodes. In the present case, the obstruction has been demonstrated radiographically (Plate 1) to be due to external compression by intrathoracic lymphadenopathy; however, concurrent microscopic involvement of the vessel wall could not be excluded.

The diagnosis of sarcoidosis in this case is supported by many factors: the initial presentation of uveitis and lymphadenopathy, abnormal pulmonary physiology which improved with oral corticosteroids, the positive Kveim, the intrathoracic biopsy material obtained, and the raised serum ACE during the current recrudescence. The subsequent course of the illness and response to therapy would not be typical for any other granulomatous disease.

To the authors' knowledge, this case is the first to present on two separate occasions with intra-thoracic venous obstruction, the second with an increase in activity of disease. At present, the symptoms have resolved with anticoagulation and corticosteroid therapy.

References

3. Rockoff SD, Rohatgi PK. Unusual manifestations of thoracic sarcoidosis AJR 1985; 144: 513-528.

Bilateral endobronchial metastases due to a chondroblastic osteosarcoma

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Introduction

Endobronchial metastases from non-pulmonary neoplasms are rare. When they occur, the primary site of the tumour is most commonly breast, colon or kidney (1), although other sites have been reported, including sarcoma (2). The present case report describes a case in which bilateral metastases secondary to a chondroblastic osteosarcoma of the spine presented acutely with dyspnoea, and removal of these lesions resulted in great symptomatic improvement. However, the patient's subsequent rapid deterioration and death highlights that the prognosis in such cases is poor.
PLATE 1. Histology of the primary lesion showing tumour osteoid production adjacent to hypercellular cartilage with mildly pleomorphic nuclei, some of which are binucleate. (Haematoxylin and eosin stain, original magnification × 200).

Case Report

A 30-year-old man initially presented in late 1994 with a 6-month history of back pain, 2 months of leg weakness and urinary incontinence for 4 days. Magnetic resonance imaging revealed an extradural tumour extending from the L3 to the S1 level involving the exit foramina and infiltrating the left lliac muscles. He underwent a decompressive laminectomy, and multiple biopsies revealed the tumour to be a chondroblastic osteosarcoma (Plate 1). This failed to respond to chemotherapy with cisplatinum and Adriamycin, so he was given second-line drugs using ifosfamide and etoposide, as well as concurrent radiotherapy. This resulted in an incomplete response. In August 1995, he represented with bladder dysfunction and foot drop. This developed into complete paraplegia despite further radiotherapy.

Seven months later, he presented as an emergency with a 1-week history of haemoptysis and a 2-day history of dyspnoea and wheeze. Examination revealed a pale, dyspnoeic young man with stridor, a respiratory rate of 30 breaths min⁻¹, a medium-sized pleural effusion on the right lung and a large right hypochondriac mass. Investigations showed haemoglobin of 6.4 g dl⁻¹, and chest X-ray confirmed a medium-sized right-sided pleural effusion and also revealed two calcified masses in the left midzone. Peak flow was 160 l min⁻¹ and blood gas analysis showed a pH of 7.34, PO₂ 5.7 and PCO₂ 4.8, whilst breathing air.

His symptoms failed to settle completely with bronchodilators, steroids, blood transfusion and aspiration of his effusion. Fibre-optic bronchoscopy revealed bilateral pedunculated endobronchial masses; one in the right bronchus intermedius and one in the left main bronchus. These filled most of the lumen and moved with respiration.

He was referred for laser ablation of these lesions but, because of a sudden deterioration, he underwent an emergency rigid bronchoscopy, during which his oxygen saturation fell to 40%, and he needed rapid piecemeal removal of both lesions (Plate 2) with forceps. This resulted in good clearance and his oxygen saturation rose to 100%. Histology of the endobronchial lesions showed them to be metastases from the chondroblastic osteosarcoma. Following the procedure, his breathing improved greatly, his FEV₁ rose from 0.5 to 2.15 l post procedure and he was able to be discharged home.

Five weeks later, he was re-admitted with a generalized deterioration but no clinical or physiological evidence of an endobronchial recurrence. He died peacefully 4 weeks later.

Discussion

Endobronchial metastases occur in only 2% of subjects with pulmonary metastatic disease (3,4). Breast, colon and renal primary tumours are responsible for about 80% of these, whilst other reported primary sites include nasopharynx, bladder, thyroid, ovary and prostate (1). Endobronchial metastases arising from sarcoma have only rarely been reported before, and the authors are unaware of any previous reports where chondroblastic osteosarcoma has presented in this way. Bilateral synchronous endobronchial metastases have been reported occasionally in large series (1,5), but a literature review failed to identify any caused by sarcoma (2).

The presence of endobronchial metastases has usually been taken to denote a poor prognosis as widespread extrabronchial metastases are also usually present. This was certainly the case here, where there was evidence of both pleural and abdominal spread and where, in spite of excellent endobronchial clearance, there was a rapid
deterioration to death in less than 3 months. However, with obvious radiological evidence of both intrapulmonary and pleural disease, it was important to avoid the pitfall of ascribing his respiratory symptoms to these, and failing to consider endobronchial disease. Stridor in this context should always prompt a search for a lesion in the main airway, as the advent of newer endobronchial therapies, such as stenting, brachytherapy or laser (6), may allow useful symptom palliation.

Some reports suggest better outcomes in patients treated for endobronchial metastases. The report of three patients with endobronchial metastases from sarcoma showed a moderate survival, with one patient dying 5 months after the discovery of endobronchial metastases and the other two still being alive 11 and 14 months after this finding. The present case is in stark contrast to this. The poorer outcome in this case reflects not only the extensive nature of this patient's extrabronchial disease, but also that he had previously failed to respond markedly to aggressive chemotherapy and radiotherapy.

Response of dermatomyositis co-existing with non-small cell lung cancer to chemotherapy

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Introduction

Non-small cell lung cancer (NSCLC) is a common cause of death world-wide and is less responsive to chemotherapy than small cell lung cancer. Dermatomyositis is a rare condition in Chinese and is classically associated with underlying malignancy, although this association has been recently doubted by some authors (1,2). Association of dermatomyositis with lung cancer is less doubtful, although a causal relationship has not been firmly established (3). Whilst it is known that cancer-associated dermatomyositis is often more refractory to standard treatment such as the administration of systemic corticosteroids, little is known of the response of NSCLC-associated dermatomyositis to chemotherapy directed at the underlying lung cancer. The present case report describes a case of a 58-year old man who presented with dermatomyositis and was found to have extensive metastatic NSCLC. Both the NSCLC and dermatomyositis responded well to a combination of mitomycin-C (M), ifosphamide (I) and cis-platinum (P).

Case History

A 58-year old ex-smoker presented with a 2-month history of an erythematous and pruritic rash affecting his face and limbs which was resistant to topical steroid therapy (Plate 1). Deterioration and ulceration of the skin was accompanied by development of progressively disabling symmetrical limb girdle weakness (including inability to stand from sitting position and flex the neck), swallowing difficulty and weight loss, although there were no respiratory symptoms. Physical examination revealed an erythematous rash affecting the described areas, symmetrical limb girdle weakness (4/5) and nasal regurgitation, but there was no other abnormalities.

A chest X-ray showed a right hilar shadow which was revealed on thoracic computerized tomography (CT) (Plate 2) to be a 3 cm irregular mass situated anterior to the

References