Rev Port Cardiol. 2013;32(2):149–152

Revista Portuguesa de
Cardiologia
Portuguese Journal of Cardiology
www.revportcardiol.org

CASE REPORT

A rare cause of pericardial disease☆

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Received 23 February 2012; accepted 3 May 2012

KEYWORDS
Constrictive pericarditis;
Mesothelioma

Abstract Among cardiovascular diseases, pericardial disease has specific characteristics. Its etiology, diagnosis and medical management are not as well understood as in coronary and valvular heart disease. In most cases, its cause is benign, although the proportion decreases with more severe clinical presentation.

The authors present the case of a 35-year-old man with no relevant past medical history, who went to the emergency department with what appeared to be an idiopathic case of acute pericarditis. However, over the following five months, there was an unfavorable evolution to constrictive pericarditis, requiring pericardiectomy. The final diagnosis was only made following surgery – a rare case of a primary pericardial tumor, a mesothelioma.

PALAVRAS-CHAVE
Pericardite constritiva;
Mesotelioma

Causa rara de doença pericárdica

Resumo As doenças do pericárdio apresentam-se como uma patologia particular do foro cardiovascular. Os seus componentes etiológicos e a gestão diagnóstica e terapêutica não estão tão bem compreendidos e estudados, comparativamente com outras áreas, como a doença coronária ou valvulopatias. Maioritariamente, a etiologia é benigna, mas a sua proporção diminui à medida que a apresentação e evolução clínicas são mais exuberantes.

Os autores descrevem um caso de um homem de 35 anos de idade, sem antecedentes clínico-patológicos de relevo conhecidos, que se apresenta num Serviço de Urgência com o que aparenta ser um episódio de pericardite aguda de etiologia idiopática. Contudo, ao longo de cinco meses, evolui desfavoravelmente, com necessidade de orientação para pericardiectomia por pericardite constritiva. Apenas no bloco operatório foi feito o diagnóstico etiológico final. Tratava-se de um caso muito raro de neoplasia primária do pericárdio, um mesotelioma.

☆ Please cite this article as: Ramos V, et al. Causa rara de doença pericárdica. Rev Port Cardiol. 2013. http://dx.doi.org/10.1016/j.repc.2012.05.024.

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Introduction

The etiology and management of pericardial disease are often unclear, since they do not enjoy the consensus surrounding other types of cardiovascular disease. However, prognosis is generally favorable and invasive intervention or extensive investigation is usually unnecessary.[1]

Malignant primary cardiac tumors are rare, particularly those originating in the pericardium, which is more likely to be affected by metastases.[2] Thus, defining the typical clinical presentation and diagnostic and medical management are hindered by the scarcity of cases described in the literature.[2]

Case report

A 35-year-old man, Caucasian, a construction worker, with no relevant past medical history and taking no regular medication, went to the emergency department in November 2008 for persistent crushing chest pain radiating to the shoulders and worsening on deep breathing and in dorsal decubitus; he had no other symptoms and no abnormalities on physical examination. The electrocardiogram showed sinus rhythm, diffuse concave ST-segment elevation and PR-segment depression in the inferior leads. Laboratory tests and chest X-ray were within normal parameters. A diagnosis of idiopathic acute pericarditis was made, and the patient was medicated with intravenous aspirin, which improved his symptoms. He was prescribed aspirin, discharged home and referred for cardiology consultation. On assessment a month later, he was asymptomatic and echocardiography showed a moderate pericardial effusion but no other significant changes. He was prescribed colchicine and ibuprofen.

Four months later, in February 2009, he again went to the emergency department for interscapular pleuritic pain and new-onset dyspnea on moderate exertion of 15 days' evolution. Cardiac auscultation was normal; pulmonary auscultation revealed decreased breath sounds and vocal fremitus in the right lung base, and jugular venous distension at 45°, with no Kussmaul sign or paradoxical pulse. Echocardiographic reassessment (Figure 1) showed preserved biventricular systolic function and no dilatation, with pericardial thickening and a small circumferential effusion of organized appearance, and no significant variation in transvalvular flow over the respiratory cycle. The electrocardiogram revealed diffuse T-wave inversion but no ST- or PR-segment alterations. Laboratory tests were similar to the previous results and the chest X-ray showed a small right basal pleural effusion. The patient was admitted for investigation of the organized pericardial effusion which appeared to be evolving to constrictive pericarditis of unknown etiology.

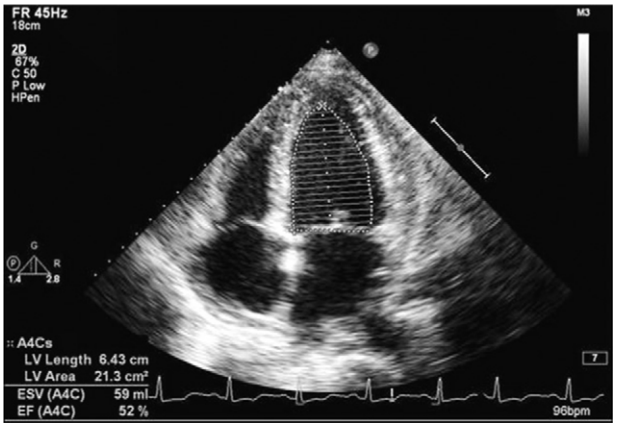

Figure 1 Echocardiogram after the patient's second visit to the emergency department, showing pericardial thickening and a small effusion of organized appearance.

Thorough etiological study, including thoracentesis, screening for sepsis (serology for infectious agents, blood cultures and microbiological analysis of pleural fluid), tuberculin test and thyroid function, was negative. Thoracic, abdominal and pelvic computed tomography was also performed, which showed diffuse pericardial thickening with no significant effusion, enlarged paratracheal lymph nodes (28 mm maximum diameter), apparently of an inflammatory nature, and bilateral pleural effusion, more pronounced on the right. The patient was referred for cardiac catheterization, which showed elevation and equalization of atrial and ventricular diastolic pressures (32 mmHg), with intraventricular pressure curves showing the square root sign and respiratory variation suggestive of ventricular interdependence, but no angiographic coronary artery or valve disease. Pericardiocentesis was not performed since the pericardial effusion was not significant and thus the window to perform it safely was small. The patient was discharged home, clinically improved, with a diagnosis of constrictive pericarditis, to await early elective pericardiectomy. However, he again suffered clinical worsening with decompensated heart failure and episodes of intense retrosternal pain associated with hypotension, and was readmitted a week after discharge.

During this hospitalization, therapeutic thoracentesis was performed twice for marked pleural effusion (more severe on the right), as well as colonoscopy due to new-onset abdominal pain with rectal bleeding, which revealed friable, congested and bleeding sigmoid mucosa, histologically compatible with ischemic colitis.

The patient was transferred to a referral surgical center (five months after onset of the clinical setting) for pericardiectomy. Intraoperatively, a fibrotic and infiltrative neoplastic process was observed in the heart, more marked in the right atrium and great vessels, making resection impossible. In view of the patient's hemodynamic instability, requiring invasive vasopressor and ventilatory support, he was transferred to the intensive care unit. Control echocardiography showed moderate biventricular systolic dysfunction with paradoxical interventricular septal motion, a moderately large pericardial effusion and significant respiratory variation in transmitral flow. The patient died three days later.

Anatomopathological study revealed a malignant epithelial neoplasm with a storiform pattern and trabecular areas, as well as spindle cells (Figure 2). Immunohistochemical analysis showed immunoreactivity of the tumor cells to cytokeratin 7 and AE1/AE3 and multifocal areas positive for calretinin and cytokeratin 5, which, together with the absence of pleural involvement, led to a final diagnosis of a primary pericardial biphasic mesothelioma.