Constrictive pericarditis caused by primary liposarcoma

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Abstract
A 30 year old man presented with symptoms of constrictive pericarditis. Echocardiography and computed tomography showed a mass extending from the pericardium to surround the heart and penetrating the left ventricular apex. An unresectable pleomorphic liposarcoma arising from the pericardium was found at thoracotomy.

Primary liposarcoma of the pericardium has not been reported previously. We describe constrictive pericarditis caused by primary liposarcoma in a 30 year old man.

Case report
A 30 year old white man presented with a two month history of progressive dyspnoea on exertion, left sided chest pain that radiated to the left shoulder, general malaise, weight loss, anorexia, night sweats, fever, and pronounced peripheral oedema. He was transferred to us from a small regional hospital with a diagnosis of suspected right sided endocarditis. On examination the patient was ill with severe dyspnoea at rest. His blood pressure was 120/85 mm Hg with pulsus paradoxus of about 30 mm Hg. The heart rate was regular at 116 beats/min and the temperature was 37.8°C. Venous pressure was visible at the angle of the jaw and his abdomen was swollen and tender with considerable hepatomegaly and ascites. Severe pitting oedema of the legs extended to the sacrum. There were broadened precordial pulsations, an apex beat palpated outside the midclavicular line, and very faint heart sounds, with a grade 2/6 systolic murmur along the right sternal border and a pericardial friction rub near the apex.

A chest radiograph showed enlargement of the heart, with irregular margins extending to the left thoracic wall, and a pleural effusion. An electrocardiogram was low voltage and showed non-specific T wave changes. Echocardiography showed a huge, irregular tumour mass surrounding the greater part of both ventricles and infiltrating the apex of the left ventricle. There was considerable limitation of diastolic filling. Computed tomography showed no extension outside the pericardium. An anterolateral thoracotomy was performed for diagnostic purposes and to decompress the myocardium. The heart was encircled by a large, richly vascular, mucoid, multilobular, green-yellow, glistening mass, encapsulated by the pericardium and attached to the apex of the left ventricle.

Frozen section examination of the tumour showed liposarcoma. Resection was impossible and the patient was allowed to die under anaesthesia according to his explicit wish before surgery if the tumour was malignant and inoperable. At necropsy the heart was encased by irregular green and yellow fatty and mucoid tissue, 25 cm in diameter, seen to contain small quantities of mucoid fluid when sectioned. The heart and tumour weighed 2750 g. The tumour was continuous with the visceral and parietal pericardium, infiltrating the apical part of the anterior and lateral left ventricle (fig 1). No metastases were identified. Light microscopic examination showed irregularly shaped cells, with large pleomorphic nuclei and slightly eosinophilic cytoplasm often containing grape like organised vacuoles, set in a stroma (fig 2). The vacuoles in many of the tumour cells stained positively with oil red O, confirming that they were fat (lipoblasts). Electron microscopic examination confirmed the diagnosis of pleomorphic liposarcoma.

Discussion

Liposarcomas occur principally in the retroperitoneal space and lower extremities and less commonly in the abdomen, buttock, vulva, neck, and mediastinum. Primary liposarcoma of the heart has been reported only twice—arising within the right ventricle and the right atrium. This is the first case report of a primary liposarcoma of the pericardium. Lagrange et al described one case of cardiac metastases from a myxoid liposarcoma and referred to four other cases.

Primary malignancies of the pericardium are rare and include mesothelioma, fibrosarcoma, angiosarcoma, and malignant teratoma. This case shows features similar to those of other primary malignant pericardial tumours, with a tendency to encase the pericardium, obliterate the pericardial space, and cause constriction of the heart. Of the diagnostic procedures we performed, echocardiography and computed tomography appeared to be the most useful. Malignant pericardial tumours typically present late, as in this case, which makes successful surgical removal unlikely.