Case Report

Recurrent Constrictive Pericarditis after Pericardiectomy: Case Report

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Introduction

Constrictive pericarditis (CP) consists of a chronic inflammation leading to dense fibrosis and adhesion of the pericardial layers, resulting in rigid unmalleable pericardium, with consequent restriction of the diastolic ventricular filling. Possible etiologies for CP are tuberculosis, collagenoses, neoplasias, and heart surgery, which can have different presentation according to location, extension and thickening degree. Tuberculosis is the most common cause of pericardial disease in the world, especially in underdeveloped countries, where it is endemic and it is frequently associated with immunodeficiency. In this report, we describe a case of recurrent CP that occurred several years after incomplete pericardiectomy.

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A 36-year-old male was admitted due to dyspnea on moderate efforts and orthopnea for one year. He had a history of idiopathic CP treated by pericardiectomy 13 years earlier at another service, its limits being the left and right branches of the phrenic nerve, anterolaterally. He denied medical history of tuberculosis, radiotherapy or autoimmune diseases. On physical exam, elevated jugular venous pressure and hepatomegaly were identified, as well as a protodiastolic sound suggestive of pericardial “knock” on cardiac auscultation.

Chest radiography revealed severe calcification of the pericardial layers, as shown in Figure 1. Transthoracic echocardiogram (TTE) showed biatrial enlargement, thickened and calcified pericardium (Figure 2), dilated vena cava with absent inspiratory collapse and preserved e’ velocities (septal e’ 15 cm/sec and lateral e’ 13 cm/sec) suggesting recurrence of the pericardium constriction. Completing the evaluation, the patient underwent computed tomography (CT), which showed severe biatrial enlargement and extensive thickening and calcification of the pericardial sac in its superior, left lateral, and diaphragmatic faces, findings consistent with CP with severe hemodynamic repercussion (Figure 3).

Keywords

Pericarditis, Constrictive; Pericardiectomy; Myocardium/atrophy; Recurrence.

The patient presented good response to the clinical treatment with loop diuretics, becoming asymptomatic and being discharged to outpatient follow-up.

Discussion

The incidence of different etiologies of CP has changed in the past decades, with pericardiectomy outcomes (i.e., survival and recurrence). Although tuberculosis is still the leading cause of CP in developing countries, most cases in developed countries are idiopathic, being possibly related to previous viral pericarditis. In addition, cardiac surgery and radiation therapy have emerged as important causes. Post-surgical and, to a higher extent, post-radiation CP, are associated with poorer outcomes.

Pericardiectomy is the treatment of choice for CP, as liberating the restricted heart improves cardiac function, leading to compensation and, in most cases, to resolution of the symptoms. For a complete pericardiectomy, excision of the pericardium should be done with the phrenic nerves as the posterior extent, the great vessels as the superior extent, and the diaphragmatic surface as the inferior extent. Some authors consider that a truly complete resection should also include the pericardium posterior to the left phrenic nerve, portion that still covers the left ventricle. In both definitions, constricting layers of epicardium are also removed, if technically feasible.

Chowdhury et al., Ling et al., and Bertog et al. found rates for incomplete resection of 14.4%, 11.0%, and 27.0%, respectively. Although technical difficulty of a complete pericardiectomy and resolution of symptoms in many cases after partial resection are among the arguments for favoring the latter, the operative risk for both procedures has been shown to be similar. In addition, partial pericardiectomy is more associated with heart failure relapse, probably due to constriction by the remaining pericardium, and 30-day mortality of a reoperation is higher compared to primary surgery. Thus, total resection should be chosen, considering its decreased mortality, less post-surgical low output syndrome, shorter hospitalization and better long-term survival compared to partial pericardiectomy.

Chowdhury et al. reported 3.7% and 20.9% of recurrence (symptoms NYHA III or IV) for patients who underwent total (phrenic-to-phrenic) and partial pericardiectomy, respectively. On the other hand, Ling et al. found that most recurring patients (symptoms NYHA III or IV) – which corresponded to 1/3 of the entire sample – had undergone total pericardiectomy. In this series, age, ascites, and radiation were identified as independent late predictors of recurrence. Previous radiation therapy is associated with limited pericardial removal, damage to adjacent structures, and neoplasm recurrence, which leads...
to worse outcomes.\textsuperscript{6,7} As well as in previous heart surgery, it also promotes more difficult resection.\textsuperscript{6}

As our patient underwent an incomplete resection, the most probable mechanism for his symptoms was recurrent constriction by the pericardium of the diaphragmatic surface and by the portion posterior to the left phrenic nerve, which were not removed. However, recurrence due to incomplete resection occurs more frequently in the first year.\textsuperscript{9} As it occurred several years after the resection, it is also possible that incomplete pericardiectomy was not the only related factor, and other pathologic processes, such as cardiomyopathy and constriction by surrounding scar tissue, could also be involved.\textsuperscript{9}

As identified in the above-mentioned series, recurrent symptoms can occur after complete pericardiectomy, indicating that constriction is not the only mechanism of its pathophysiology.\textsuperscript{4,6,7} Patients who recur without history of radiation, heart surgery or incomplete pericardium resection have, among the possibly involved causes, myocardial atrophy due to prolonged constriction, myocardial involvement by the same pathological process, and fibrous invasion of the myocardium.\textsuperscript{4,6,7}

### Conclusion

The case described in this report, in association with current evidence in the literature, reinforces the importance of complete pericardiectomy for treatment of CP and the possibility of relapsing symptoms, which can happen even when total resection is achieved. Besides incomplete pericardiectomy, post-surgical or post-radiation resection, as well as myocardial impairment, due to different factors, can be related to the recurrence of symptoms.
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Figure 3 – Computed tomography of the chest showing biatrial enlargement, ventricular narrowing, and pericardial thickening and calcification (arrow).

Authors’ contribution

Research development and design: Holanda AC, Santos ECL, Markman Filho B, Lima SG, Lordsleem ABMS, Pedrosa FMQ;
Data acquisition: Holanda AC, Santos ECL, Markman Filho B, Lima SG, Lordsleem ABMS, Pedrosa FMQ;
Data analysis and interpretation: Holanda AC, Santos ECL, Markman Filho B, Lima SG, Lordsleem ABMS, Pedrosa FMQ; Manuscript drafting: Holanda AC, Santos ECL, Markman Filho B, Lima SG, Lordsleem ABMS, Pedrosa FMQ; Critical revision for important intellectual content: Holanda AC, Santos ECL, Markman Filho B, Lima SG, Lordsleem ABMS, Pedrosa FMQ.

References


Potential Conflicts of Interests

There are no relevant conflicts of interests.

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