

CLINICAL VIGNETTE

A Case of Asymptomatic Patient with Pericardial Calcification and Constrictive Pericarditis

Andrew Mathew, MD, Ramin Tabibazar, MD and Ravi Dave, MD

A 38-year-old male presented with “fluttering in the chest”, over the past several weeks. His symptoms were sporadic and resolved spontaneously. His physical examination included unremarkable vital signs. He had elevated jugular venous pressure, but no hepatic congestion, peripheral edema, or chest crackles. His resting EKG showed normal sinus rhythm, without ectopy.

The patient underwent transthoracic echocardiogram which showed pericardial calcification and findings concerning for constrictive pericarditis. Cardiac computed tomography confirmed extensive pericardial calcification, and cardiac MRI showed calcified and thickened pericardium with constrictive cardiac physiology. The etiology for pericardial calcification and constrictive pericarditis remained undetermined despite infectious and rheumatologic evaluation. After consultation with CT surgery, the patient deferred surgical intervention at this juncture as he has remained essentially asymptomatic.

Background

Constrictive pericarditis occurs when the pericardial sac becomes inflamed and fibrosed. As the pericardium stiffens, it becomes less capable to adapt to volume changes. This causes greater ventricular interdependence which may lead to cardiovascular symptoms. Constrictive pericarditis is more common in men. The incidence and prevalence have not been well established.¹ Not infrequently, patients with constrictive pericarditis may be undiagnosed or misdiagnosed with other conditions such as congestive heart failure or chronic liver disease.

While tuberculosis is the most common etiology in developing countries, the majority of cases in developed countries are idiopathic or post cardiac surgery.¹ In several large studies, over 40% of cases were eventually classified as idiopathic.²⁻⁵ History of cardiac surgery, trauma, radiation exposure, tuberculosis, viruses, neoplasms, and connective tissue disorders should be considered before concluding an idiopathic etiology. Inaccessibility of the pericardial tissue may pose a diagnostic challenge. Nevertheless, idiopathic constrictive pericarditis is a diagnosis of exclusion after comprehensive evaluation.

While pericardial calcification is not always seen in patients with constrictive pericarditis, its presence is highly suggestive in the correct clinical context. Pericardial calcification is seen in approximately 25%-30% of patients.^{1,2,5,6} These patients are

more likely to have idiopathic pericardial disease, longer duration of symptoms, pericardial knock, larger atria or atrial arrhythmia.⁶ In addition, pericardial calcification is associated with higher perioperative mortality when undergoing corrective surgery.⁶

Diagnosis

Patients may present with a range of clinical symptoms resulting from fluid overload and/or diminished cardiac output. Symptoms may include peripheral edema, ascites, effusions, anasarca, fatigue, chest pain, dyspnea, atrial arrhythmia, and/or abdominal complaints. Elevated jugular venous pressure is seen in the majority of patients, while pulsus paradoxus, Kussmaul's sign, and a pericardial knock are seen less often.³ Congestive hepatopathy has also been observed with a disproportionate elevation in prothrombin time compared to serum bilirubin, suggesting a cardiac etiology.⁷

Initial evaluation with chest radiographs may show pericardial calcifications. The diagnosis is typically made with transthoracic echocardiogram (TTE), showing increased pericardial thickness with or without calcifications. Other findings include abnormal septal bounce with inspiration, in early diastole ventricular septal notching on M Mode tracing, and flattened posterior motion of the posterior left ventricle wall.⁸ Computed tomography can further assess the extent and position of calcifications. Patients with non-diagnostic imaging may undergo cardiac catheterization with classic hemodynamic findings of increased atrial pressures with marked x and y descents, and equalization of end-diastolic pressures with the square root sign.

Some patients may be asymptomatic, presenting with occult constrictive pericarditis on imaging, or initially misdiagnosed with a primary liver disease. These patients should have exercise testing and measurement of their maximal oxygen consumption. Jugular venous pressure should be documented and liver function tests should be monitored. Worsening jugular venous pressure, reduced exercise tolerance, or hepatic insufficiency are all indications for treatment.⁹

Management

Patients with transient pericarditis may respond to a trial of medical treatment with NSAIDs, steroids, diuretics, and other etiology specific medication. Conservative treatment should be

attempted for several months before considering surgery.¹⁰ Patients with signs of chronic constriction including hepatic dysfunction and pericardial calcification benefit from earlier surgical intervention.¹¹ Diuretics can be used as palliative care for patients who are not candidates for surgery. Diuretics can also be used preoperatively in order to optimize the patients' hemodynamically.

Constrictive pericarditis is typically chronic and progressive. Definitive treatment is pericardectomy to remove all thickened pericardium. The majority of patients have a significant improvement in functional status and symptoms. Idiopathic etiologies have a higher five year survival rate of around 80%.^{2,4,5} Older age, post-radiation etiology, and worse NYHA class adversely affect survival.^{2,3,11} Pericardectomy is associated with a high operative mortality rate between 4-8%.²⁻⁵ Patients with mild disease should be treated conservatively to avoid the morbidity and mortality associated with surgery. Similarly, pericardectomy in patients already with advanced disease and a poor prognosis may do more harm than good.²⁻⁵

Patients with calcification are managed similarly, but may require additional procedures. Calcifications are most commonly found on the surface of the diaphragm and on the right ventricle causing myocardial adherence and calcium penetration.⁶ Options to remove the adherent calcified plaques include wedge excision or ultrasonic decalcification. In severe cases, cardiopulmonary bypass or sacrificing the phrenic nerve may be needed.^{1,6,12,13}

Conclusion

Constrictive pericarditis is often idiopathic and can be associated with pericardial calcifications. Along with clinical symptoms of heart failure, diagnosis is typically made with TTE. Despite high perioperative mortality, pericardectomy is the standard of treatment with most patient having significant relief of symptoms. Pericardial calcification is often associated with idiopathic etiologies which have been associated with better survival rates. However, calcifications are also independently associated with increased perioperative mortality.

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