Diffuse malignant mesothelioma presenting as bilateral malignant lymphangitis

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Malignant mesothelioma commonly presents a radiological picture of unilateral irregular pleural thickening. When accompanied by a pleural effusion, as is often the case, the underlying tumour may be obscured. Less commonly, single or a few rounded opacities may be present in the underlying lung.1–4 Although microscopic lymphatic invasion and spread to both regional lymph node groups and underlying lung is well described,5 mesothelioma presenting as malignant lymphangitis with a clear clinical and radiological picture is very rare and has been described only with the contralateral lung affected.6 In this case of bilateral malignant lymphangitis complicating diffuse malignant mesothelioma a negative radiograph taken shortly before the diagnostic radiograph proved a useful pointer to the correct diagnosis.

Case report

In October 1982 a 40 year old motor company secretary presented with a history of left sided pleuritic chest pain that followed a severe attack of influenza 15 months previously.

On examination he was noted to have a mildly flattened left chest with compensatory scoliosis. No other clinical abnormalities were detected. Radiographic screening showed partial fixation of the left diaphragm. The chest radiograph showed mild linear atelectasis in the left lower zone. A minor degree of pleural shadowing was present at the left base, which extended up the lateral chest wall. This picture suggested either pleural nodules or intrapulmonary lesions. Bronchoscopy and bronchography were unhelpful. Cytological examination of the sputum yielded negative results. At that time the symptoms and signs were thought to be postinflammatory and the patient was treated symptomatically.

Six months later, in May 1983, he returned complaining of considerable abdominal discomfort, right sided chest pain, and severe dyspnoea. He appeared ill, and was febrile (38°C), and there was evidence of recent weight loss. He had widespread wheezes and prominent basal crackles on auscultation of the chest. He had no abdominal masses or lymphadenopathy. The chest radiograph showed widespread coarse abnormal shadowing suggestive of interstitial infiltration, which was present bilaterally in all lung zones, affecting the bases more than the upper zones. A moderate right sided pleural effusion was also present (figure). Bronchoscopy showed no abnormality. A blood stained pleural effusion was drained and pleural biopsy was performed. Preliminary histological reports on the bronchial washings, pleural fluid, and pleural biopsy specimens indicated poorly differentiated malignant cells suggesting either an adenocarcinoma or a mesothelioma. His condition continued to deteriorate and he died one week later.

Necropsy findings

At necropsy both pleurae were macroscopically thickened by tumour, which obscured the outline of the thoracic aorta. Metastatic and continuous tumour deposits were present in both lungs. The regional lymph glands and both kidneys were infiltrated macroscopically by tumour. Microscopic examination of sections of the pleurae showed neoplastic infiltrations by sheets of cells typical of a mixed type of mesothelioma. In the lungs this cellular pattern was present in alveoli and in perivascular lymphatic channels. The kidney lesions were identical to those in the pleurae and lungs. A mesotheliomatous nodule was found macroscopically in the atrial wall. No malignant infiltrations were found in any other organs, and no evidence of asbestosis was present. An asbestos fibre count of pulmonary tissue revealed over 3 million asbestos fibres per gram of wet tissue and over 23 million asbestos fibres per gram of dry tissue. The diagnosis of diffuse malignant pleural mesothelioma of mixed histological type was confirmed unanimously by the five pathologists serving on the South African Asbestos Tumour Reference Panel.

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History of asbestos exposure
The patient was born and raised on his family’s farm in the North West Cape, South Africa, 10 kilometres from the Cape blue asbestos mine at Pomfret. The only time that he or any family member worked at the mine was when the patient, at the age of 21 years, was employed for a spell of three months in the milling section of the crocidolite mine. Thereafter he moved away from the area, returning only for short intermittent visits to the farm.

Discussion
In view of the absence of any intrapulmonary shadows on the first radiograph, the presence of the diffuse coarse bilateral interstitial shadows seen six months later could not have been attributed to asbestosis since advanced asbestosis is not likely to have developed with such speed. As the patient’s earliest exposure to asbestos had occurred 40 years previously, signs of asbestosis would have been expected on the initial radiograph. It is of interest that despite the high asbestos fibre concentrations found in the lungs no asbestosis was present histologically. The radiographic changes were attributable to malignant lymphangitis. To the best of our knowledge this is the first reported case of bilateral malignant lymphangitis complicating malignant mesothelioma. While malignant lymphangitis is known to occur as a microscopic phenomenon,5 we believe that its presence as a radiological and clinical entity has not previously been recognised.

References