Primary pleural liposarcoma

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Primary intrathoracic liposarcoma is uncommon. Razzak et al in 1971 indicated that 44 cases had been recorded, of which 43 originated in the mediastinum and one in the pulmonary hilum. Sawamura et al in 1982 reported seven cases of primary pulmonary liposarcoma. We report a further case, which displays features suggesting that it was of pleural rather than mediastinal or pulmonary origin.

Case report

A 61 year old man with a 30 year history of asthma and a two year history of ischaemic heart disease was admitted for investigation of a left sided pleural effusion. He gave a four week history of increasing dyspnoea on exertion associated with pain in the left side of the chest. He had never smoked and denied haemoptysis. On examination he had a cushingoid appearance as a result of long term steroid treatment for his asthma and had signs of a left pleural effusion. The haemoglobin concentration was 12.1 g/dl, the total white cell count 10.0 x 10^9/l, and erythrocyte sedimentation rate 120 mm in one hour. The plasma urea and electrolyte concentrations and results of liver function tests were normal. Examination of the sputum for tubercle bacilli, neoplastic cells, and asbestos bodies was negative. The chest radiograph showed evidence of cardiomegaly with a large left pleural effusion. A chest radiograph taken two months earlier had shown blunting of the left costophrenic angle. After admission pleural aspiration produced 500 ml of heavily blood stained fluid. An intercostal tube was inserted and thick jelly like material was obtained, which eventually blocked the tube. Two days after admission the patient developed intractable left ventricular failure and died.

Postmortem examination showed a recent myocardial infarct and thromboembolism in right and left pulmonary arteries. The greater part of the left lung was encased in soft mucinous tumour, which covered the pleural surface in a sheet varying from a few millimetres to 4.5 cm in thickness. The tumour was adherent to the visceral pleura and extended along the interlobar fissure and medial surface of the lung to affect part of the pericardium. It did not form a macroscopically evident mediastinal mass and it compressed the lung but did not invade the lung parenchyma. There were light friable adhesions to the parietal pleura but no evidence of tumour of the chest wall. Dissection of retroperitoneum and abdominal and pelvic viscera showed no evidence of neoplasm, neither did inspection and palpation of the extremities.

Microscopically the tumour consisted of somewhat pleomorphic, cytologically malignant cells dispersed in a stroma possessing a rich capillary network and containing large quantities of mucin, which was distributed diffusely but in places formed cell free "lakes." The cytoplasm of the tumour cells contained clear vacuoles, which often compressed the nuclei and which gave strongly positive reactions for lipid when fat stains were performed on frozen sections. The tumour pattern was typical of myxoid liposarcoma (figs 1 and 2).

![Fig 1](image1) Low power view showing strands of tumour cells separated by lakes of pooled mucin. (x 125.)

![Fig 2](image2) High power view showing pleomorphism and cytoplasmic vacuolation of lipoblasts with signet ring formation. (x 320.)

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Discussion

Liposarcomas are the commonest soft tissue sarcomas of adult life, with a peak age incidence between 40 and 60 years, and are classified microscopically into well differentiated, myxoid, round cell, and pleomorphic types. Myxoid liposarcoma is said to account for 40–50% of all liposarcomas. It very rarely originates from pre-existing lipoma, the tumour arising from primitive mesenchymal cells in most cases. The two main primary sites of origin are the extremities (especially the lower) and the retroperitoneum.3 The chest wall,4 mediastinum, and lung1-3,5-8 are far more rare primary sites (of 1067 cases of liposarcoma recorded at the Armed Forces Institute of Pathology, Washington, during 1970–9 only 29 were located in the “chest”).3

In our case there was no evidence of tumour of the chest wall. The lung parenchyma was shown, macroscopically and microscopically, not to be infiltrated by tumour and no tumour was found at any extrathoracic site. It seems likely therefore that this was either a primary pleural or primary mediastinal liposarcoma. We suggest that the extensive neoplasm enveloping the pleura of one lung, with hardly any tumour adhering to the mediastinum (it affected the adjacent pericardium only), and the clinical pattern indicate a primary pleural origin as being the more likely, though we concede that the alternative of a small mediastinal tumour progressing to extensive malignancy of the pleura cannot with certainty be excluded.

References