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CASE REPORT

Effusive-constrictive pericarditis as the manifestation of an unexpected diagnosis☆

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KEYWORDS
Effusive-constrictive pericarditis;
Heart failure;
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Abstract Constrictive pericarditis is a clinical condition characterized by the appearance of signs and symptoms of right heart failure due to loss of pericardial compliance. Cardiac surgery is now one of the most frequent causes in developed countries, while tuberculosis remains the most prevalent cause in developing countries. Malignancy is a rare cause but usually has a poor prognosis. The diagnosis of constrictive pericarditis remains a clinical challenge and requires a combination of noninvasive diagnostic methods (echocardiography, cardiac magnetic resonance and computed tomography); in some cases, cardiac catheterization is needed to confirm the diagnosis. The authors present the case of a 51-year-old man, hospitalized due to cardiac tamponade. Diagnostic investigation was suggestive of tuberculous etiology. Despite directed medical therapy, the patient developed effusive-constrictive physiology. He underwent pericardiectomy and anatomopathologic study suggested a neoplastic etiology. The patient died in the postoperative period from biventricular failure.

PALAVRAS-CHAVE
Pericardite efusiva-constritiva;
Insuficiência cardíaca;
Ecocardiografia;

Pericardite efusiva-constritiva como manifestação de um diagnóstico inesperado

Resumo A pericardite constritiva é uma entidade clínica caracterizada pelo aparecimento de sinais e sintomas de insuficiência cardíaca direita, secundários à perda da *compliance* pericárdica. Atualmente, a cirurgia cardíaca tornou-se numa das etiologias mais frequentes nos países desenvolvidos, mantendo-se a tuberculose como a causa mais prevalente nos países em vias de desenvolvimento. As etiologias neoplásicas são mais raras e habitualmente de pior

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prognóstico. O diagnóstico desta entidade mantém-se um desafio clínico, sendo necessária a integração dos achados dos métodos de diagnóstico não invasivos (ecocardiografia, ressonância magnética e tomografia computorizada) e por vezes o recurso ao cateterismo cardíaco. Os autores apresentam o caso clínico de um homem de 51 anos de idade, internado por tamponamento cardíaco. A investigação etiológica foi sugestiva de etiologia tuberculosa, que apesar da terapêutica médica dirigida, evoluiu para fisiologia efusiva-constritiva. Foi submetido a pericardiectomia e o exame anátomo-patológico sugeriu etiologia neoplásica. O doente veio a falecer no pós-operatório em falência biventricular.

Introduction

Constrictive pericarditis is a clinical condition characterized by the appearance of signs and symptoms of right heart failure due to loss of pericardial compliance.[1] Cardiac surgery, radiotherapy and idiopathic pericarditis are now the most common causes in developed countries, but tuberculosis remains the most prevalent cause in developing countries.[1]

Diagnosis is usually established by Doppler echocardiography, but doubtful cases may require other diagnostic methods, such as cardiac magnetic resonance (CMR), computed tomography (CT) or invasive hemodynamic assessment by cardiac catheterization.

Treatment is based on administration of diuretics and directed therapy for the underlying condition. However, pericardiectomy is the only definitive and potentially curative treatment and is recommended in cases with signs of chronicity.

Case report

A 51-year-old man, black, regularly participating in sports and with no relevant medical history, came to the emergency department for progressively worsening fatigue, occasional sharp chest pain, increased abdominal volume, loss of appetite and food intolerance with episodes of vomiting for around a week. Three days before admission, he had an episode of syncope and noticed significant jugular distension. He denied fever, chills, cough, expectoration or weight loss. On physical examination, his blood pressure was 118/78 mmHg and heart rate was 98 bpm; he was apyretic and dyspneic, with jugular distension and faint heart sounds, but no peripheral edema. The electrocardiogram (ECG) showed sinus rhythm, with low QRS voltage in the classical and limb leads and an S1Q3T3 pattern (Figure 1). Laboratory tests showed slight elevation of inflammatory parameters (leukocytes 11.6×10^9/l, C-reactive protein 3.9 mg/dl), marked D-dimer elevation (5153 ng/ml), hemoglobin 13.3 g/dl, creatinine 1.3 mg/dl and negative myocardial necrosis markers. Chest CT angiography excluded pulmonary thromboembolism but revealed a large right pericardial and pleural effusion. Echocardiography showed a large, circumferential pericardial effusion, cloudy in appearance, with a maximum size of 32 mm, and partial right chamber collapse (Figure 2).

Pericardiocentesis was performed under fluoroscopic guidance for diagnostic and therapeutic purposes, a total of 4000 cc of bloody fluid being drained; cytochemical study revealed this to be an exudate, with normal adenosine deaminase levels. Cytology revealed no cancer cells and microbiological study, both direct and in culture, was negative, including for mycobacteria. Various markers of autoimmune disease (antinuclear antibody, rheumatoid factor, anti-cardiolipin, anti-beta-2 glycoprotein and C3 and C4 levels) and blood cancers (carcinoembryonic antigen, cancer antigen 19.9, prostate-specific antigen, alpha-fetoprotein, beta-2 microglobulin and peripheral blood smear) were analyzed, none being positive for diagnosis. Investigation of viral antibodies (echovirus, Coxsackie, adenovirus, influenza A and B, human immunodeficiency virus, and hepatitis B and C) was negative or not suggestive of recent infection. Chest-abdominal-pelvic CT revealed polyserositis only, with no adenopathy or other suspicious masses. A subsequent interferon-gamma release assay was positive, and so a diagnosis of tuberculous pericarditis was assumed and therapy was begun with antituberculosis drugs.

After two months of antituberculosis therapy, repeat echocardiography showed signs of pericardial constriction (septal bounce and significant respiratory variation in cardiac valve and hepatic vein flows – Figure 3), together with a thin rim of pericardial effusion. CMR confirmed these findings and revealed pericardial thickening, with no signal intensification after contrast injection and an extensive area of late enhancement, as well as areas of pericardiamyocardial adherence and signs of constrictive physiology (septal bounce) (Figure 4). Clinically, predominantly right-sided heart failure was observed during the third month of medical therapy, prompting rehospitalization; this rapidly worsened and the patient developed anasarca, liver failure, exudative enteropathy and cachexia. In view of the diagnosis of effusive-constrictive pericarditis, with rapid and progressive clinical deterioration and signs of chronicity, he was referred for surgical pericardiectomy. Intraoperatively, complete pericardial symphysis was observed, with a macroscopic appearance suggestive of tumor infiltration