

CLINICAL VIGNETTE

A Case of Asymptomatic Patient with Pericardial Calcification and Constrictive Pericarditis

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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 pericardiectomy 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} Pericardiectomy 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, pericardiectomy 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, pericardiectomy 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.

REFERENCES

1. **Syed FF, Schaff HV, Oh JK.** Constrictive pericarditis--a curable diastolic heart failure. *Nat Rev Cardiol.* 2014 Sep;11(9):530-44. doi: 10.1038/nrcardio.2014.100. Epub 2014 Jul 29. Review. Erratum in: *Nat Rev Cardiol.* 2015 Dec;12(12):682. PubMed PMID: 25072910.
2. **Bertog SC, Thambidorai SK, Parakh K, Schoenhagen P, Ozduran V, Houghtaling PL, Lytle BW, Blackstone EH, Lauer MS, Klein AL.** Constrictive pericarditis: etiology and cause-specific survival after pericardiectomy. *J Am Coll Cardiol.* 2004 Apr 21;43(8):1445-52. PubMed PMID: 15093882.
3. **Ling LH, Oh JK, Schaff HV, Danielson GK, Mahoney DW, Seward JB, Tajik AJ.** Constrictive pericarditis in the modern era: evolving clinical spectrum and impact on

outcome after pericardiectomy. *Circulation.* 1999 Sep 28;100(13):1380-6. PubMed PMID: 10500037.

4. **Szabó G, Schmack B, Bulut C, Soós P, Weymann A, Stadtfeld S, Karck M.** Constrictive pericarditis: risks, aetiologies and outcomes after total pericardiectomy: 24 years of experience. *Eur J Cardiothorac Surg.* 2013 Dec;44(6):1023-8; discussion 1028. doi: 10.1093/ejcts/ezt138. Epub 2013 Jun 12. PubMed PMID: 23761416.
5. **George TJ, Arnaoutakis GJ, Beaty CA, Kilic A, Baumgartner WA, Conte JV.** Contemporary etiologies, risk factors, and outcomes after pericardiectomy. *Ann Thorac Surg.* 2012 Aug;94(2):445-51. doi: 10.1016/j.athoracsur.2012.03.079. Epub 2012 May 22. PubMed PMID: 22621875; PubMed Central PMCID: PMC3610598.
6. **Ling LH, Oh JK, Breen JF, Schaff HV, Danielson GK, Mahoney DW, Seward JB, Tajik AJ.** Calcific constrictive pericarditis: is it still with us? *Ann Intern Med.* 2000 Mar 21;132(6):444-50. Erratum in: *Ann Intern Med* 2000 Oct 17;133(8):659. PubMed PMID: 10733443.
7. **White TJ, Leevy CM, Brusca AM, Gnassi AM.** The liver in congestive heart failure. *Am Heart J.* 1955 Feb;49(2):250-7. PubMed PMID: 13228357.
8. **Engel PJ, Fowler NO, Tei CW, Shah PM, Driedger HJ, Shabetai R, Harbin AD, Franch RH.** M-mode echocardiography in constrictive pericarditis. *J Am Coll Cardiol.* 1985 Aug;6(2):471-4. PubMed PMID: 4019932.
9. **Hoit BD.** Treatment of Pericardial Disease. Cardiovascular Therapeutics: A Companion to Braunwald's Heart Disease, 4th edition. Philadelphia: Elsevier; 2013.
10. **Haley JH, Tajik AJ, Danielson GK, Schaff HV, Mulvagh SL, Oh JK.** Transient constrictive pericarditis: causes and natural history. *J Am Coll Cardiol.* 2004 Jan 21;43(2):271-5. PubMed PMID: 14736448.
11. **Vistarini N, Chen C, Mazine A, Bouchard D, Hebert Y, Carrier M, Cartier R, Demers P, Pellerin M, Perrault LP.** Pericardiectomy for Constrictive Pericarditis: 20 Years of Experience at the Montreal Heart Institute. *Ann Thorac Surg.* 2015 Jul;100(1):107-13. doi: 10.1016/j.athoracsur.2015.02.054. Epub 2015 May 13. PubMed PMID: 25979240.
12. **Ghavidel AA, Gholampour M, Kyavar M, Mirmesdagh Y, Tabatabaie MB.** Constrictive pericarditis treated by surgery. *Tex Heart Inst J.* 2012;39(2):199-205. PubMed PMID: 22740731; PubMed Central PMCID: PMC3384050.
13. **Yetkin U, Kestelli M, Yilik L, Ergunes K, Kanlioglu N, Emrecaan B, Ozbek C, Gurbuz A.** Recent surgical experience in chronic constrictive pericarditis. *Tex Heart Inst J.* 2003;30(1):27-30. PubMed PMID: 12638667; PubMed Central PMCID: PMC152831.

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