Unusual association of diseases/symptoms

Effusive-constrictive pericarditis in the absence of overt pericardial disease on multiple imaging modalities

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Summary

Effusive-constrictive pericarditis (ECP) is a rare condition that may prove fatal without appropriate treatment. In ECP, there is concomitant existence of a pericardial effusion and CP, that together lead to impaired cardiac filling. Therapeutic pericardiocentesis only addresses part of the problem; surgical pericardiectomy may be required to relieve the constrictive element. Imaging in ECP characteristically demonstrates calcification or thickening of the pericardium. The authors describe a case of ECP where a number of imaging modalities (including echocardiography, cardiac magnetic resonance and CT) did not identify overt pericardial disease. The patient underwent surgical pericardiectomy that led to a rapid resolution of symptoms and full recovery. Histopathological analysis of the pericardial biopsy indicated a non-specific pericardial fibrosis, in keeping with the final diagnosis of ECP.

BACKGROUND

Effusive-constrictive pericarditis (ECP) is a rare condition that may prove fatal without appropriate treatment. In ECP, there is concomitant existence of a pericardial effusion and CP, that together lead to impaired cardiac filling. Therapeutic pericardiocentesis only addresses part of the problem; surgical pericardiectomy may be required to relieve the constrictive element. ECP should be strongly suspected when neck veins continue to be dilated after drainage of a pericardial effusion. ECP usually presents with symptoms of heart failure; it is often idiopathic but may occur as a consequence of malignancy, medication, post-radiotherapy and in connective tissue disorders.^{1 2} Although the pericardium may uncommonly be of normal thickness in ECP, there are usually other signs of pericardial disease such as calcification of the pericardium. We describe a case of ECP where a number of imaging modalities revealed normal thickness pericardium without calcification, initially raising doubts about the underlying diagnosis despite suggestive clinical features and haemodynamic data.

CASE PRESENTATION

A 62-year-old Caucasian gentleman with long-standing rheumatoid arthritis was admitted with increasing shortness of breath, reduced exercise tolerance and intermittent atypical cardiac chest pain. On examination, he was haemodynamically stable, the jugular venous pressure was elevated and peripheral oedema extended to the abdomen. An echocardiogram was performed and demonstrated good left and right ventricular function, a sizeable pericardial effusion but no echocardiographic evidence of cardiac tamponade; the pericardium appeared to be of normal thickness. Diagnostic pericardiocentesis confirmed an exudative effusion with no evidence of malignancy or infection (including tubercle infection). Blood tests revealed a slight anaemia with mildly elevated inflammatory markers; levels of rheumatoid factor and anticitrullinated

protein antibody were raised. He underwent a CT pulmonary angiogram that revealed the pericardial effusion in the absence of thickening or calcification of the pericardium (figure 1); bilateral pleural effusions were present but no evidence of pulmonary embolism. Initially, he was treated with diuretic therapy. One week later, he developed signs of cardiac tamponade. A pericardial drain was inserted and despite drainage of 450 mls of fluid and echocardiographic resolution of the effusion, he remained markedly symptomatic with exertional dyspnoea and residual peripheral oedema.

Figure 1 Image from CT pulmonary angiogram showing a pericardial effusion, absence of thickening/calcification of the pericardium and bilateral pleural effusions.

Figure 2 Simultaneous right atrial and left ventricular pressure tracings demonstrating equalisation of the pressure tracings during diastole and the 'square root' or 'dip and plateau' sign.

INVESTIGATIONS

He underwent left and right heart cardiac catheterisation and cardiac MRI (CMR) after pericardiocentesis. Figure 2 demonstrates simultaneous right atrial and left ventricular pressures that show the classical 'dip and plateau' or 'square root' sign which can be seen in both CP and restrictive cardiomyopathy; a pattern that reflects the rapid increase in pressure associated with early diastolic filling which abruptly plateaus as a result of impaired further filling secondary to the constrictive or restrictive process. In this patient, there was also equalisation of the pressures in the left and right heart during diastole, which is seen in CP. Figure 3 is a still image from the CMR that revealed normal cardiac chamber size and normal pericardial thickness of 2 mm. On inspiration, there was flattening of the septum and increased size of the right ventricle indicating ventricular interdependence. Despite lack of imaging evidence of abnormal pericardial anatomy, the cardiac catheterisation data and abnormal cardiac physiology indicated probable CP. These signs were unmasked after drainage of the effusion which is characteristic of ECP. In the context of normal pericardial anatomy, restrictive cardiomyopathy is an important differential diagnosis. Although systolic function is often normal in restrictive cardiomyopathy, there is usually left ventricular hypertrophy with marked atrial dilatation; neither of these features was present in this case. In restrictive cardiomyopathy, the septum is involved in the pathological 'stiffening' process and as a result, the septal bounce is not seen.

OUTCOME AND FOLLOW-UP

Our patient underwent surgical pericardiectomy that led to a rapid resolution of symptoms and full recovery. Again, on

Figure 3 Cardiac MRI showing the residual pericardial effusion, normal chamber size and absence of thickening/calcification of the pericardium.

raw inspection, the pericardium was not thickened and there was no evidence of calcification. Histopathological analysis of the pericardial biopsy indicated a non-specific pericardial fibrosis, in keeping with the final diagnosis of ECP.

DISCUSSION

ECP has been described in a number of case reports^{3 4} and more recently, these have highlighted the value of imaging.⁵⁻⁷ Thickening and/or calcification of the pericardium

have been a consistent finding in these reports. As far as we are aware, this is the first case of ECP with no evidence of overt pericardial pathology, such as thickening or calcification, on multiple imaging modalities. The importance lies in the fact that these features, while characteristic of CP, do not exclude the diagnosis. Talreja *et al*⁸ described normal pericardial thickness in 18% of patients, as identified postresection, in a retrospective study of 143 patients undergoing pericardiectomy for CP. Even though the pericardium was of normal thickness in this small cohort of CP patients, there was evidence of pericardial disease such as calcification on chest x-ray, CT or echocardiography. The diagnosis of ECP in our patient was made belatedly based on persisting symptoms, a failure of venous pressures to drop postpericardiocentesis and the haemodynamic data.

Learning points

- ▶ This case illustrates that in ECP the pericardium may have a normal thickness and be 'acalcific' but still constricts the heart.
- ▶ Haemodynamic data and evidence of ventricular interdependence, together with clinical signs and suspicion, remain important to timeously reach the diagnosis.
- ▶ Timely identification of ECP facilitates early, potentially life-saving treatment.

Competing interests None.

Patient consent Obtained.

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