Surgical treatment and outcome of cardiac cystic echinococcosis

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Abstract

OBJECTIVES: Cardiac cystic echinococcosis (CE), or hydatid cyst is exceptionally uncommon. We review the experience of surgical treatment of cardiac CE.

METHODS: Twenty-six patients (11 females; mean age, 28.9 ± 7.6 years) with cardiac CE undergoing surgical treatment from February 1978 to April 2013 were reviewed. The operative methods mainly included puncture-aspiration cystectomy, intact endocyst enucleation and total cyst resection.

RESULTS: Cardiac CE was located in the myocardium in 16 cases, pericardium in 8 and both myocardium and pericardium in 2. There were 21 cases with solitary hydatid cyst including at the left ventricle in 7, right ventricle in 5, right atrium in 2, interventricular septum in 1 and at the pericardium in 6; 5 cases were with multiple cysts. There was no operative mortality. All patients received albendazole post-operatively. The mean follow-up time was 68 ± 21 months (range 7–195 months) except for 4 who were lost to follow-up. There were three recurrences and one late death.

CONCLUSIONS: Cardiac cystic echinococcosis (CE) remains a very infrequent zoonotic infection. Presenting symptoms of cardiac hydatid disease are variable depending on the size, number and location of the cyst. Echocardiography, corroborated with computed tomography or magnetic resonance imaging, affords the best diagnostic and follow-up confirmation. Surgical treatment is associated with a low morbidity and mortality, and the selection of proper technique is very important to completely remove the hydatid cyst and prevent recurrence. Postoperative oral antiparasitic therapy is necessary for a definitive cure.

Keywords: Cardiac • Hydatid disease • Myocardium • Pericardium • Surgery • Incisions • Exposure • Techniques

INTRODUCTION

Human echinococcosis is a zoonotic infection caused by larval forms of tapeworms of the genus Echinococcus found in the small intestine of carnivores. The most common, cystic echinococcosis (CE) or hydatid cyst is the designation for the larval phase of Echinococcus granulosus [1]. Some carnivores (most often dogs) are the definitive host for this tapeworm. The eggs of these tapeworms excreted by carnivores may infect various species of natural intermediate host animals such as cattle and sheep, and occasionally humans. Human CE is caused by the ingestion of contaminated food or by close contact with infected animals or contaminated soil. The ova hatch into hexacanth in the small intestine and penetrate the intestinal wall, pass through the portal vein into the liver, lungs and other tissues and develop into a hydatid cyst. The life cycle is completed when carnivores eat the cyst-bearing organs of the intermediate hosts. Cardiac CE is exceptionally uncommon and is reported in 0.5–2% from all registered echinococcosis cases in comparison with liver (70%) or lung (20%) involvement [2, 3]. Although some reports of surgical management with cardiac hydatid disease have been published in isolated cases [2, 4, 5], few relatively large series of patients have been reported. We reviewed our 35-year experience of the diagnosis and the surgical treatment of 26 patients with cardiac CE, and provide diagnostic and surgical therapeutic recommendations.

PATIENTS AND METHODS

From February 1978 to April 2013, 26 consecutive patients with cardiac CE were surgically managed at the First Affiliated Hospital of Xinjiang Medical University. There were 15 males and 11 females, and ages ranged between 8 and 60 years, with a mean of 28.9 ± 7.6 years. Six patients were sheep-farmers and all the cases lived in an endemic area at least once. At the time of admission, 3 patients were non-symptomatic (diagnosis of cardiac CE was...
made following investigations for other causes). Twenty-three of them presented with symptoms. Major symptoms were exertional dyspnoea in 15, chest pain in 11, palpitations in 8, cough in 7 and fever in 2. Chest radiographs were normal in 10 patients, and 3 had evidence of pulmonary hydatidosis. Cardiomegaly was present in 13 patients including generalized in 5, left ventricular in 5 and right ventricular in 3. Pericardial calcification was seen in 2 patients. Electrocardiograms were reported as normal in 7 patients, and with left ventricular hypertrophy in 5, right ventricular hypertrophy in 2, incomplete right bundle branch block in 3, incomplete left bundle branch block in 1 and non-specific ST/T changes in 8. Eosinophilia (>400 cells/mm³) was present in 15 patients (58%). Casoni intradermal test was positive in 15 (65%, 15/23) patients. Serological evaluations including the indirect haemagglutination (IHA) test and enzyme-linked immune sorbent assay (ELISA) were positive in 15 (79%, 15/19) and 10 (83%, 10/12) patients, respectively. Coronary angiography was performed in 3 cases and the findings were normal. The diagnosis of cardiac CE was established mainly by trans-thoracic echocardiography. Seven patients also underwent transoesophageal echocardiography (TOE) preoperatively. Computed tomography (CT) was carried out in addition to echocardiography in 10 patients, and magnetic resonance imaging (MRI) in 3; the results confirmed the echocardiographic findings and identified new extracardiac cyst locations in 4.

In our group with 26 cases, cardiac CE was located in the myocardium in 16 cases, pericardium in 8, and both myocardium and pericardium in 2. There were 21 cases with solitary hydatid cysts including left ventricular cyst in 7, right ventricular cyst in 5, right atrial cyst in 2, interventricular septum cyst in 1 (Fig. 1) and pericardial cyst in 6; and there were 5 multiple cysts (≥2 cysts) including left ventricular cysts in 1, left ventricular combined with pericardial cysts in 2 and multiple pericardial cysts in 2 (Figs 2–4).

The exact distribution of cardiac CE in our group was as follows: there were 18 myocardial cysts (including left ventricular combined with pericardial in 2 cases) which were localized in the left ventricle in 10 cases (38%), right ventricle in 5 (19%), right atrium

Figure 1: Two-dimensional transthoracic echocardiography demonstrated a cystic mass measuring 4.3 × 4.5 cm (arrows) within the interventricular septum.

Figure 2: Two-dimensional transthoracic echocardiography demonstrated a huge multiloculated cystic mass measuring 9.3 × 12.4 cm (arrows) in the pericardium.

Figure 3: Contrast-enhanced CT showed two well-defined cystic masses in the pericardium with axial (A) and coronal view (B). A unilocular cyst (3.1 × 4.7 cm) was located close to the left atrium and a second multilocular cyst (8.7 × 7.9 cm) with multiple daughter cysts adjacent to the left ventricle compressing the anterolateral wall (same patient as in Fig. 2).
in 2 (8%) and interventricular septum in 1 (4%), and there were 10 pericardial cysts including solitary cysts in 6 (23%) and multiple cysts in 4 (15%). Transthoracic echocardiography revealed the cysts with a mean diameter of 5.4 cm (range 1.0–13.5 cm). Seven patients had concomitant extracardiac hydatid disease: involvement of the liver in 3, one or both lungs in 3 and lungs and liver in 1.

All operations were carried out under general anaesthesia and median sternotomy was used in 20 and left anterolateral thoracotomy in 6. Eleven operations were performed with the beating-heart technique, while 15 were carried out under cardiopulmonary bypass (CPB), moderate hypothermia and crystalline cardioplegia. The operative field was wrapped with sponges moistened with 20% hypertonic saline solution, which was also instilled into the cysts, washing the residual cavity and the pericardial cavity as a larvicidal agent.

There were 18 patients with myocardial cysts including: solitary cysts in 15 (ventricular cysts in 12, right atrial cysts in 2 and interventricular septum cyst in 1), and multiple cysts in 3 (left ventricular in 1, left ventricular combined with pericardial in 2). In 6 cases with minor and superficial ventricular cysts, ‘intact endocyst enucleation’ was performed on the beating heart in 3 cases and under cardioplegic arrest with CPB in 3 cases, starting with an incision in the epicardium (or myocardium) where the cyst was nearest the surface of the heart, followed by careful and gentle incision of the ectocyst. When the proper depth of incision was reached, the whitish endocyst protruded from the incision and was carefully enucleated without rupture. The epicardial or myocardial pouch resulting from cyst enucleation was closed by primary suturing.

In the other 9 ventricular cysts, ‘puncture–aspiration cystectomy’ was performed under CPB with cardioplegic arrest in 8 and on the beating heart in 1. The cysts were punctured with a wide-bore needle, connected with a suction device, and the cystic contents were aspirated. After sterilization by instillation of 20% hypertonic saline solution, the cyst was opened and the endocyst and remaining daughter cysts were removed. The residual cavity was obliterated by multiple interrupted prolene sutures.

In 2 right-intra-atrial cyst cases, standard CPB with bicaval cannulation under cardioplegic arrest was used. On opening the right atrium, one intra-atrial cyst was attached to the root of the tricuspid valve, and the other one was located in the right lateral atrial wall in close proximity to the superior vena cava. Puncture-aspiration cystectomy was performed in the 2 cases; the cystic mass was then carefully opened and its contents were aspirated and the ectocyst excised.

In the case of the one interventricular septum cyst, after cardioplegic arrest under CPB, an incision adjacent to the left anterior descending artery was made, without entering any cardiac chambers. Then, puncture–aspiration cystectomy was performed, and the endocyst was removed but not the ectocyst, to avoid impairment of atrioventricular conduction. The defect of the ventricular septum was closed with a Dacron patch. The ventriculotomy incisions were sutured linearly using Teflon felt strips with 2–0 Ethibond.

There were 10 patients with pericardial cysts, including solitary cysts in 6 and multiple cysts in 4 (multiple pericardial cysts in 2, and both pericardial and left ventricular cysts in 2). ‘Total cyst resection’ (an enlarged resection made to remove both of the endocysts and ectocysts as a whole and usually a part of adjacent pericardium) was performed in 8 cases. Puncture–aspiration cystectomy was successfully performed in 1 patient with two cysts about 9 and 5 cm in diameter (Fig. 5). In 1 patient with multiple pericardial cysts (five cysts) combined with secondary cyst infection and pericardial thickening and calcification, puncture–aspiration cystectomy was performed and the endocysts were removed, but some ectocysts remained due to intensive calcification and adhesion.

RESULTS

There was no operative mortality. Pathology reports all indicated intact or degenerated hydatid cysts. The postoperative period was uneventful for all patients. Patients were checked postoperatively at least every 6 months. The mean follow-up time was 68 ± 21 months (range 3–158 months), except for 4 who were lost to follow-up (more than 10 years after operation). One late death due to chronic right heart failure occurred in a patient who had a right atrial hydatid cyst after 10 months of follow-up. There were three recurrences including one of multiple pericardial cysts and

Figure 4: MRI shows multilocular and unilocular lesions that are hyperintense on T2-weighted axial and coronal non-contrast images, which is in the same location as that shown by CT (same patient as in Figs 1 and 2).
two of myocardial cysts after a mean of 12 months; these patients underwent reoperation.

In 7 patients with extracardiac cysts in addition, 3 of them had lung cysts and were referred for surgery within 6 months after the initial cardiac operation; and 1 patient had left ventricular cysts concomitant with left lower lobe and hepatic cysts and underwent puncture-aspiration cystectomy for the ventricular and lung cysts in one stage through a left anterolateral thoracotomy, and was then referred for hepatic CE surgery 3 months after the initial cardiopulmonary operation.

As recommended, all patients received albendazole (10–15 mg/kg/day orally, for 28–30 days with intervals of 14 days in between treatments, for 1–6 courses) postoperatively [6]. This was then discontinued in cases with solitary or degenerated cysts and when there was no evidence of extracardiac hydatidosis. Otherwise, the anthelmintic drug was prescribed for at least 6 months, and as long as there remained evidence of inoperable systemic hydatidosis.

**DISCUSSION**

Human CE is of worldwide importance and presents medical, veterinary and economic problems for developing countries. The Xinjiang region in north-western China, among other sheep-farming areas in the world, is still considered endemic for human CE [7]. Cardiac involvement in CE can be divided into myocardial and pericardial cysts, which are either primary or secondary. Primary involvement of the heart usually occurs via coronary circulation; the intestinal lymphatics, thoracic duct, upper and lower venae cavae, pulmonary capillary network and patent foramen ovale may also be a pathway [8]. Secondary involvement occurs from hydatid disease of adjacent organs, such as lungs and parts of the mediastinum, or from the dome of the liver in which abdominal cysts prolapse through the diaphragm [9]. Most studies found that the distribution of cardiac CE parallels that of coronary blood flow and most lesions (40–60%) occur within the left ventricle, followed by the right ventricle (15–30%), the interventricular septum, the left atrium, the right atrium and the interatrial septum (2–10%, respectively). Primary pericardial cysts are rare (2–10%), and they generally develop secondary to intrapericardial rupture of a myocardial cyst or to spillage of cystic contents during surgical removal [2–4, 6, 10–12]. In our group, the cardiac CE were localized in the left ventricle in 10 cases (38%), right ventricle in 5 (19%), right atrium in 2 (8%), interventricular septum in 1 (4%) and pericardium in 10, including solitary cysts in 6 (23%) and multiple cysts in 4 (15%).

The hexacanth embryo starts to develop in cardiac tissue and within 1–5 years forms the actual cyst. The average duration of the life of CE in humans is 10–20 years and the annual growth rate of the cyst is usually about 1–5 cm in diameter [6, 12, 13]. The hydatid cyst is composed of two parts: the endocyst (the hydatid parasite) and the ectocyst (the adventitia that surrounds the intact laminated layer). The endocyst is composed of two layers: the germinal layer, the function of which is to elaborate the different elements of the hydatid, and the laminated layer. The ectocyst results from the host's organ reaction against the hydatid parasite considered as a foreign body, which is an adventitial layer that surrounds the intact laminated layer [1]. In myocardial hydatid cysts, the ectocyst tissue becomes progressively more abundant and eventually forms an inseparable portion of the myocardium. Because of the configuration of the heart and the density of the myocardium, particularly the ventricular wall, the growth of a hydatid cyst is sometimes restrained; whereas, in pericardial hydatid cysts, the size of the cysts can be much bigger, even more than 10 cm in diameter.

**Figure 5:** The puncture-aspiration cystectomy technique for a huge pericardial cyst: the cysts were punctured with a wide-bore needle, connected with a suction device, and the cystic contents were aspirated (A). After sterilization by instillation of 20% hypertonic saline solution, the cyst was opened and the endocyst and remaining daughter cysts were removed (B). Washing the residual cavity and the pericardial cavity with 20% hypertonic saline solution (C). The resected endocyst and daughter cysts (D).
The diagnosis of cardiac CE can be difficult and must be suspected in any patient from endemic or sheep farming areas with a cystic tumour of the heart. Presenting symptoms of cardiac hydatid disease are variable depending on the size, number and location of the cysts. In the early period, the cysts are very small and grow slowly without causing any symptoms. As the cysts reach a reasonable size, the common primary symptoms that can occur are palpitations, dyspnoea and chest pain [10]. Further, some hydatid cysts can result in serious consequences. The most dangerous complication is cyst perforation. Usually, left ventricular cysts perforate out of the cavity resulting in acute cardiac tamponade and right ventricular cysts perforate into it, leading to drastic anaphylactic reaction or pulmonary, intracerebral or peripheral arterial embolic complications [14]. The frequency of intracardiac perforation is very high (25–40%). After cyst perforation 75% of the patients died from septic shock or embolic complications [15]. In addition, the cysts can give rise to symptoms such as those associated with the compression of a coronary artery, with a disturbance in valvular mechanisms, or with outflow tract obstruction or a variety of conduction defects caused by the involvement of the interventricular septum [16, 17]. In a few cases, the cysts may become stagnant or inactive and may degenerate with calcification or infection.

For the radiological diagnosis of cardiac CE, chest X-ray, echocardiography, CT or MRI might be used. On transthoracic echocardiography classic cardiac CE lesions reveal well-defined unilocular or multivesicular cysts with uniform anechoic content, whereas those that have become solidified, calcified or degenerated can be inhomogeneous hypechoic or hyperechoic, and thus mimic semisolid or solid mass lesions [18–20]. It is possible to get more detailed images with TOE. According to this group, echocardiography is the preferred diagnostic method because of its low cost and availability. However, echocardiography is operator-dependent, with a limited field of view; in certain thoracic deformities and variants of the patient’s habitus, the area behind the sternum may be difficult or impossible to examine. CT and MRI are superior to echocardiography for the evaluation of pericardial masses and their relationship to surrounding tissues and extension to the myocardium. Coexisting pulmonary, mediastinal and new extracardiac hydatid cystic disease may also be detected by CT and MRI [9]. Specific signs of hydatid cysts include calcification of the cyst wall, multivesicular nature of the cystic mass and membrane detachment, which indicate a true and accurate diagnosis. CT best shows wall calcification, whereas MRI depicts the exact anatomical location [21]. In our group, CT was carried out in addition to echocardiography in 10 patients, and MRI in 3. The results confirmed the echocardiographic findings and identified new extracardiac cyst locations in 4 patients. Therefore, if available, echocardiography, corroborated with CT or MRI, affords the best diagnostic and follow-up confirmation.

Routine laboratory tests are not specific and may reveal normal or abnormal values. The blood count may show eosinophilia; the Caroni intradermal test and serological tests (IHA or ELISA techniques) are useful but are subject to false negativity in the diagnosis of hydatid disease [19, 22]. We found it possible to establish reliable diagnoses with the combined use of echocardiography and serological tests.

Surgical removal of the cyst is the definitive treatment to prevent potentially life-threatening complications [4, 5, 10]. Median sternotomy is the most common surgical access to the heart, although access by left anterolateral thoracotomy was utilized in 6 of our cases. In patients concomitant with extracardiac cysts (lung and liver), it is possible to combine the cardiac operation with a thoracic or abdominal exploration to remove accompanying pulmonary or hepatic cysts, as demonstrated in one of our patients. Alternatively, surgical interventions are recommended to be performed in separate steps with intervals of a few months. The general principle is to give priority to the cysts prone to complications. We prefer first removal of pulmonary cysts, followed by cardiac cysts and hepatic cysts due to likely earlier rupture of pulmonary cysts under general anaesthesia and mechanical ventilation.

In patients with pericardial cysts, surgical interventions usually can be done on a beating heart. In patients with myocardial cysts, the use of CPB under cardioplegic arrest is still our favourite choice with the least risk of spillage of cyst contents during the procedure, although some superficially located cysts can be removed with a beating-heart technique. It is, however, also possible to remove superficial cysts using CPB with an empty beating heart. In the operation on cardioplegic arrest, cross-clamping of both the aorta and the pulmonary artery is beneficial to prevent dissemination of the parasite into the left ventricular cavity or via the ventricular septum into the pulmonary circulation.

The main purpose of the operation is to remove the endocyst and try to avoid heart tissue injury. Based on the anatomical features and according to the size, location and complications of the cardiac CE, there are three main methods for surgical removal of cardiac cysts: puncture–aspiration cystectomy, intact endocyst enucleation and total cyst resection.

In patients with pericardial cysts, total cyst resection should be the first choice, which is to remove both the endocyst and ectocyst as a whole. In large or multiple pericardial cysts, there are more chances of tight adhesion or calcification which may result in cyst rupture during operation. Therefore, puncture–aspiration cystectomy should be performed in these patients. The endocyst is decompressed through puncture–aspiration and removed, followed by the ectocyst resection if possible.

Most myocardial cysts are characterized by solitary intact cysts that are embedded in the myocardium and may grow towards the endocardium or towards the adventitia. Owing to systolic and diastolic motion of the heart, the ectocyst tissue may become progressively more abundant and eventually forms an inseparable portion of the myocardium. We suggested that intact endocyst enucleation or puncture–aspiration cystectomy be used for myocardial cysts other than total cyst resection. Furthermore, some myocardial cysts adjacent to coronary arteries and some cysts can be intracardiac (atrial cysts or interatrial/interventricular septum cysts), which may be close to heart valves or conductive tissue. To avoid aggressive surgery, we ensure the resection of the endocyst but not all of the ectocyst in those cysts mentioned above.

The technique of intact endocyst enucleation completely obviates the possibility of dissemination through spillage, which is better be performed safely on a heart arrested by cardioplegia with CPB in patients with minor (<4 cm in diameter) and superficial myocardial cysts, although this can also be done successfully on a beating heart. Endocyst rupture may occur during performing the intact endocyst enucleation technique. If this occurs, the residual and pericardial cavity should be filled for 15 min with 20% hypertonic saline solution, which provides reliable decontamination. If the surgeon estimates that there is danger of cyst rupture during enucleation, it is advisable to resort to puncture–aspiration cystectomy. The technique of puncture–aspiration cystectomy is relatively simple and can be performed in all kinds of cardiac cysts, mainly applied to large or multiple pericardial cysts,
large or deep ventricle cysts, as well as atrial or septal cysts, and also infected or calcified cysts.

Risks at surgery include anaphylaxis and dissemination of protoscoleces due to spillage of the cystic fluid. When a hydatid cyst is to be removed, the use of protoscolicidcs such as 70–95% ethanol, 15–20% hypertonic saline solution or 0.5% cetrimide solution is recommended. For optimal efficacity, the substances have to be left in the cyst cavity for at least 15 min [6, 23]. We use 20% hypertonic saline solution as a larvicidal agent.

About 10% of all hydatid cysts tend to recur after surgery, but this rate may decrease with proper medical treatment. Chemotherapy with albendazole or mebendazole is reserved for patients not suitable for surgical intervention, and may also be given to reduce the risk of recurrence, especially in the event of intracardiac rupture. Chemotherapy is contraindicated for large cysts with a risk of rupture or for inactive or calcified cysts [6, 7, 24]. Although it is recommended that medical treatment be started a few weeks prior to surgery, it is important to understand that chemotherapy may lead to a higher risk of cyst wall destruction and rupture, especially in cardiac cysts [25]. Therefore, we suggest prophylactic chemotherapy with albendazole several days before, and continued for several weeks after surgery, the duration of which can be regulated according to the number of cystic lesions and extent of organ involvement.

Conflict of interest: none declared.

REFERENCES