Chronic Pericarditis

Chronic pericarditis is long-lasting, gradual inflammation of the pericardium, causing accumulation of fluid in the pericardial space or thickening of the pericardium. The chronic varieties of pericarditis are rare. Chronic pericarditis is usually preceded by acute pericarditis (although acute pericarditis is more common and usually self-limiting).

There are two main types encompassed by this term:

- Chronic effusive pericarditis
- Chronic constrictive pericarditis

Effusive-constrictive pericarditis (persistence of symptoms and signs of constrictive pericarditis after removal of pericardial fluid) suggests a clinical continuum initiated by acute pericarditis and progressing through pericardial effusion, chronic effusive pericarditis, effusive-constrictive pericarditis to chronic constrictive pericarditis.

Anatomy

The normal pericardium has two layers (outer fibrous pericardium and inner serous pericardium). There are approximately 50 ml of fluid in the intrapericardial space or pericardial cavity, ie the space between the serous pericardium next to the heart and the serous pericardium next to the fibrous pericardium.

The pericardium:

- Helps cardiac efficiency by limiting dilatation, aids atrial filling, etc.
- Protects the heart by reducing external friction and providing a barrier to extension of infection and malignancy.
- Fixes the heart anatomically through ligamentous connections.

Aetiological factors

The range of aetiologies should be considered alongside prevalence as many on the list are possible but unlikely diagnoses:

Idiopathic

- It is common for no antecedent diagnosis to explain the inflammatory process.
- Unrecognised viral pericarditis (Coxsackievirus, echovirus and adenovirus) may explain some of these, which constitutes the most common aetiological factor for all forms of pericarditis (both acute and chronic), particularly in developed countries.

Infection

- Infection can be viral, bacterial, fungal or parasitic.
- Tuberculosis (TB) is the main cause of constrictive pericarditis in developing nations but is less common in developed countries.
Inflammation

- Many different inflammatory conditions can affect the pericardium.
- Between 11% and 50% of patients with rheumatoid arthritis have pericarditis at autopsy.
- Systemic lupus erythematosus (SLE), scleroderma, sarcoidosis, granulomatosis with polyangiitis (Wegener's granulomatosis) and others can all cause chronic pericarditis.

Metabolic

- Renal failure (35-50% of patients with uraemia, pre-dialysis have pericarditis).
- Hypothyroidism.
- Cholesterol pericarditis (gold paint pericarditis).

Cardiovascular disease

- Myocardial infarction causes acute pericarditis (transmural infarcts, persistent ST elevation).
- Dressler's syndrome (2-3 weeks after myocardial infarction).
- Aortic dissection.

Neoplastic

- 5-17% of pericarditis.
- This is caused mostly by metastatic disease (lung 33%, breast 25%, haematological 15%).

Miscellaneous

- Drugs: doxorubicin; cyclophosphamide; drug-induced SLE (methylldopa, isoniazid, hydralazine); methysergide; smallpox vaccination; dantrolene, phenytoin; minoxidil.
- Irradiation.
- Post-pericardiotomy syndrome (ie after cardiac surgery 10-40%).
- Trauma, especially with oesophageal rupture or pancreatitis.

Chronic effusive pericarditis

Aetiology

This is most commonly idiopathic but can follow any of the causes of acute pericarditis, particularly malignancy (most commonly breast and lung), TB - more common in developing countries - or hypothyroidism.

Presentation

Patients with effusive-constrictive pericarditis have both a pericardial effusion and constrictive pericarditis. They usually present with a pericardial effusion and evidence of increased filling pressure with cardiac tamponade or constriction.

- Variable but dyspnoea on exertion, particularly in severe cases.
- Cardiovascular symptoms include chest pain, pressure or discomfort, syncope, light-headedness and palpitations.
- Many patients are asymptomatic until the disease is advanced (up to 2 L of fluid can build up).
- Respiratory symptoms, including cough and hoarseness, may occur.
- Other symptoms include fatigue, hiccoughs, anxiety and confusion.
- Attention should be given to any history of specific antecedent conditions (eg, cardiac surgery, renal failure, radiation treatment, malignancy, TB).

Signs may include hypotension, elevated jugular venous pressure (JVP) and diminished heart sounds (Beck's triad or acute compression triad described in 1935 with cardiac tamponade). Signs may develop towards those of cardiac tamponade (Beck's triad, pulsus paradoxus, Kussmaul's sign, etc). Cardiac dullness is an unreliable sign. Pericardial friction rub is best heard in the supine position at the end of exhalation. Hepatojugular reflux may be observed.

Chronic constrictive pericarditis

Aetiology

These parallel those of antecedent acute pericarditis and hence are many and varied. The risk of constrictive pericarditis is higher following bacterial forms of pericarditis, intermediate for post-pericardiotomy syndromes and systemic inflammatory diseases and low for viral and idiopathic cases of pericarditis[1].

Constrictive pericarditis is underdiagnosed, mainly because of the difficulty in differentiating it from restrictive cardiomyopathy and other causes of right heart failure[2].
Constrictive pericarditis can occur after any pericardial disease process. The more common causes of constrictive pericarditis are:

- Idiopathic.
- Viral.
- TB (the highest total incidence, as common in developing countries).
- Mediastinal irradiation (5-10 years later).
- Post-surgical (including cardiac catheterisation).

Less common causes are:

- Other infections.
- Neoplasms.
- Uraemia.
- Connective tissue disorders.
- Drugs.
- Trauma.
- Cardiovascular disease.

Other aetiologies are rare - hereditary, chemical trauma, etc.

Presentation

A history of cardiac surgery or a systemic disease that affects the pericardium makes the diagnosis of constrictive pericarditis more likely\[3\].

Typically, there is a very gradual onset (usually months, occasionally days). The pericardium becomes thickened and fibrotic (and later 'eggshell' calcification may be visible on CXR):

- In the early stages signs are subtle and easily missed.
- In advanced disease the patient may be ill with jaundice, cachexia and muscle wasting.
- Similar to right heart failure, commonly dyspnoea (which may be relatively slight), peripheral oedema, JVP elevated (classically with a prominent y descent (Friedreich's sign), and doesn't fall with inspiration (Kussmaul's sign)).
- Additionally, there may be pulsatile hepatomegaly (in as many as 70% of patients), reduced apical impulse and a pericardial 'knock' (early diastolic sound).
- Pulsus paradoxus, a finding in cardiac tamponade, is uncommon\[2\].
- A patient with intense venous congestion but no heart enlargement or valvular disease, should lead one to suspect constrictive pericarditis.
Differential diagnosis

Chronic constrictive pericarditis
The main differential diagnosis is restrictive cardiomyopathy. Echocardiographic techniques such as speckle-track imaging, velocity vector imaging, as well as cardiac computerised tomography and cardiac MRI can help differentiate constriction from restriction with high sensitivity and specificity.

Others include:

- Dilated cardiomyopathy
- Pericardial effusion
- Cardiac tumours

Chronic effusive pericarditis

- Cardiac tamponade
- Dilated cardiomyopathy
- Constrictive pericarditis
- Constrictive-effusive pericarditis
- Pulmonary oedema

Investigations

- Calcification of the pericardium on CXR strongly suggests constrictive pericarditis in patients with heart failure.
- Echocardiogram (tissue Doppler imaging and colour M-mode echocardiography) is usually diagnostic and helps distinguish from restrictive cardiomyopathy.
- MRI can estimate thickness of the pericardium.
- Cardiac catheterisation is sometimes used to measure pressures.
- CT scans may be performed in some cases, as CT can demonstrate increased pericardial thickness and calcification.
- A pericardial biopsy may be indicated, especially if infective, malignant or granulomatous causes are suspected.
- The heart is not enlarged. ECG may be low voltage with nonspecific T-wave changes.
- Echocardiographic techniques such as tissue Doppler imaging (TDI) and 2D-speckle tracking, dual source CT and also tagged cine MRI with the analysis of phase contrast angiography sequences are all promising novel techniques to improve the diagnosis.

Management

The underlying cause should be treated where possible. Chronic constriction has a definite surgical treatment, whereas transient cases may be reversible with empirical anti-inflammatory therapy.

Chronic effusive pericarditis
Chronic effusive pericarditis can be treated by catheter pericardiocentesis or by surgical drainage and pericardiectomy.

Constrictive pericarditis
Although a significant number of patients will require pericardiectomy, some patients have a predominantly inflammatory and reversible pericardial reaction and may improve with the treatment of the underlying cause and the use of anti-inflammatory medications.

Patients should therefore be observed for the improvement on medical treatments for a period, whenever possible, before proceeding to pericardiectomy.

Although pericardiectomy is the gold standard for chronic constrictive pericarditis, it is still associated with a significant operative mortality of around 10%. As symptoms of constrictive pericarditis may persist after partial pericardiectomy, it is very important that the pericardiectomy is complete, with as much of the pericardium removed as possible.

Prognosis

Long-term survival after pericardiectomy for constrictive pericarditis is related to the underlying aetiology and overall clinical condition of the patient. There is a relatively good survival in patients with idiopathic constrictive pericarditis.

Prognosis is strongly linked to the underlying cause but long-term survival is more likely with surgery and the best results are achieved if surgery is offered early. The surgical results are poor in patients with:

- Organ failure (eg, renal and hepatic particularly).
- Ascites.
- Untreated coronary artery disease.
- Elderly age.
- New York Heart Association (NYHA) class IV heart failure symptoms.
- Post-radiation pericarditis.
- Myocardial fibrosis.
Historical notes

- Richard Lower (notable for the first human blood transfusion in England in 1667) described a patient with dyspnoea and intermittent pulse, in 1669.
- Lancisi (notable for suggesting the connection between malaria and mosquitoes) reported on the constrictive syndrome in 1828.
- Corrigan is credited with the pericardial knock in 1842.
- Franz Volhard collaborated with Viktor Schmieden in 1923, leading to the first pericardiectomy for constrictive pericarditis.

Further reading & references

7. 2015 ESC Guidelines for the diagnosis and management of pericardial diseases: European Society of Cardiology (August 2015).

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