



Case Report

Multiple hydatid cystectomy of the heart necessitating LIMA to LAD anastomosis in a young patient

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Abstract

Cardiac hydatid disease is very rare, even in endemic regions. Clinical manifestations included chest pain, anaphylactic shock, constrictive pericarditis, congestive heart failure, and arterial embolism. Surgery is the exclusive therapy, where the cysts are excised during open-heart surgery. The surgical approach therefore must be performed carefully, given the potential complications that surgery may bring. Because of the risk of potentially lethal complications, early diagnosis and definitive treatment are important. A 32-year-old male patient was admitted with chest pain, weight loss, lethargy, and dizziness. On the transesophageal echocardiography study, a cystic mass (2.5×3×4.5 cm in dimension adjacent to the left ventricular posterior wall) that was divided into two by a septum was noted. Diagnosis of hydatidosis was confirmed with serologic tests (ELISA and indirect immunofluorescence). Echinococcosis, also known as hydatid disease, is common in several regions of the world, for example, the Mediterranean countries, the Middle East, South America, and East Africa. While performing pericystectomy in the anterior left ventricular wall, we noticed that there were three cysts, contrary to the preoperative diagnosis pointing a single one, and it was impossible to effectively complete the procedure without compromising anterosuperiorly displaced left anterior descending artery (LAD). We decided to go on bypass, arrest the heart, and complete the pericystectomy at the cost of injuring LAD and grafting the left internal mammary artery to LAD. Microscopic examination of the cyst showed a germinal layer and an avascular, eosinophilic, chitinous layer that confirmed the diagnosis of hydatid cyst. The patient was discharged on the fifth postoperative day on albendazole medication. © 2007 Published by Elsevier Inc.

Keywords: Hydatid disease; Coronary artery bypass grafting; Cystectomy; Echinococcus

1. Introduction

Echinococcosis, also known as hydatid disease, is common in several regions of the world, for example, the Mediterranean countries, the Middle East, South America, East Africa, and some areas in Canada and Australia [1,2]. The incidence and prevalence of this disease are estimated to be high in countries where sheep farming is widespread such as Turkey. Echinococcosis is a human infection caused by

the larval stage of *Echinococcus granulosus*. The liver and the lungs are the most common sites of infection [3,4].

Cardiac cyst hydatid is a rare pathology and is seen in 0.5–2% of echinococcal infections [1]. A total of 77% of large cysts and 50% of medium-sized cysts are symptomatic [1]. It is therefore legitimate to state that the larger or more numerous the cysts are (covering a large surface of the myocardium), the more important is their effect on the functioning of the heart and thereby the severity of the symptoms and signs. We report a patient with multiple cysts adjacent to the myocardium, which were formerly diagnosed as a solitary cyst associated with superior displacement of the left anterior descending artery (LAD) with compression. This condition necessitated a LAD–left internal mammary artery (LIMA) bypass graft in our case.

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2. Case report

A 32-year-old male patient was admitted with chest pain, weight loss, lethargy, and dizziness. On the transesophageal echocardiography (TEE) study, a cystic mass (2.5×3×4.5 cm in dimension adjacent to the left ventricular posterior wall) that was divided into two by a septum was noted (Fig. 1).

TEE revealed the same findings. An MRI scan, which is a more reliable diagnostic procedure than the computed tomography (CT) scan for diagnosing cardiac hydatid cyst (CHC), was performed, and it revealed an apical left ventricular cyst with a dimension of 5×4.5 cm and noted pericardial thickenings that were anterosuperiorly and anteroinferiorly located with dimensions of 3.5×2 and 2.5×2 cm, respectively (Fig. 2).

Diagnosis of hydatidosis was confirmed with serologic tests (ELISA and indirect immunofluorescence).

After a 2-month treatment with albendazole, he was referred for surgery for the removal of the cyst within the heart that did not regress with medical therapy.

Cardiopulmonary bypass (CPB) was started; the heart was emptied, making exploration for apically and posteriorly located cysts easier (Fig. 3).

The cysts were then punctured and emptied with a syringe; they were sterilized by injecting a hypertonic saline solution. After a brief pause, the cysts were incised and emptied of their content. While performing pericystectomy in the anterior left ventricular wall, we noticed that it was impossible to effectively complete the procedure with effacement of the residual cavity without compromising the anterosuperiorly displaced LAD (Fig. 4). We decided to go on bypass, arrest the heart, and complete the pericystectomy and effacement of the cavity at the cost of injuring LAD and grafting LIMA to LAD. The residual cavities were closed by 3-0 polypropylene sutures.

Microscopic examination of the cyst showed a germinal layer and an avascular, eosinophilic, chitinous layer (Fig. 5)

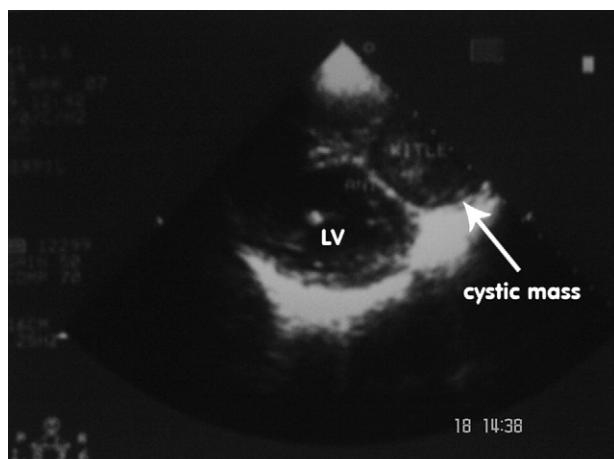


Fig. 1. Transthoracic two-dimensional echocardiogram of parasternal long-axis view showing a hydatid cyst (arrow).



Fig. 2. Nuclear magnetic resonance imaging, direct coronal section, shows an apical left ventricular cyst.

that confirmed the diagnosis of hydatid cyst and pericystic area consisting of fibrovascular tissue with mononuclear inflammation and foreign-body reaction (Fig. 6).

3. Discussion

Hydatid disease is a parasitic infestation most frequently caused by the larval form of the tapeworm *E. granulosus*, which uses the dog as the definitive host [2,3]. After ingestion, the larvae go through the duodenal wall to the portal blood system and into the liver, where they are found in about 60% of cases [5]. Some of them (ca. 20%) may escape hepatic filtration and enter the pulmonary circulation. A small percentage may reach the systemic circulation, resulting in infection and cyst formation in any organ [2,3]. Isolated cardiac involvement by echinococcosis at any age is very rare and occurs in 0.5–3% of the cases [2] as in our case.

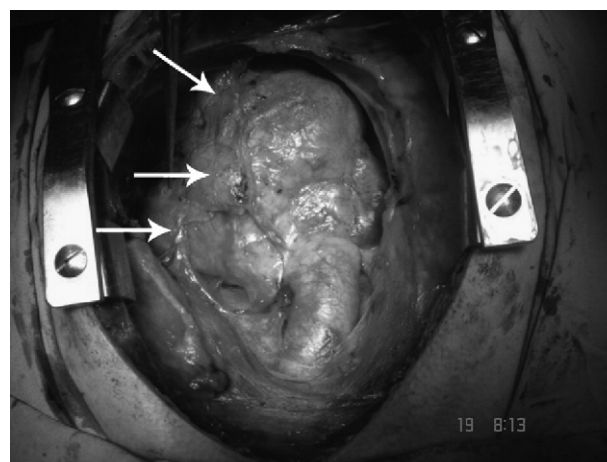


Fig. 3. Arrows point to the location of multiple cysts. Upper arrow: posteroapically located cyst; middle arrow: anteromedially located cyst; lower arrow: anterolaterally located cyst.

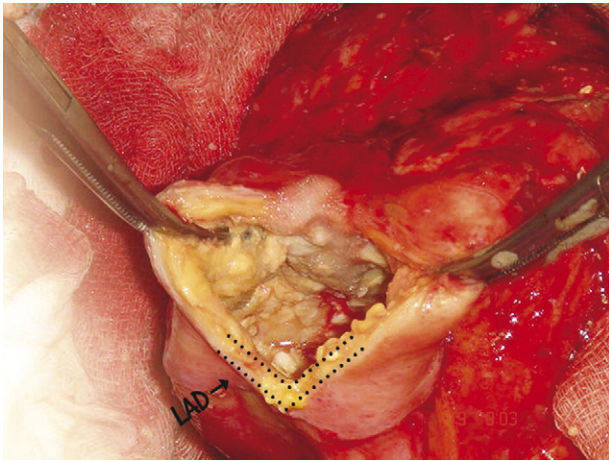


Fig. 4. LAD courses between dotted lines.

There is a strong supply of blood to the myocardium via coronary circulation. Cardiac cysts are commonly situated in the left ventricular free wall and interventricular septum because of their thickness and rich blood supply, although less frequent involvement of the right ventricle, atria, and pericardium has also been reported [6].

Consistent with the literature, the three cysts were localized to the anterior and apical left ventricular wall in our case. Precordial pain is the most common symptom and is most often vague and did not resemble that of angina pectoris. Typical angina is due to the compression of the coronary arteries. ECG often displays inverted T waves or right bundle branch blocks, but these changes are not specific. Eosinophilia is not specific. High titers of antiechinococcal antibodies are helpful but inconstant. The diagnostic capacity of transthoracic two-dimensional echocardiography is excellent, and this technique is the method of choice to detect CHCs and to determine their location, number, size, and relation to adjacent structures. When a hydatid cyst ruptures into a cardiac chamber, it undergoes a degenerative process and the typical echolucent appearance

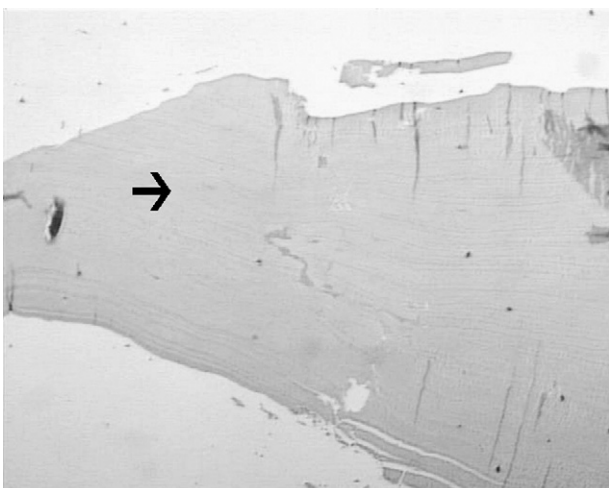


Fig. 5. Avascular, eosinophilic, chitinous layer (H&E, ×200; arrow).

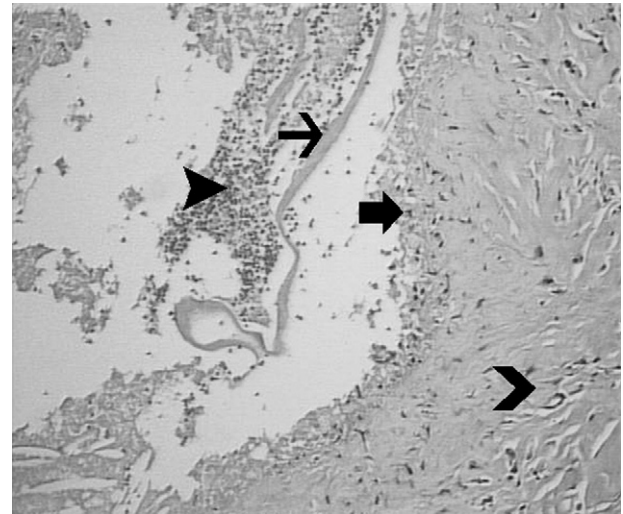


Fig. 6. Germinal layer surrounded with fibrous tissue with foreign-body reaction and inflammation (H&E, ×200). Arrows indicate (from left to right) inflammation zone, germinal layer, foreign-body reaction and fibrous tissue.

of the cyst becomes echogenic and the mass becomes solid and sometimes calcific if the patient survives and can be more difficult to differentiate from other heart tumors. CT may be a helpful diagnostic method. However, no information different from that acquired via two-dimensional echocardiography can be obtained using this technique.

Nuclear magnetic imaging (NMI) provides valuable information for the diagnosis, determines the location of the hydatid cyst, and allows evaluation of tumor extension and tissue involvement in the region of the hydatid cyst. In our case, the apically located hydatid cyst was more precisely detected via NMI than via echocardiography. However, NMI misdiagnosed the two other (anterolaterally and anteromedially located) cysts, which we detected intraoperatively and diagnosed as soft tissue thickenings.

Coronary arteriography is not advisable in all cases of CHCs but may be helpful in patients with typical angina pectoris or when the surgeon believes that it may influence the operative strategy [7].

The pathology of cardiac hydatid disease is similar to that described in other involved organs. Grossly, the capsule of the hydatid cyst is white, gelatinous, and glistening, and the cavity is filled with liquid. Smaller daughter cysts are often seen within the dominant cyst. Microscopically, the hydatid cyst is composed of a hyalinized and laminated outer “cellular” layer and a characteristic nucleated “inner” germinative layer. The inner layer contains protoscolices with distinctive, refractile sickle-shaped hooklets. Demonstration of scolices or hooklets in hematoxylin–eosin-stained sections or Giemsa-stained smears of cystic fluid is diagnostic for hydatid disease. Hydatid hooklets are acid fast, and the Ziehl–Neelsen stain can enhance the presence of hooklets when screening cyst fluid [5].

A CHC operation is classified as a surgical emergency because serious complications may occur (cerebral embolism,

global heart failure). The current surgical treatment of CHC is only via CPB. In some cases, it is possible to perform surgery without extracorporeal circulation, but the necessary equipment should be readily available in the operating theater [8]. Delivery of the cyst with an intact membrane (i.e., without prior puncture and emptying of the content of the cyst) is dangerous because of the high risk of rupture. Pericystectomy consists of the removal of the protruding dome and possibly the whole cyst if it is thin, superficial, and far from the dangerous vascular zones. Pericystectomy is done for precaution and should save the healthy myocardial fibers. Treatment of the residual cavity constitutes the most important, most delicate step and requires an empty, motionless heart. Effacement of the residual cavity is a necessary precaution, as an untreated cystic cavity is a fragile myocardial zone that could be the starting point of a possible postoperative aneurysm or even a fatal ventricular rupture. These procedures could not have been performed without compromising the course of LAD in our case. Perioperative mortality ranges from 5% to 10% in most series [6,7]. In the residents of these regions where echinococcosis is endemic, life-threatening, acute complications that may cause sudden death can be encountered. The mechanism of this fatal outcome may be caused by an anaphylactic reaction to cyst fluid leakage [3,4] or by disturbance of the vital structures of the cardiovascular system and related complications, such as cardiac tamponade or embolic phenomena [5–7].

Recurrences in the myocardium are exceptional [6], as this is not the usual site of secondary CHCs. Pericardial recurrences are more frequent [7]. Some complications are related to extracardiac secondary cysts. Their gravity depends on their localization and their number. If they are not numerous and if they are situated on organs easily accessible to surgery, the outcome is generally satisfactory. If they are disseminated or situated on delicate organs, such as

the brain, their course can be punctuated with life-threatening complications.

4. Summary

Cardiac hydatid disease is very rare, even in endemic regions. Superior displacement of LAD with compression was noted, and this condition necessitated a LAD–LIMA bypass graft in our case.

Acknowledgment

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