Complicated Pericardial Cyst: Atypical Anatomy and Clinical Course

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Summary: Pericardial cysts are usually detected by chance and are clinically silent in most cases. Nevertheless, symptoms and serious complications may occur. We describe a case of pericardial cyst diagnosed in an 8-year-old boy who was admitted with chest pain. Echocardiography revealed a mild to moderate pericardial effusion and a 7.5×5.5 cm intrapericardial echo-free lesion consistent with a pericardial cyst. Surgery was carried out 3 days afterward because of the patient's worsening condition, the progressive increase of pericardial effusion, and the onset of initial signs of cardiac tamponade. The cyst showed a long and easily movable vascular pedicle and inflammatory areas involving the pericardial surface. Like the pericardial effusion, the contents of the mass appeared as serosanguineous fluid on aspiration. Histologic examination confirmed the diagnosis of pericardial cyst and showed findings according to ischemia-related lesions of the cyst. The coexistence of pericardial cyst and cardiac tamponade is very unusual. The atypical anatomy and clinical course suggest a distinct and so far undescribed pathogenetic mechanism for this association: the torsion of a vascular pedicle and the subsequent development of ischemia-related lesions of the cyst.

Key words: pericardial cyst, cardiac tamponade, echocardiography

Introduction

Pericardial cysts are the most common primary tumors of the pericardium and account for 7% of all mediastinal tumors.^{1,2} The etiology is usually congenital for abnormal development of mesenchymal tissue during the embryogenic state, so that a portion of the parietal pericardium is pinched off and disconnected from the pericardial sac. The usual site of pericardial cysts is the cardiophrenic right angle (51–70%), but they can also be localized in the mediastinum, namely, in the left cardiophrenic angle (22–38%) or superior mediastinum (8–11%) in the paratracheal area or near the aortic arch.^{3–5} The cysts appear as round or elliptic masses, with thin and smooth walls; most of them are unilocular, often with trabeculated internal lining. In the absence of superimposed phlogistic process, the fluid content is a clear transudate. Their anatomic structure, like the pericardium, consists of collagen and scattered elastic fibers lined by a layer of mesothelial cells; foci of mesothelial hyperplasia and an area of calcification and chronic inflammation have been found. Pericardial cysts are usually asymptomatic and appear as homogeneous radiodensity on a chest x-ray performed routinely or for other clinical suspicions. Symptoms are described in about one third of the cases and include chest pain, dyspnea, and coughing;⁴ spontaneous rupture and cardiac tamponade have been reported occasionally.^{6–8}

We describe an uncommon case of pedunculated pericardial cyst with pericardial effusion and clinical signs of cardiac tamponade.

Case Report

An 8-year-old boy was admitted to the emergency room for recurrent episodes of chest pain appearing the day before admission. He had no previous history of cardiac disease. At the age of 5 years, a chest x-ray performed for asthma was normal. At the age of 7 years, he presented an episode of intense chest pain with fainting during physical activity. Further electrocardiographic (ECG) and laboratory tests were normal. On

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FIG. 1 Two-dimensional echocardiography. (A) Apical four-chamber view, (B) parasternal short-axis view at papillary muscles level. LA = left atrium, LV = left ventricle, PE = pericardial effusion, PM = pericardial mass, RA = right atrium, RV = right ventricle.

admission, physical examination was unremarkable except for muffled cardiac tones and tachycardia. The ECG showed sinus tachycardia (110 beats/min), and a slight ST-segment elevation was present in both inferior and anterior leads. Two-dimensional transthoracic echocardiography (Fig. 1) revealed a mild to moderate pericardial effusion and a 7.5×5.5 cm intrapericardial echo-free lesion adjacent to the left ventricular free wall and consistent with a biloculate cystic structure. Other echocardiographic findings were normal, as were routine laboratory tests. The patient was then transferred to the Department of Cardiovascular Surgery to confirm the diagnosis and decide the treatment. At magnetic resonance examination (Fig. 2), a pericardial mass adjacent to the diaphragm further down and to the obtuse cardiac margin medially was confirmed. The mass was connected with the pericardium by a long pedicle originating in the upper mediastinum between the left pulmonary artery and the inferior side of the aortic arch. The upper margin of the mass was not well defined from the left branch of the pulmonary artery. Pericardial effusion was confirmed and a progressive increase in volume was shown by daily sonographic controls.

Surgery was carried out three days after hospitalization due to the patient's worsening condition and the onset of initial signs of cardiac tamponade. After median thoracotomy, 400 ml of serosanguineous fluid were aspirated from the pericardial sac; a nonadherent, well-encapsulated, biloculate tumor, 9×10 cm in size, situated on the left ventricular obtuse margin appeared. It was connected to the inferior side of the aortic arch by a long, thin, and easily movable vascular pedicle. The wall of the mass showed both necrotic and inflammatory reaction extending to the whole visible epicardial surface. Like the pericardial effusion, the contents of the mass appeared as serosanguineous fluid on aspiration. The vascular pedicle was then ligated as close as possible to its insertion and the mass was resected. Histologic examination confirmed the diagnosis of pericardial cyst with no evidence of malignancy or different tissue other than pericardium. Mesothelial cells with below

granulation tissue, fibrin, and focal hemorrhage were found according to ischemia-related lesions. Cultures of both cystic and pericardial fluids were negative.

The subsequent hospital course was uneventful and the patient was discharged on Day 8. Diuretic therapy was given at discharge after an almost complete resolution of the pericardial effusion. Five weeks later, a new hospital admission was necessary for the appearance of interscapular pain emphasized when lying on the left side. High temperature was also present. Echocardiography again showed mild pericardial effusion with normal cardiac function. Symptoms were ascribed to a postpericardiotomy syndrome. Antiphlogistic therapy was started, resulting in complete recovery within a few days.

Serial echocardiograms and chest radiograms over a 4-year follow-up showed no evidence of recurrence, and at present the patient is asymptomatic.

FIG. 2 Magnetic resonance imaging in the coronal plane. The arrow indicates the cyst pedicle. Ao = ascending aorta, LV = left ventricle, PA = left branch of pulmonary artery, PM = pericardial mass.

Discussion

Pericardial cysts have a benign course in the majority of cases, but serious complications may occur related to enlargement of the cyst, including compression of the heart with hemodynamic deteriorations,^{9, 10} airway obstructions,¹¹ and erosion of cardiac wall and vascular structures.^{12, 13} Secondary infections may develop an inflammatory process spreading from the cyst wall to the surrounding tissues; one case of sudden death due to necrotic damage of the myocardium involving the conduction system has been reported.¹⁴ It is widely accepted that pericardial cysts may remain asymptomatic in the majority of cases; spontaneous resolution is unusual, although two cases of presumed spontaneous resolution have been reported.^{6, 7}

We describe a patient with a pedunculated pericardial cyst who developed cardiac tamponade. Only one other case of pericardial cyst complicated by cardiac tamponade has been reported.⁸ These authors hypothesized that a small patent communication between the cyst and the pericardium caused pericardial effusion. Later, this connection spontaneously sealed itself off by obliterative fibrosis. As in our patient, the same serosanguineous fluid was found both in the pericardium and inside the cyst.

In our case, the distinct clinical presentation and the anatomical characteristics of the cyst suggest a different explanation of this rare association of cardiac tamponade with a pericardial cyst. We hypothesize that a sudden torsion of the long and easily movable cyst pedicle observed at surgery produced ischemia-related cyst lesions with a subsequent considerable increase in cyst size and the appearance of pericardial effusion quickly developing into cardiac tamponade. The possibility of a reversible torsion with transient ischemia of the cyst could also explain the episode of intense chest pain with lipothymia experienced about 1 year previously. One case of twisted pedunculated pleuropericardial cyst has also been reported.¹⁵ As in our patient, the clinical presentation was an acute and intense chest pain and the content of the cyst was hemorrhagic fluid. No pericardial effusion was described.

Our patient underwent thoracotomy for the sudden appearance of cardiac tamponade and because the pericardial effusion suggested the presence of malignancy; in such conditions a radical intervention was thought to be more appropriate. Surgical treatment for every pericardial cyst is still controversial; the surgical removal of asymptomatic and proven pericardial cyst is probably unjustified. In selected nonurgent cases, video-assisted thoracoscopy may represent a valid alternative to thoracotomy, especially in high-risk surgical patients.^{16, 17}

Conclusion

We report a very unusual case of pericardial cyst complicated by pericardial effusion and cardiac tamponade. The presence of a long and easily movable vascular pedicle and ischemia-related lesions of the cyst suggests that torsion could be the pathogenetic mechanism causing this rare complication.

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