Management of Cardiac Hydatid Disease

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Abstract

Hydatid disease also known as echinococcosis is an endemic zoonotic infestation in cattle-raising regions of the world formed by the larval forms of cestodes belonging to the genus Echinococcus. This disease is mainly caused by the larval form of Echinococcus granulosus and Echinococcus alveolaris.

Although hydatidosis is most often occurred in the liver [60%-70%] and lung [20%-30%], it can occur in any organ or tissue from head to toe, including heart. Cardiac involvement is rare and accounts for only 0.5%-2% of all hydatidosis cases. Initially there may be no remarkable symptom in patients with cardiac hydatidosis. When the cyst reaches a significant size, some symptoms and complications may be manifested. Chest pain, palpitation, and shortness of breath are the main symptoms associated with cardiac hydatidosis. Although serologic tests are a helpful supplement for diagnosis, false negative results are possible. Definitive diagnosis of cardiac hydatidosis is mainly based on echocardiography. Even, asymptomatic patients can be diagnosed incidentally on echocardiography. The differential diagnosis of cardiac hydatidosis includes ventricular aneurysm, pericardial and cardiac cysts, myocardial abscess, pericardial hematomas, and cystic degenerative tumors.

The combination of both surgical and medical treatment should be done in all patients with cardiac hydatid disease. Surgical treatment can be performed with or without cardiopulmonary bypass (CPB) according to the localization of the cardiac cysts. Although median sternotomy is preferred incision for surgical intervention, a mini anterolateral left thoracotomy incision can also be used successfully in patients with apical and apical-lateral myocardial hydatid cysts. If the protoscolices in the cystic fluid are accidentally poured into the surgical field during operation, formation of secondary cysts is possible. Hence, the whole of the surgical field must be protected by washing with scolicidal agents to prevent secondary cystic development.

Although patients with cardiac hydatidosis may be asymptomatic for many years or have non-specific complaints, they may have lethal complications, including tamponade, anaphylactic shock and sudden death. Therefore, all patients with cardiac hydatidosis should be operated on as soon as possible.

INTRODUCTION

Hydatid disease also known as echinococcosis is an endemic parasitic infestation in cattle-raising regions of the world formed by the larval forms of cestodes belonging to the genus Echinococcus family Taeniidae [1-3]. It has been known the time of Galen and Hippocrates. The term hydatid cyst (watery vesicle) was first used by Rudolphy in 1808 [4] and was derived from Greek word ‘hudatis’ (drop of water) [5]. Cardiac hydatidosis was first mentioned in 1836. Arturico et al. [6], performed the first successful surgical intervention using cardiopulmonary bypass (CPB) in 1962.

Four species of hydatidosis have been identified that create important public health concerns. Human hydatid disease is mainly caused by the larval form of Echinococcus granulosus and Echinococcus alveolaris [1,7,8]. This disease is one of the most neglected public health problems in people [9].

The adult parasite lives in the intestinal mucosa of the definitive hosts, such as dogs, foxes and jackals. Herbivores such as sheep, cattle and goats, are the natural intermediate hosts. These animals have been reported as the most important sources of infestation in the life cycle of E. granulosus [9]. Humans are accidental intermediate hosts [2,7,10]. This tapeworm produces eggs (ova) that are passed into the definitive hosts’ stool. If these eggs are ingested by intermediate hosts (humans or ungulate intermediate hosts), the embryos (oncospheres) released from the eggs penetrate the intestinal wall until they reach a small vessel. Most of them are stuck in the hepatic sinusoids, where they
transform into a hydatid cyst (metacestode stage). The liver and the lungs are the most common sites of this infestation. Rarely, a few of embryos may escape from two filters [portal system and pulmonary capillaries] and may reach the myocardium through the coronary artery circulation [3,11, 12]. Furthermore, larvae might reach the heart through the lymphatic circulation, superior and inferior caval veins, and the hemorrhoidal veins of the large intestine [11,13]. Cardiac hydatidosis may also develop by the rupture of pulmonary cysts into the pulmonary vein or due to direct extension from the adjacent structures [14]. Airborne infestation might be possible, if any scolices may reach the alveoli and pierce the alveolo-capillary membrane through the pulmonary veins [15].

The life cycle of the parasite is completed by the ingestion of an infected organ of intermediate hosts by any definitive hosts. Humans do not play a role in the continuation of the life cycle of parasite as an accidental intermediate host, because the bodies of dead people are buried [16].

Although hydatidosis is most often occurred in the liver (60%-70%) and lung (20%-30%), it can occur in any organ or tissue from head to toe, including heart [14,17]. Cardiac involvement is rare and accounts for only 0.5%-2% of all hydatidosis cases [11]. The left ventricle free wall and interventricular septum are commonly involved in patients with cardiac hydatidosis, because of their thickness and rich blood supply [18]. The distribution in the left ventricle, right ventricle, interventricular septum, and left atrium was reported as 60%, 15%, 9%, and 8% of cardiac cases, respectively [3]. The pericardium is involved in 8% of cases [19]. Pericardial hydatid cysts are usually the result of rupture of cardiac cysts [20]. The pericardial hydatidosis may also develop when the subepicardially located cardiac cysts are opened into the pericardium [21]. The opening of the cystic content into the pericardial space may lead to tamponade due to an exaggerated exudative reaction of the pericardium even in patients with small cyst [20]. Rupture into the pericardial space may also cause constrictive pericarditis due to an inflammatory reaction [5,22].

**CYST STRUCUTRE**

Hydatid cyst consists of three layers with water inside. The first layer is the pericyst or adventitia, which is the fibrous host reaction against the parasite. The second layer is the outer layer of the cyst also known as the laminated membrane, which is an acellular mucopolysaccharide layer. The third layer (inner layer) is also known as the germinative membrane. Brood capsules and protoscolices are budded from the germinal layer. [7,10,23]. Hydatid cyst fluid contains carbohydrates, proteins, lipids, electrolytes, vitamins and trace elements that may have a role in the metabolism and growth of the cyst [24].

**SYMPTOMS**

Initially there may be no remarkable symptom in patients with cardiac hydatidosis. Thameur et al. [20], reported that only 60% of patients with cardiac hydatidosis were symptomatic. Symptoms are closely related to the size and location of the cysts. When the cyst reaches a significant size, some symptoms and complications may be manifested [19,25].

Chest pain, palpitation, and shortness of breath are the main symptoms associated with cardiac hydatidosis [11]. If cardiac cyst reaches a large diameter, it may lead to severe symptoms such as angina, palpitation, and arrhythmias [13]. Cardiac hydatidosis growing toward the epicardium may cause symptoms suggesting coronary artery disease, while those growing toward the septum may lead to conduction defects [26]. Patients with epicardial hydatid cysts compressing the small coronary arteries might be misdiagnosed as coronary artery disease [11]. Palpitations are frequently sensed due to a transitory cardiac arrhythmia, often without electrical signs [20]. Mechanical compression of the left ventricle and the fibrotic reaction may lead to heart failure symptoms [27]. If the cardiac hydatid cysts expand into the cardiac cavity, they may cause murmurs suggesting valvular lesions.

Hydatid cysts of the right ventricle [usually subendocardially located] expand into the cardiac cavity, whereas those of left ventricle [usually subepicardially located] expand toward the epicardium and pericardium, because left ventricle has a thicker and denser myocardial tissue and higher pressure in it [20,28]. Rupture into the pericardial space may lead to tamponade due to an exaggerated exudative reaction of the pericardium even in patients with small cyst, while rupture into the cardiac cavity causes systemic or pulmonary embolic complications [20]. Thus, even in asymptomatic patients, cardiac involvement may lead to sudden death due to fatal complications [18].

**DIAGNOSIS**

Chest radiographs might reveal cardiomegaly, mass on borders and deformation of the cardiac silhouette in patients with cardiac masses [22,25]. ST segment and T wave alterations may be found in electrocardiograms, reflecting the localization of the hydatid cyst. Loss of QRS voltage and Atrioventricular conductions defects may be found with left ventricular and interventricular septal involvements, respectively [13]. However, definitive diagnosis of cardiac hydatidosis is mainly based on echocardiography. Even, asymptomatic patients with cardiac hydatidosis can be diagnosed incidentally on echocardiography.

Two-dimensional echocardiography is a reliable diagnostic method for the cardiac hydatid cysts and can also show hemodynamic function of heart. In patients with cardiac hydatidosis, surgical intervention can safely be based on the echocardiographic findings. Other imaging techniques [CT, MRI] may be required in the case of unreliable images and especially to investigate other cystic locations [20]. Occasionally, the echoluent and multi-septate nature of cardiac hydatid lesions may be absent [29]. CT or MRI should be performed in these cases. Additionally, CT and MRI are superior to echocardiography for the evaluation of internal structure of the mass lesions, their relationship to adjacent tissues, and coexisting lung or mediastinal hydatidosis [19,25]. MRI is a more reliable diagnostic tool than the CT scan for diagnosing of cardiac hydatidosis [20]. Important information can be obtained with nuclear magnetic imaging (NMI) for the diagnosis. NMI determines the location of the cysts, and allows evaluation of cystic extension [30].

Although serologic tests are a helpful supplement for diagnosis of cardiac hydatidosis, false negative results are possible [18]. IgG ELISA and IHA are the most sensitive serologic
tests [31]. Serologic tests can be false-negative in 10% to 20% of patients with hepatic hydatidosis, 40% with pulmonary hydatidosis, and 50% with cardiac hydatidosis [32]. Even in a ruptured cyst into the cardiac cavity, serological tests might be negative, whereas hyperesinophilia may be found positive [20]. However, hyperesinophilia is not always present in patients with hydatidosis [11,33]. Sometimes, false positive serological results may be possible, because of other parasitic infestations, malignancy and chronic immune disorders [5].

**DIFFERENTIAL DIAGNOSIS**

The differential diagnosis of cardiac hydatidosis includes ventricular aneurysm, pericardial and cardiac cysts, myocardial abscess, pericardial hematomas, and cystic degenerative tumors [19]. Sometimes myocardial hydatid cyst remains inactive and loses its fluidity and degenerates in time. If this degenerated cyst that protrudes into the outflow tract is covered with fibrin, it may mimic atrial myxoma [15].

**TREATMENT**

The combination of both surgical and medical treatment should be done in all patients with cardiac hydatid disease [34]. Benzimidazoles [mebendazole and albendazole] and praziquantel are used in medical treatment. Birinciöglu et al. [15], suggested that albendazole should be prescribed to all patients both preoperatively and postoperatively. Albendazole should be prescribed 400 mg twice daily after meals for 4 weeks course. After a 2-week break, another cure should be done again. One preoperative and 3 postoperative course should be suggested [15]. Drug therapy should be given for at least 2 years after surgical intervention and these patients should be monitored for at least 10 years [5].

Preoperative albendazole therapy might be useful to avoid contamination, if cyst contents spread the surrounding operative field [15]. In contrast, some suggested that preoperative drug therapy should not be performed, because the cyst may become more friable and even may rupture during surgical intervention [34].

PAIR (puncture, aspiration, injection and reaspiration) procedure may lead to serious complications in patients with cardiac hydatidosis. Therefore, we believe that it should be preferred in patients with abdominal hydatidosis instead of cardiac hydatidosis.

Surgical treatment of cardiac hydatidosis can be performed with or without CPB according to the localization of the cyst. Median sternotomy is preferred incision for surgical removal of the cardiac hydatidosis. Furthermore, this incision can be successfully applied in heart and bilateral lung cysts [35]. Although median sternotomy is preferred incision for surgical intervention, a mini anterolateral left thoracotomy incision can also be used successfully in patients with apical and apical-lateral myocardial hydatid cysts [36]. Video-assisted thoracoscopic surgery (VATS) might be performed especially in patients with dead pericardial or epicardial hydatidosis. However, every measure must be taken.

Birinciöglu et al. [15], suggested that all cysts without ventricular connection can be operated on without CPB. In contrast, some suggested that CPB and cross-clamping of aorta should be performed to prevent embolic complications [14,37]. In patients with right-sided cardiac hydatidosis, both the aorta and pulmonary artery should be clamped to avoid systemic and pulmonary embolism. Furthermore, a supplementary filter to venous circuit should be added to avoid passage of cystic particles toward the heart-lung machine [20]. We also think that it is useful to add a filter to arterial line to avoid passage of unseen cystic particles toward the heart-lung machine.

It has been suggested that off-pump hydatid cyst operation has an advantage, if surgeon accidentally enters intracardiac cavity. In this circumstance, cystic embolization does not occur into the cardiac cavity. In contrast, embolization might occur during on-pump surgery due to low intraventricular blood pressure [15].

Enucleation of the cardiac hydatid cyst with its intact membrane has been found dangerous due to high risk of rupture [30]. To prevent accidental tear of cysts in to the ventricular cavity, cysts, which do not protrude any direction, needle aspiration may be useful to guide the dissection towards where the cyst fluid comes [15]. Cystic space should be punctured and emptied with a syringe (Figure 1). Then it should be sterilized by injecting a scolicidal agent. If the protoscolices in the cystic fluid are accidentally poured into the surgical field, formation of secondary cysts is possible [38]. Hence, whole operative field should be surrounded with sponges soaked with scolicidal agents to prevent secondary cystic development. Then, an incision is made and whole cystic content removed. Ventricular arrhythmias leading to sudden death may occur after surgical intervention due to scar formation [21]. To avoid this complication, the ventricular incision should be as small as possible. Some suggested that the residual cystic cavity should be eliminated to prevent a possible postoperative ventricular aneurysm or even a fatal myocardial wall rupture [30]. In contrast, Birinciöglu et al. [29], suggested that residual pouches should be left open, because, these pouches will be closed with secondary healing. Although they suggested that sutures used for closure may also cut myocardium [15], the strong cyst-related fibrotic tissue facilitates the suturing of the

![Figure 1](image1.png)
myocardial incision without tearing the myocardial wall in most patients [39].

CONCLUSION

Although patients with cardiac hydatidosis may be asymptomatic for many years or have non-specific complaints, they may have lethal complications, including tamponade, anaphylactic shock and sudden death [40]. Therefore, all patients with cardiac hydatidosis should be operated on as soon as possible.

REFERENCES


