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Primary cardiac angiosarcoma causing rupture of the heart and spontaneous bilateral pneumothorax

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ABSTRACT A patient with a primary cardiac angiosarcoma developed two previously unreported manifestations: cardiac rupture, which has not been reported with any primary cardiac tumour; and spontaneous pneumothorax, which is well recognised with other tumours but has not been reported with a cardiac angiosarcoma.

Cardiac angiosarcoma is a rare tumour, which may present with a range of cardiac complications.

Case report

A 75 year old man presented with a one month history of haemoptysis and anorexia, and a 3 kg weight loss; he had had mild left anterior chest pain but no dyspnoea. He had smoked 30 cigarettes a day for about 30 years, but had stopped 20 years previously. Clinical examination showed nothing remarkable. The forced expiratory volume in one second (FEV_1) was 2·4 l and the forced vital capacity (FVC) 3·0 l.

Chest radiography showed a 50% left pneumothorax and three masses in the right upper lobe, right mid zone, and left mid zone. Computed tomography of the chest showed multiple small, peripheral, and ill defined opacities throughout both lungs and a mass arising from the right atrium.

The left pneumothorax was treated by intercostal tube drainage. A biopsy was then performed percutaneously on the lesion in the right upper zone with a single pass of a Rotex needle. This yielded a single crushed piece of lung parenchyma with no evidence of neoplasia. Two chest radiographs taken in the 24 hours after the needle biopsy showed only a small left apical pneumothorax. Three days later, however, chest radiography showed a new pneumothorax in the right apex, which had increased to a 50% pneumothorax by the next day, when pleural fluid was also detected. Twelve hours later he became acutely short of breath and hypotensive, with signs of both air and fluid in the right pleural space. Intercostal drainage yielded 100 ml of blood and a brisk initial air leak, but no improvement in his haemodynamic state. The patient died 50 minutes later.

At necropsy the right hemithorax contained 1300 ml of fresh blood, and clot covered the anterior and posterior surfaces of the right lung. The chest drain was situated anterior to the blood clot with the tip at the apex of the pleural space. It had not pierced the surface of the lung.

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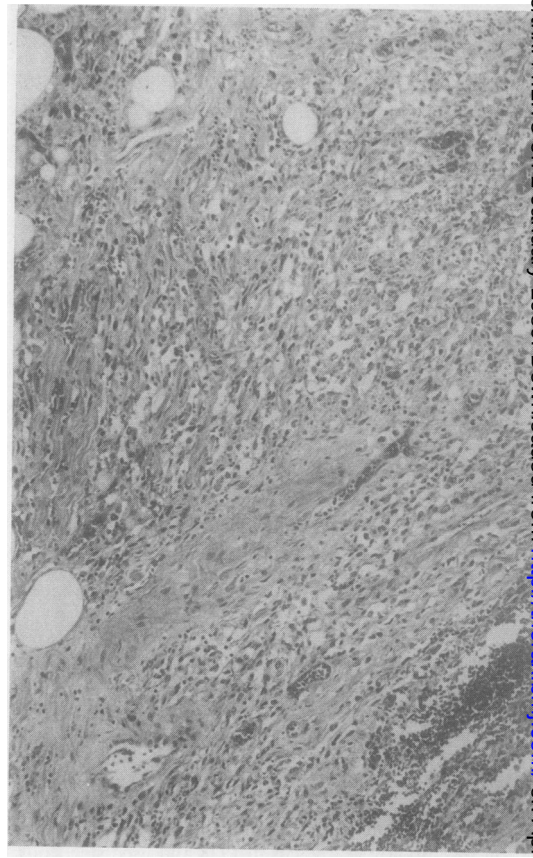

Tumour composed of pleomorphic spindle cells forming variably sized vascular channels. (Haematoxylin and eosin.)

Multiple, small soft nodules up to 1 cm in diameter were scattered throughout the parenchyma of all lobes and visible subpleurally. Similar tumour nodules were present on the diaphragm. The dilated right atrium contained a haemorrhagic, necrotic tumour measuring 10 × 8 × 4 cm (including attached thrombus) with a 0·5 cm perforation. This defect was 15 cm away from the site of the needle aspirate of the lung carried out four days earlier and was situated posteriorly, communicating with the right pleural cavity. The pericardial space was obliterated by dense fibrous adhesions. Tumour arose in and distorted the atrium in the region of the superior vena caval opening, with extension downwards

towards the fossa ovale, which was normal, and anterior to the epicardium. There was no obstruction of venous inlets or of the tricuspid valve by intracavitary tumour.

Sections of right atrial tumour and lung metastases showed poorly differentiated angiosarcoma. There was much necrosis and superimposed thrombus on the primary cardiac tumour but viable areas of tumour showed characteristic anastomosing vascular channels (figure). The lung metastases showed some solid spindle cell foci. Multiple tumour emboli were noted within peripheral pulmonary artery branches. Immunohistochemical staining for factor VIII, which gives a positive result with some angiosarcomas, was negative. Special stains excluded mesothelioma. Mediastinal lymph nodes were free of tumour. Histological examination showed no evidence of tumour in other organs.

Discussion

Cardiac rupture has been recognised for over 300 years. Both Harvey in 1649 and Morgagni in 1761 give accounts of cardiac rupture or tears. Morgagni is even said to have died from this complication. Cardiac rupture due to tumour is extremely uncommon[1-3] and is previously unreported with a cardiac angiosarcoma. An extensive search of published reports has yielded no further cases and none was reported in two large series of cardiac metastases.[4,5]

Angiosarcoma is the most common primary malignant tumour of the heart. It is usually situated in the right atrium and commonly invades the vena cava, tricuspid valve, and pericardium.[6,7] There have been no reports of associated cardiac rupture. Pulmonary metastases, evident in our patient, occurred in 61%[6] and 75%[7] of patients in two large series. Whether the pleura is affected by local extension of the primary tumour or by metastases is not clear.

Spontaneous pneumothorax is a rare but well documented feature of pulmonary tumours, both primary bronchial carcinoma and pulmonary metastases, particularly sarcomas.[8] In a review of 1143 patients presenting with spontaneous pneumothorax 10 were attributed to malignant lung neoplasms.[8] Four were due to primary bronchial carcinoma and six to metastases, all from a sarcoma. Other metastatic tumours that cause pneumothorax in rare cases include malignant melanoma,[9] carcinoma of the cervix,[10] Wilm's tumour,[10] and adenocarcinoma of unknown origin.[10] None has been secondary to cardiac angiosarcoma.

Our patient presented with a spontaneous left pneumothorax. The right pneumothorax was also probably spontaneous as there had been no detectable air leak 24 hours after a single pass with a Rotex aspiration needle in a patient shown to have multiple subpleural metastases.

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