Primary pericardial mesothelioma presenting as tuberculous pericarditis

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Summary: Ante-mortem diagnosis of primary pericardial mesothelioma is very rare. We report a case which presented clinically as tuberculous constrictive pericarditis. The patient underwent pericardial resection with an immediate haemodynamic benefit, although the malignant process progressed, and he died 14 weeks later.

Introduction

Malignant mesotheliomas may arise in the pleura (70–75%), the peritoneum (20–25%), the pericardium (4%), or, very rarely, the tunica vaginalis.¹ In all sites, the symptoms and signs are related to infiltration of adjacent structures, malignant effusion, and ultimately obliteration of the serous cavity. Metastatic spread is usually a clinically minor problem. We present the case of a patient who reported a previous diagnosis of pericardial tuberculosis, and presented with signs of constrictive pericarditis, but who was found to have a primary pericardial mesothelioma.

Case report

A 29 year old Pakistani clothing merchant presented with a short history of increasing breathlessness. He said that 4 months previously he had been diagnosed as having pericardial tuberculosis in Pakistan and had been on anti-tuberculous therapy since. He gave no history of exposure to asbestos. He was dyspnoeic with an elevated jugular venous pressure, crepitations in the bases of both lungs, and peripheral oedema. There was left cervical lymphadenopathy. He was admitted and the antituberculous therapy continued with the addition of steroids. Baseline biochemical and haematological indices showed an albumin of 29 g/l, total protein 81 g/l, bilirubin 21 μmol/l, and an alkaline phosphatase of 336 μmol/l. He had an erythrocyte sedimentation rate of 25 mm/h. The chest X-ray showed bilateral pleural effusions and the electrocardiogram a low voltage pattern. An echocardiogram showed a grossly thickened pericardium with a small posterior pericardial effusion (Figure 1). A pericardiocentesis was performed under echocardiographic control and 150 ml of blood-stained fluid was removed. However, following the procedure he developed increasing dyspnoea, cyanosis, further elevation of the jugular venous pressure, a tachycardia of 120/min, and a falling systolic pressure to 90 mmHg with a pulsus paradoxus of 20 mmHg. At emergency pericardectomy, through a bilateral sub-mammary incision with cardiopulmonary bypass standby, the mobility of the heart was noted to be markedly restricted and the parietal and visceral pericardium irregularly thickened to about 2 cm. The lungs and pleura were normal and bilateral serous effusions were drained. The pericardium was resected, apart from two strips containing the phrenic nerves, and the central venous pressure fell from 18 to 8 mmHg.

Histological examination of the resected pericardium showed a neoplasm (Figure 2) composed of pleomorphic spindle and cuboidal cells with abundant cytoplasm, arranged in fascicles, solid nodules with cleft formation, and a few areas with a papillary pattern. The mitosis count was 30 per 10 high power fields (magnification x 400) with occasional abnormal mitoses. The tumour cells showed positive staining with CAM5.3, a marker for cytokeratin, and were negative for carcinoembryonic antigen (CEA). Stains for acid mucopolysac-
Primary pericardial mesotheliomas are rare, with only one diagnosed in 496,795 consecutive hospital admissions, and one in 18,328 autopsies at the Henry Ford Hospital, USA, and a calculated annual incidence of one in 41 million in Canada. Pericardial mesothelioma may occur at any age. There are too few published cases for an assessment of aetiological factors. The wide age range and lack of a consistent occupational history preclude a widespread association with asbestos, although there is one report of intrathoracic mesothelioma in a patient who had pericardectomy and application of asbestos for angina pectoris, and one report of pericardial mesothelioma in a patient with pleural plaques and ferruginous bodies in the lungs.

The histological features of primary mesothelioma of the pericardium do not differ from those of mesothelioma arising in the pleura or elsewhere. Most cases are of a mixed type with both spindle cell and glandular elements. The major differential diagnoses are from a benign reactive proliferation of the mesothelium, in which necrosis, infiltration and cytological atypia are not prominent, and from secondary malignancies, especially adenocarcinoma. Hyaluronidase-labile mucin production is variable, and the most helpful, commonly available immunohistochemical stains are CAM5.2 and CEA. If the latter is negative, a diagnosis of adenocarcinoma is unlikely.

Pericardial mesothelioma can present in such a way that mistaken diagnoses of mitral stenosis in rheumatic fever and atrial myxoma have been made, the true nature of the lesion becoming apparent only at operation or post-mortem. The most common presentation is with compromised cardiac function due to pericardial effusion, or due to constriction by an infiltrating mass of tumour.

The differential diagnosis of subacute constrictive pericarditis in a patient from the Indian sub-continent is wide and a common cause is tuberculosis. Tubercle bacilli are demonstrated in less than half of these cases. One case of pericardial mesothelioma with coexistent pericardial tuberculosis has been reported from India. We have no independent evidence of tuberculosis in our case. Cultures of pleural and pericardial fluid were negative. Although the history, signs, and echocardiographic findings were consistent with tuberculous pericarditis, persistence or worsening of symptoms and effusion in a patient on effective anti-tuberculous therapy, with steroids, should perhaps alert the clinician to the possibility of another aetiology. As in pleural mesothelioma, primary pericardial mesothelioma is an aggressive disease and in our patient this was responsible for his death within 4 months of diagnosis.

Discussion

Figure 1  Echocardiogram showing markedly thickened pericardium and pleural effusion. P = pericardium; PE = pleural effusion; RV = right ventricle; LV = left ventricle; LA = left atrium; Ao = Aorta.

Figure 2  Photomicrograph showing predominant spindle cell area of pericardial malignant mesothelioma. Cellular pleomorphism and several mitotic figures are evident. (Haematoxylin and Eosin) (Bar = 30 μm).

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