CASE REPORT Fibrous Pericardial Mass Signifying the Importance of Advanced Cardiovascular Imaging: A Case Report

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Background: Pericardial masses are an extremely rare group of diseases which can be classified based on etiology. The presentation of pericardial masses varies considerably from one individual to another, ranging from an asymptomatic presentation with an incidental finding on imaging, to presenting with non-specific signs and symptoms. Due to the enigmatic nature and presentation of pericardial masses, diagnostic imaging is mandatory.

Case Presentation: A 69-year-old patient presented to our cardiology clinic complaining of intermittent shortness of breath upon moderate exertion in the absence of chest pain, paroxysmal nocturnal dyspnea, orthopnea, dizziness, palpitations, or lower limb edema. The patient's past medical history was significant because of his history of pericarditis associated with pericardial effusion 6 years prior to presentation at our clinic. Despite adequate medical treatment, the patient complained of a relapsing and remitting pattern of symptoms that mandated the performance of advanced cardiovascular imaging, namely, cardiac magnetic resonance imaging, which revealed the presence of a profound pericardial mass.

Conclusion: Despite the fact that relapsing pericarditis is a well-established complication following acute pericarditis, the presentation of a complication such as a fibrous pericardial mass evident on cardiac MRI has essentially been unreported in the literature previously. Keywords: pericardial mass, cardiovascular imaging, cardiac magnetic resonance imaging

Introduction

Pericardial masses are an extremely rare group of diseases which can be classified based on etiology into neoplastic, which is further classified into primary and secondary, and non-neoplastic, such as thrombus and simple cysts.¹ The presentation of pericardial masses varies considerably from one individual to another, ranging from an asymptomatic presentation with an incidental finding on imaging, to presenting with non-specific signs and symptoms such as chest pain and dyspnea mimicking other pathologies of pericardial tissue, including pericarditis, pericardial effusion and cardiac tamponade.²

Due to the enigmatic nature and presentation of pericardial masses, diagnostic imaging is mandatory. Initial diagnostic imaging evaluation typically involves chest radiographs and transthoracic echocardiography (TTE). However, further assessment with cardiac magnetic resonance imaging (CMR) is of significance due to its superiority in visualizing and detecting pericardial masses in comparison to TTE.³ Moreover, the importance of CMR lies in its ability to characterize a pericardial mass, providing insights on the composition of the mass and potential management options,^{4,5}

We report a rare case of a 69-year-old male patient, with a previous history of pericardial effusion several years prior to his presentation, presenting with nonspecific clinical features that led to a journey of clinical assessment, evaluations, and management until the diagnosis of a fibrous pericardial mass was eventually established.

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Case Presentation

A 69-year-old male patient presented to our cardiology clinic complaining of intermittent shortness of breath upon moderate exertion in the absence of chest pain, paroxysmal nocturnal dyspnea, orthopnea, dizziness, palpitations, or lower limb edema for a couple of months. The patient's past medical history was significant because of a history of pericarditis associated with pericardial effusion 6 years prior to his presentation at our clinic, which was resolved with appropriate pharmacological treatment consisting of NSAIDs and colchicine and of which he had not complained since. Also, he had an anterior wall myocardial infarction dating to several years ago, which required immediate intervention and stenting to his left anterior descending artery. Additionally, the patient is a heavy smoker and is diabetic, and hypertensive, both conditions are under pharmacological intervention and well-controlled.

At first visit, the patient's results were as follows: blood pressure 131/67 mmHg, heart rate 85 bpm, respiratory rate 18bpm, oxygen saturation rate 95%, and oral temperature 36.5 C. A cardiopulmonary examination showed normal S1 and S2 with no murmurs and clear lung fields to auscultation bilaterally. Laboratory tests included negative outcomes for tuberculosis, ANA, and rheumatoid factor double-stranded DNA thus making any rheumatological disease less likely. ECG showed a sinus rhythm with q waves in the anteroseptal leads. Subsequently a transthoracic echocardiogram (TTE) was performed and revealed an ejection fraction of 60%, no wall motion abnormality and mild left ventricular hypertrophy. Additional findings noted were grade 1 diastolic dysfunction, moderate aortic stenosis, mild aortic regurgitation and a moderate pericardial effusion moderately circumferential with an anterior pocket of 1.8–2.4 cm with no evidence of cardiac tamponade physiology. At that point, the patient was prescribed colchicine for 3 months and NSAIDS for 14 days with a follow-up visit. Then the patient improved, and his pericardial effusion improved.

Three months following his initial presentation, the patient started complaining again of occasional symptoms of chest pain not related to physical activity or stress. However, the patient reported that his shortness of breath had diminished with time. Based on this clinical picture, the possibility of coronary artery syndrome was suspected and needed to be ruled out. The patient underwent a pharmacological myocardial perfusion stress and rest scan. The test showed no significant evidence of ischemia and a subtle decrease uptake in the distal anteroseptal wall, which represents apical thinning rather than a true perfusion defect. The ECG portion of the study was normal. The patient was treated with another course of colchicine and steroids.

Six months later, the patient presented with a third relapse of shortness of breath. Upon inquiring further, the patient revealed that he had, unfortunately, discontinued the colchicine prior to completing the intended period of six months. TTE showed relapse of pericardial effusion and an echo-dense lesion in the pericardium of the anterior wall, which was more prominent than had been seen on the previous echo. This led to performing a CMR, which revealed an echo-dense lesion in the pericardium of the anterior wall between the right atrium and the right ventricle. It was accompanied by moderate pericardial effusion consisting of multiple layered fibrous bands that were more pronounced at the right ventricle, with a maximum thickness of 26mm and the appearances of fibrotic pericarditis (non-constrictive pericarditis) with delayed enhancement (Figure 1). Accordingly, the diagnosis of a fibrous pericardial mass was established. The patient was treated with a prolonged course of colchicine.

Discussion

One of the complications of acute pericarditis is the development of relapsing or resistant pericarditis that generally affects 15–30% of individuals following an initial episode of acute pericarditis.⁶ Typically, the clinical picture of relapsing pericarditis involves two forms known as the recurrent and incessant types. Recurrent pericarditis is defined by the presence of a period of partial or complete resolution of symptoms following an acute episode of pericarditis usually lasting 4–6 weeks, whereas the persistence of clinical features with the absence of a definite symptomatology timeline is referred to as incessant pericarditis.^{6,7} The exact underlying pathophysiology of relapsing pericarditis remains unknown despite the proposition of several mechanisms. The presence of anti-cardiac antibodies alongside other non-organ-specific autoantibodies, such as anti-intercalated disk autoantibodies in patients with a history of relapsing pericarditis indicating an immunological role, constructs the basis of the current leading pathophysiologic

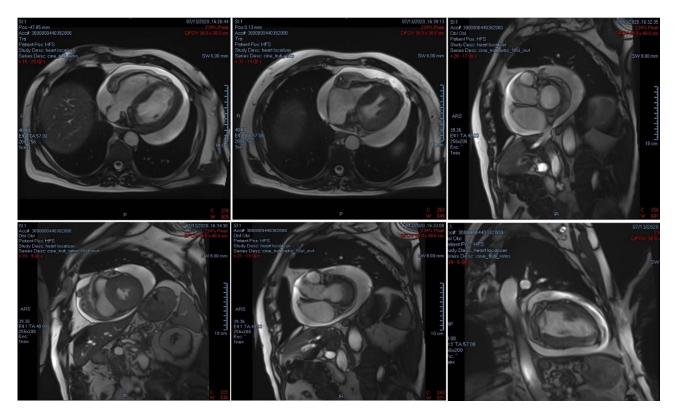


Figure I Cardiac magnetic resonance imaging in cine T2 sequences demonstrating the pericardial mass in different slices.

mechanism.⁸ Other suggested mechanisms involve the presence of a concurrent systemic disease such as malignancy,⁸ abrupt cessation of therapeutic interventions prior to complete resolution of acute pericarditis, the occurrence of a viral infection,⁹ or the presence of an underlying autoinflammatory disorder, particularly Familial Mediterranean Fever (FMF) and Tumor Necrosis Factor Receptor-Associated Periodic Syndrome (TRAPS).⁷

The diagnosis of pericardial masses is highly dependent on different cardiac imaging modalities. However, despite the fact that TTE is the standard tool of evaluation in patients presenting with pericardial pathologies, TTE alone can be unreliable in reaching a diagnosis in some cases due to some of its limitations.^{4,10} Alternatively, CMR is considered an excellent and reliable method to assess the pericardium in the absence of unwarranted exposure to radiation.

CMR demonstrates its superiority in narrowing differential diagnoses through its ability to specify and differentiate between smooth, thickened pericardium (which suggests the presence of acute or subacute pericarditis) and thickened irregular pericardium (suggestive of chronic pericarditis, pericardial fibrosis, or pericardial mass).⁴ Additionally, CMR provides essential information on the assessment of any presenting pericardial effusion, in both its static imaging (that evaluates the anatomy) and cine imaging modality (which studies cardiac motions), and is widely regarded to be superior to echocardiograms in outlining the character of the effusion.^{11,12} CMR is also especially necessary in evaluating localized/loculated pericardial effusions and in the discrimination of small effusions versus focal pericardial thickening.

CMR is also an extremely important imaging modality for evaluating pericardial masses, not only for confirmation of presence or absence of a mass but also because it is able to provide precise tissue characterization and mass vascularization assessment, ascertain the size of the mass, and distinguish between benign and malignant masses.^{13,14} Moreover, by using different sequences, CMR provides a framework to characterize tissue composition in addition to perfusion assessment of a pericardial mass, which helps in differentiating types of pericardial masses. Utilization of the T1 and T2 weighted images together with late gadolinium enhancement has been proven to precisely define some pericardial tumors.¹⁵ Furthermore, novel techniques such as native T1 mapping along with T2 mapping sequences are also capable of differentiating fibrous containing masses from thrombus and fat.¹⁶

Pericardial fibromas are usually characterized by a lack of complex vascularization, and accordingly are defined by hypointense signals on T1 and T2 weighted images.¹⁷

Relapsing pericarditis, whether in the recurrent or incessant form, is generally associated with a promising long-term prognosis.¹⁸ The occurrence of complications in relapsing pericarditis remains rare. One complication of significance, despite its extreme rarity, is the development of tamponade, as demonstrated by Robinson and Bridgen; in their study, 2 out of 33 patients developed tamponade.¹⁹ Despite the fact that constrictive pericarditis is a well-documented rare complication of acute pericarditis, it has not been documented in relapsing pericarditis.^{19,20} Similarly, the involvement of the myocardium and subsequent development of myocardial disease has not been reported in relapsing pericarditis.¹⁸

The mainstay treatment for relapsing pericarditis involves conservative measures consisting mainly of restriction of physical activity, and pharmacological interventions involving the combinational use of aspirin or NSAID and colchicine, with evidence indicating that the use of colchicine significantly reduces further recurrence.^{21,22} In patients with clinical presentations that require rapid symptomatologic control, corticosteroids, typically prednisone, are used.²³ However, corticosteroids are associated with a significant side effect profile in comparison to NSAIDs and colchicine, and there is a higher possibility of recurrence due to the likely use of inadequate dosage or rapid tapering resulting in corticosteroid dependence.^{7,22} In patients who are resistant to colchicine and are corticosteroid dependent, there is increasing evidence of the use of anakinra, which is an interleukin 1 β recombinant receptor antagonist.²⁴ Brucato et al²⁴ conducted a randomized controlled trial that demonstrated the efficacy of anakinra in preventing recurrent episodes of pericarditis; only 2 out of 11 patients who received anakinra experienced recurrence, in contrast to the placebo group in which 9 out of 10 patients experienced recurrence. Nonetheless, in the presence of severe symptomatic episodes of relapsing pericarditis that is resistant to the various therapeutic approaches, pericardiectomy is warranted.²² However, there is evidence of recurrence following pericardiectomy due to the partial removal of the pericardium,²² thus creating doubt regarding the efficacy of the procedure.

Conclusion

Although relapsing pericarditis is a well-established complication following acute pericarditis, the presentation of a complication such as a fibrous pericardial mass evident on cardiac MRI has essentially been unreported in the literature thus far. Accordingly, our case highlights the importance of considering cardiac MRI in cases of failure of resolution of symptomatology in patients with acute pericarditis, or, in certain instances, exacerbation of presenting features.

Statement of Ethics and Informed Consent

Written informed consent was obtained from the patient, including for the publication of the paper and its content. Institutional approval was not required for the publication of this work.

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Disclosure

The authors report no conflicts of interest in this work.

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