Heart Hydatid Cyst Close to the Left Descending Artery in a Thirteen-Year-Old Boy

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Introduction: Hydatid cyst is a significant health problem in underdeveloped and developing countries, particularly among sheep breeders. Although cardiac involvement is seen only in 0.2% to 3% of the cases, early diagnosis and treatment are important.

Case Presentation: A 13-year-old boy with dyspnea and atypical pericardial type chest pain for three months was referred to us. Chest X-ray revealed linear calcification in the left side of the heart. Computed tomography demonstrated a cyst with peripheral calcification and without internal septation in the lateral left ventricle (LV) myocardium. Serologic IgG test was positive for Echinococcosis. No other cyst was seen in the other organs such as the lungs and liver. By midline sternotomy on pump, an incision was made 2-cm lateral to the left ascending artery through the LV myocardium and without entering any cardiac chamber. After injecting hypertonic (5%) saline, the cyst was punctured and its fluid contents were aspirated, the cyst was enucleated, and the cavity marsupialization was done for protection of the myocardium. LV ejection fraction before operation was 40% but after operation and repairing LV myocardium, ejection fraction increased to 50% in. Histopathologic examination confirmed a hydatid cyst.

Conclusions: In myocardial hydatid cysts, we recommend a direct approach without entering the cardiac chambers to avoid dissemination of the infection. We recommend excision of the germinative membrane without capitonnage to avoid impairment of myocardial contraction.

Keywords: Echinococcosis; Myocardium; Heart; Cysts

1. Introduction

Hydatid cyst is a significant health problem in underdeveloped and developing countries, particularly among sheep breeders (1). Hydatidosis is a parasitic infection caused by the larval stage of *Echinococcus granulosus* (1). The liver (70%) and lungs (25%) are the most affected organs (2). Although cardiac involvement is seen only in 0.2% to 3% of cases, early diagnosis and treatment are important (1). Isolated cardiac involvement can occur but is extremely rare. Hydatid infection of the heart may occur via coronary circulation or from intravascular pulmonary cyst rupture (2). The clinical signs and symptoms of cardiac hydatid cyst are nonspecific and highly varied; therefore, this disease may be difficult to diagnose. Although echinococcosis is a serious health issue in some geographical regions of the world, cardiac involvement is rare (3). The mostly affected sites of the heart are the left ventricle (LV) (75%), right ventricle (RV) (18%), and the interventricular septum, whereas the pericardium as well as left and right atria are least affected. In primary

cardiac hydatidosis, larvae usually reach the myocardium through the coronary circulation (1).

2. Case Presentation

A 13-year-old boy who had no remarkable medical history was admitted to Baqiyatallah Hospital, Tehran, Iran, on December 2012 because of dyspnea and atypical pericardial type chest pain. Three months before admission, he was referred to us without fever, angina-like chest pain, or palpitation. Cardiac examination and laboratory data (including blood chemistries, cardiac enzymes) yielded normal findings. His erythrocyte sedimentation rate was 22 mm/h and complete blood count showed no significant eosinophilia. Chest X-ray revealed increased cardiothoracic ratio with linear sharp calcification in left heart border (Table 1) (Figure 1).

Physical examination findings were not indicative of disease. Electrocardiographic (ECG) findings were normal. The left ventricular ejection fraction was 40%.

Abdominal and thoracic computed tomography (CT) scans ruled out involvement of other locations (Figure 2). Transthoracic and trans esophageal echocardiography revealed a large (73 mm × 53 mm) encapsulated, intramural, heterogeneous well-defined mass with multiple small echolucency, outer wall calcification (77 mm × 53 mm) in the anterolateral part of the LV at middle and apical segments with protrusion to LV cavity, and severely decreased LV volume and diameter. There was mild central mitral regurgitation and mild to moderate pericardial effusion with systolic RV inversion without diastolic RV collapse.

Thoracic CT revealed a cystic formation of 8-cm × 6-cm dimensions in the same location as shown by chest X-ray and echocardiography. Cardiac MRI (dynamic study with gadolinium) showed a large (75 mm × 54 mm), well-demarcated intramyocardial mass with cystic or necrotic center in the anterolateral wall of LV with severe protrusion to LV cavity; post-gadolinium enhancement at the peripheral aspect of the mass showed capsular enhancement (we used ruler with CM measurement cyst and use CTS criteria for measure). MRI showed no extra-cardiac involvement. This finding was consistent with a cardiac hydatid cyst (Figure 3). Serologic test was positive for echinococcus with high titer. Prior to surgery for cyst

excision, the patient received albendazole for two weeks in order to decrease the number of living parasites.

Table 1. Patient and Cyst Characteristics

Age	Gender	Sign and Symptom	Chest X-Ray	Cyst Size, cm
13	Male	Dyspnea, atypical pericardial chest pain	Calcification	7.7 × 5.3

Figure 1. Chest X-ray

Chest X-ray revealed increased cardiothoracic ratio with linear sharp calcification in left heart border.

Figure 2. Abdominal and Thoracic Computed Tomography Scans

Abdominal and thoracic computed tomography scans ruled out involvement of other locations by disease.

Figure 3. Cardiac Hydatid Cyst. The Finding Was Consistent With a Cardiac Hydatid Cyst

Figure 4. After injecting hypertonic (5%) saline, the cyst was punctured and its fluid contents were aspirated, enucleating and cyst cavity marsupialization were performed.

Figure 5. Removal of the cyst improved myocardial compliance and myocardial perfusion, corrected the cardiac deformity, and strengthened myocardial contraction.

Via midline sternotomy on pump, a 5-cm incision was made on LV myocardium, lateral to LAD and without entering cardiac chambers, to excise the cyst. After inject-

ing hypertonic (5%) saline, the cyst was punctured and its fluid contents were aspirated. The cyst was enucleated and its cavity marsupialization was done (Figure 4). Removal of the cyst improved myocardial compliance and myocardial perfusion, corrected the cardiac deformity, and strengthened myocardial contraction (Figure 5). The patient received albendazole for one month to avoid recurrence. After two years, follow-up transthoracic echocardiography showed EF of 50% with no evidence of recurrence and decreased mitral regurgitation.

3. Discussion

In deciding upon an operative technique, the location of the cyst is of great importance. Most cysts are in the ventricular myocardium. In this reported case, we excised the germinative membrane through the myocardium without entering the cardiac chambers. Postoperative echocardiographic findings indicated improved myocardium function and an ejection fraction of 40% with mild increase in mitral regurgitation. Cardiac involvement can occur from the systemic or pulmonary circulation of parasite or direct extension from adjacent involved structures. It can occur in any part of the heart and the manifestations depend on the size, location, and integrity of the cyst. The LV is the most common site (75%), followed by the RV (15%), interventricular septum (5%-9%), left atrium (8%), pericardium (8%), pulmonary artery (7%), and right atrium (3%-4%) (3). In a study, 15 patients with cardiac hydatid cyst were followed up from ten months to 22 years. There were four recurrences of myocardial cysts after 12 months (3).

In this report, calcification of the cystic lesion in chest X-ray was the main key to diagnosis. Although the symptoms associated with echinococcosis are nonspecific, it may be indicated by an abnormal cardiac silhouette on a chest radiograph, or by echocardiographic findings of cardiac chamber deformation. Few serious consequences were encountered from obstruction of the cardiac valves by a cyst, or from compression of the conduction system or great veins. Major surgical complications from rupture of cyst were seen including systemic or pulmonary embolization, pericardial dissemination, purulent inflammation, and sepsis. These complications of surgical treatment account for a greater mortality rate than anaphylaxis and heart failure do (3). Surgical treatment of cardiac echinococcosis is urgent (3). Most patients with cardiac echinococcosis have no symptoms and the disease is often latent because of slow growth of hydatid cyst in the heart. Unless a cyst is located in a critical anatomic site, the disease is usually diagnosed late. Signs and symptoms of cardiac hydatid cysts are extremely variable and are directly related to the location and the size of the cysts. Approximately 10% of patients, especially those with large hydatid cysts, have clinical manifestations. Precordial pain is the most common symptom and is most often vague and does not resemble angina pectoris. Precordial pain also can be of the pericardial type,

especially in patients with a pericardial hydatid cyst and a pericardial reaction such as in our patient. Sudden rupture of intracardiac cysts is a frightful complication and can cause acute pericarditis or tamponade, acute pulmonary hypertension by embolization of several scolices, systemic arterial embolization, and severe anaphylactic shock. All complications can be life threatening (4).

A variety of intracardiac tumors and a congenital pericardial cyst must be considered in the differential diagnosis; the cysts may be single or multiple, uniloculated or multiloculated, and thin-or thick-walled. More specific signs include calcification of the cyst wall (Figure 1), presence of daughter cysts, and membrane detachment. The mass becomes solid and can be difficult to differentiate from heart tumors (4). Surgery remains the treatment of choice in the management of hydatid disease. Although anthelmintic drugs have been used in the preoperative and postoperative periods since 1977, excision of the lesion under cardiopulmonary bypass is recommended (4).

Heart involvement can result in many clinical features. Precordial pain and dyspnea are frequent revealing symptoms, but patients sometimes present with life-threatening conditions such as congestive heart failure, pericardial tamponade, pulmonary embolism, syncopal attacks, or superior vena cava syndrome. The combination of imaging and serology (enzyme-linked immunosorbent assay) usually provides an effective diagnosis, although sensitivity is quite low. Transthoracic and transesophageal echocardiography play a central role in diagnosing cardiac hydatid cysts. Nuclear MRI is helpful in determining extracardiac extensions of the cysts (5). Because recurrence may appear much later, such a monitoring should be pursued for at least three years (5). Albendazole is active against the germinal membrane of the cyst wall but its concentrations in cyst fluid may be suboptimal. In contrast, praziquantel reaches adequate levels in cyst fluid but is not active against the germinal

membrane at usual dosages. Some evidence suggests pre-operative combination chemotherapy with both agents to be more effective than albendazole alone. However, because of its activity against the germinal membrane, albendazole may cause cysts to leak or rupture. Therefore, in this case we did not pretreat patient with albendazole for a prolonged period (6). Perhaps, weak point of this report is reporting only one case but it was a very large cyst in lateral wall of LV and close to coronary vessel without rupture and involvement of heart chamber, which would be a strong and specific point of this research.

In conclusion, the reported case was of particular interest because it enabled a rare preliminary diagnosis of cardiac echinococcosis by Chest X-ray. In addition, technique of excision has been never reported before in connection with this entity. In myocardial hydatid cysts, we recommend a direct approach without entering the cardiac chambers in order to avoid dissemination of infection and cyst materials. We also recommend excision of the germinative membrane without capitonnage to avoid impairment of myocardial contraction.

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