Pericardial disease

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Abstract

Pericardial diseases are common but rigorous large-scale or randomized studies of current clinical practice remain limited. This article outlines a current and practical approach to the clinical assessment and investigation of pericarditis, pericardial effusions and constrictive pericarditis. The management of these conditions is discussed in line with best practice. Limited clinical trial data underlines the importance of bedside and investigative skills in assessing both the function and form of this organ and the treatment options.

Keywords Acute pericarditis; constrictive pericarditis; pericardiocentesis; pericardium; tamponade

What's new?

- A recent multicentre randomized trial has confirmed the value of colchicine, added to standard anti-inflammatory therapy, in reducing both recurrence rates and symptom persistence after acute pericarditis

Introduction

The pericardium is a continuous sac around the heart analogous to the pleura surrounding the lungs and peritoneum surrounding the abdominal viscera. Anatomically it is divided into:

- fibrous pericardium: the outermost layer in continuity with the adventitia of the great vessels
- serous pericardium: itself divided into a parietal layer that is adherent to the fibrous pericardium and a visceral layer, which covers the epicardial surface of the heart.

Between the parietal and visceral layers of the serous pericardium is a potential space, the *pericardial space*, which in physiological situations contains a small volume of *pericardial fluid*.

The function of the pericardium is unclear. It is a relatively non-distensible structure that maintains cardiac size and prevents dilatation during cardiac development. The fluid-filled pericardial space may reduce friction between the epicardial surface of the heart and surrounding structures. However, the pericardium does not seem essential for cardiac development or function (most patients with complete congenital absence of pericardium are asymptomatic) and removal of the pericardium at operation has no obvious detrimental effect.

Pericarditis[1,2]

Aetiology

Inflammation of the pericardium has multiple causes. These can broadly be divided into infectious (viral, bacterial, mycobacterial or, rarely, fungal), inflammatory (connective tissue disorders, vasculitis), metabolic (uraemia, hypothyroidism), neoplastic conditions and local injury (including trauma, postoperative, or following myocardial infarction and acute dissection). Frequently no specific cause is identifiable. An autoimmune process may be central to 'idiopathic' and, indeed, several other causes of pericarditis (including post-cardiomyotomy, rheumatic pericarditis and 'Dressler's syndrome').

Clinical presentation

This is highly variable. Acute (often viral or idiopathic) pericarditis may account for 5–10% of emergency department presentations with chest pain syndromes. The pain is classically sharp, worse on inspiration and lying flat, and may be associated with fever, tachycardia and myalgia. On auscultation, a pericardial friction rub may be heard. The classical ECG changes are scalloped ST-segment elevation with PR-depression, which often involves all leads (except aVR) but may be absent or localized to chest or limb leads (Figure 1). QRS prolongation, QT dispersion and QT shortening are absent, in contrast to ECG changes in ST-segment elevation myocardial infarction.[3] ECG changes may evolve through the disease process with resolution of ST and PR changes, and the development of widespread T-wave changes, probably reflecting some myocardial involvement. The white cell count and inflammatory markers may be elevated. Concurrent elevation of cardiac-specific biomarkers, such as troponin, reflects extension of the inflammatory process into the myocardium (*myopericarditis*). Echocardiography often demonstrates a small pericardial effusion. The relative sensitivity and specificity of both clinical findings and routine investigations have not been assessed. Invasive investigations, such as pericardial biopsy, are not usually required because acute pericarditis runs a benign course in the majority of patients.[4]

More insidious forms may present with vague symptoms, or with pericardial effusion or constriction.

Recurrence

Reported recurrence rates in acute pericarditis depend on the population studied and disease definitions. In a recent multicentre trial of patients with a first presentation of acute pericarditis, recurrence or persistence rates in the whole study population were 37.5%, driven predominantly by symptomatic episodes. Tamponade or constriction was not observed. Serious long-term sequelae, such as recurrent tamponade, constrictive pericarditis or multiple highly symptomatic recurrences, are rare.

Bacterial and tuberculous pericarditis

Purulent pericarditis is rare but often fatal unless treated promptly by appropriate-spectrum antibiotics and either pericardial drainage and washout or pericardiectomy. Bacterial

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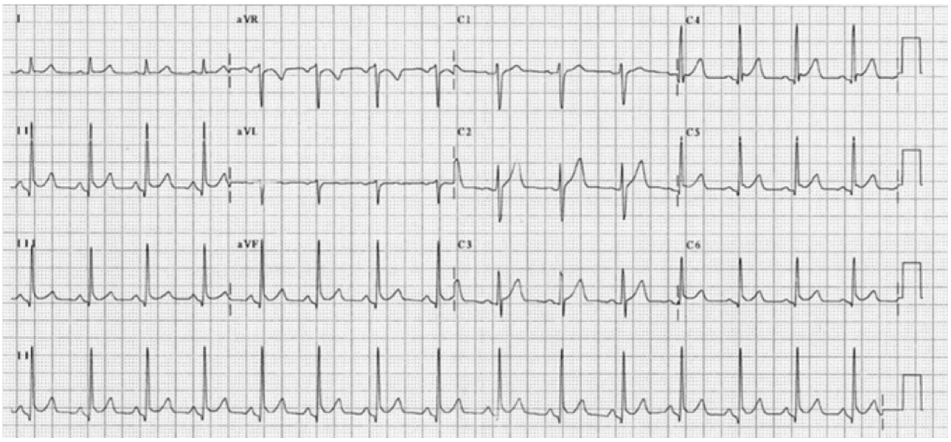

Figure 1 Typical electrocardiograph (ECG) findings in acute pericarditis. There is widespread scalloped ST-elevation with downsloping PR-depression. (Image courtesy of Dr. Jim Newton).

pericarditis usually complicates infection elsewhere, especially in the context of immunosuppression.

Tuberculous pericarditis is now less common in developed countries but has a high mortality without treatment. Diagnosis is achieved by identification of *Mycobacterium* species directly or on culture, or of mycobacterium DNA by polymerase chain reaction from pericardial tissue or fluid. Elevation of adenosine deaminase activity in the pericardial fluid has been shown to have 90% sensitivity and 74% specificity for tuberculous pericarditis. Other potential diagnostic tests include pericardial lysozyme and interferon-γ. Treatment is with appropriate antituberculous agents but the subsequent incidence of pericardial constriction is as high as 30–50%. Up to one half of tuberculous pericarditis occurs in the context of HIV infection.

Treatment

General: patients with idiopathic or viral pericarditis can be safely managed in an outpatient setting once a significant pericardial effusion has been excluded. Specific therapies, such as antimicrobial and antituberculous agents or immunosuppression, may be appropriate depending on the underlying cause.

NSAIDs: in most patients, symptomatic relief can be gained with non-steroidal anti-inflammatory agents such as ibuprofen (300–800 mg 6–8 hourly) but aspirin (650 mg 6 hourly) may be preferred in patients with pericarditis complicating myocardial infarction. There are no randomized data available to demonstrate the efficacy of these agents or to judge duration of therapy. In general, treatment is continued until symptoms, pericardial effusion and markers of inflammation have resolved or returned to normal.

Colchicine: the ICAP multicentre randomized double blind trial of colchicine (weight-adjusted 0.5–1 mg once or twice daily) in 240 patients (2013) followed the earlier COPE study and showed a significant 65% reduction in incessant or recurrent pericarditis and symptom persistence at 72 hours, together with reductions in both hospitalization and individual patient recurrences. Therapy was added to either aspirin or ibuprofen. Serious adverse effects were not observed, although in some patients[4] gastrointestinal effects, in particular diarrhoea, can limit dosage and duration of therapy.[5]

Corticosteroids: in a systematic review of therapy for acute pericarditis, corticosteroids were associated with a trend for increased risk of recurrence but, conversely, low-dose corticosteroids were superior to high-dose with regard to treatment failure or recurrences. Corticosteroids are recommended only for treatment of pericarditis associated with underlying autoimmune or connective tissue disease. Use of corticosteroids in tuberculous pericarditis has been proposed, to improve outcome or prevent progression to constriction, but a number of small randomized studies have not demonstrated clear benefit.[6]

Pericardiectomy: this is reserved for recurrent pericarditis refractory to medical therapy.

Pericardial effusion and tamponade

The pericardial space normally contains only a few millilitres of fluid although this volume may increase in a number of situations. The fibrous nature of the pericardium renders it relatively indistensible in the short term although gradual remodelling and dilatation can occur over longer periods. Pericardial pressure rises if the rate of blood or fluid accumulation exceeds the rate at which the pericardium can remodel; if that pressure exceeds the pressure within the cardiac chambers, external compression occurs. Compromised cardiac filling in diastole, especially during inspiration, further compromises left ventricular filling and cardiac output falls, resulting in *cardiac tamponade*. Tamponade is a life-threatening medical emergency treated by urgent drainage of the pericardial space (*pericardiocentesis*).[7]

Acute versus chronic pericardial effusions

The size of a pericardial effusion varies considerably and does not reflect its haemodynamic importance. Chronically acquired effusions often exceed a litre in volume whereas acute effusions of only a hundred millilitres may cause haemodynamic compromise.