

11 Years Experience with Cardiac Hydatid Cyst Operation at Rajae Heart Center

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Abstract

Background- Cardiac involvement in hydatid disease is uncommon (<2%). Symptoms depend on the location, size and integrity of cyst and the patients may be asymptomatic or in profound circulatory collapse.

Method- In this descriptive, cross-sectional study from March 1992 to July 2003, we had 13 proved cardiac hydatid cysts in our center. We evaluated clinical signs, symptoms, diagnostic procedures and treatment results of these patients.

Results- Average age of the patients was 36 ± 3.2 years, 61.5% were female and 38.5% were male. Dyspnea was the most common symptom; followed by dysrhythmia. Inverted T- wave and widening of QRS complex existed in 30%. Cardiomegaly was seen in 61%. Echocardiography showed cystic lesion in all cases. The most common sites of cardiac involvement were interventricular septum (46%), followed by right atrium (15.3%), LV free wall (15.3%), pericardium (7.7%), RV free wall (7.7%), and left atrium (7.7%). We used hypertonic saline solution for evacuation of fluid cyst. IABP insertion was used in 15.3%. Permanent pacemaker insertion was used in 7.7%. Hospital mortality was 7.7%.

Conclusion- There are a wide range of clinical signs and symptoms of cardiac hydatid disease depending on the site, size and shape of the cyst. There was not any advantage for CT-scan compared to echocardiography. Because Iran is an endemic area for hydatid disease, cardiac involvement should be a differential diagnosis of any cardiovascular symptom (*Iranian Heart Journal 2004; 5(4): 40-44*).

Key Words- cardiac hydatid disease ■ echinococcus ■ mebendazole ■ albendazole ■ CPB

Echinococcosis is an infection of humans caused by the larval stage of *Echinococcus granulosus* and *multilocularis*). The tapeworm species is endemic in many sheep-raising areas of the world and in Iran. This disease has both intermediate (sheep, cattle, human...) and definitive hosts (dogs). The adult *E. granulosus* is a tapeworm that resides in the jejunum of dogs. Eggs are passed in the stool and ingested by cows, sheep, and man...¹ After humans ingest the eggs, embryos enter the portal circulation and are filtered by the liver and occasionally by the lungs.¹

Cardiac involvement in hydatid disease is uncommon (<2%) and usually is intramyocardial in the interventricular septum and LV free wall.² A myocardial cyst may degenerate and calcify, develop daughter cysts, or rupture. Rupture of the cyst is the most dreaded complication; rupture into the pericardium may result in acute pericarditis, which may progress to chronic constrictive pericarditis. Rupture into the cardiac chamber may result in systemic or pulmonary emboli.³ Rapidly progressive pulmonary hypertension may occur with rupture of right-sided cysts.³ Surgical excision is generally

recommended, even for asymptomatic patients.¹ The liberation of hydatid fluid into the circulation may produce profound fatal circulatory collapse due to an anaphylactic reaction to the protein constituents of the fluid. Unless the disease is recurrent or inoperable, patients with cardiac hydatid disease must undergo surgery to avoid life-threatening complications such as cyst rupture, anaphylactic shock, tamponade⁴, pulmonary^{1,5,6,7}, intracerebral^{8,9} or peripheral arterial embolism,¹⁰ acute coronary syndrome,^{11,12} arrhythmias¹³ and infection.¹⁴ The efficacy of alternative medical therapies is not well established. The aim of this work was to study the clinical/pathological features and the indications and results of surgery over 11 years in our unit.

Methods

In this descriptive, cross-sectional study from March 1992 to July 2003, we had 13 proved cardiac hydatid cyst patients in the Rajaei Heart Center. There were five male and eight female patients, with a mean age of 36 ± 7.2 years. The patients were analyzed with regard to the demographics, clinical presentation, type of surgical resection, operative mortality, recurrence, late complications and long-term survival.

Clinical Features and preoperative evaluation: Our hospital is a referral hospital, so most of the patients had symptoms. Dyspnea was the most common symptom which was seen in 61.5%, dysrhythmia was seen in 30.7%, and 15.3% of patients were asymptomatic. Inverted T-waves in precordial leads were seen in 3 cases, and widening of QRS complex existed in another one. Casoni skin test was performed in 5 cases, of which 3 cases were positive. CXR, echocardiography and CT-scan were performed in all cases.

Operative technique: At operation the aim was to achieve complete clearance of hydatid cyst without uncontrolled rupture. At exploration, the pericardium was carefully packed with pads around the cysts to reduce the risk of pericardial soiling. The operation was performed with median sternotomy in all patients. Standard cardiopulmonary bypass (CPB) techniques using moderate hypothermia and cardioplegic arrest were used in 11 patients. 2 patients with subepicardial cyst and/or pericardial cyst were operated on without using CPB. The cysts were reached via ventriculotomy in 8 and atriotomy in 5, and hypertonic saline (10% NaCl), was used as scolicidal agent.

Results

Increased cardiothoracic ratio in CXR was seen in 8 cases and in 2 of them co-existent right side pleural effusion was found. Echocardiography showed cystic lesion in all cases and there was no superiority of CT-scan except for detection of other organ involvement. Cardiac hydatid cyst was isolated in 11 of cases and in 2 cases, liver involvement existed. The most common site of cardiac involvement was the interventricular septum in 6 cases (46%), followed by right atrium in 2 cases (15.3%), LV free wall in 2 cases (15.3%), pericardium in 1 case (7.7%), RV free wall in 1 case (7.7%), and left atrium in 1 case (7.7%). Mean cross clamp time was 56.55 ± 9.01 in min and mean pump time was 102.61 ± 21.18 in min. Weaning from CPB was done with inotropic support in 53.8% of the cases and with intra-aortic balloon pump in 15.3% of the cases. Permanent pacemaker implantation was used in one patient (7.7%). Hospital mortality was 7.7% (1 patient). This occurred in the operating room in a patient with anaphylactic shock due to rupture of hydatid cyst. Overall hospital stay was 25 ± 11.2 days (range: 15-55): ICU stay was

3.4±2.1 days²⁻¹⁰ and post-operative admission was 11.4 ±3.3 days.⁷⁻¹⁵ Mean follow up of the patients was 4.3 years (1-10±1.4). During this follow-up period, all patients had echocardiographic examination twice annually.

Discussion

Human echinococcosis is a zoonotic infection caused by the tapeworm of the genus *Echinococcus*. Hydatid disease is endemic in most sheep-raising countries in Asia, Europe, South America, New Zealand, and Australia.^{1, 15} Exposure to food and water contaminated by the feces of an infected definitive host or poor hygiene in areas of infestation can lead to echinococcosis.¹ Of the 4 known species of *Echinococcus*, *Echinococcus granulosus* is the most important and causes cystic echinococcosis (CE). The incidence of CE in endemic areas ranges from 1-220 cases per 100,000 inhabitants.¹ Morbidity is usually secondary to free rupture of the echinococcal cyst (with or without anaphylaxis), infection of the cyst, or dysfunction of affected organs. Examples of dysfunction of affected organs are biliary obstruction, cirrhosis,¹⁶ bronchial obstruction, renal outflow obstruction, increased intracranial pressure secondary to mass, and hydrocephalus secondary to cerebrospinal fluid outflow obstruction.¹ In CE, mortality is secondary to anaphylaxis, systemic complications of the cysts (sepsis, cirrhosis, respiratory failure), or operative complications. Many hydatid cysts remain asymptomatic, even into advanced age. Parasite load, the site, and the size of the cysts determine the degree of symptoms.¹ A history of living in or visiting an endemic area must be established. Also, exposure to the parasite through the ingestion of foods or water contaminated by the feces of a definitive host must be determined. Theoretically, echinococcosis can involve any organ. The liver is the

most common organ involved, followed by the lungs. These two organs account for 90% of cases of echinococcosis.¹ In CE, symptoms can be produced by mass effect or cyst complications. Symptoms due to pressure usually take a long time to manifest, except when they occur in the brain or the eyes. Most symptomatic cysts are larger than 5 cm in diameter. Organs affected by *E. granulosus* are the liver (63%), lungs (25%), muscles (5%), bones (3%), kidneys (2%), brain (1%), and spleen (1%). Secondary complications may occur as a result of infection of the cyst or leakage of the cyst. Minor leaks lead to increased pain and a mild allergic reaction characterized by flushing and urticaria. Major rupture leads to a full-blown anaphylactic reaction, which is fatal if not treated promptly. A rupture into the biliary tree can lead to obstruction by the daughter cysts, producing cholangitis. Rupture into the bronchi can lead to expectoration of cyst fluid.¹ Cardiac hydatid disease is seen in any age and sex group,^{2, 4, 17} although it is more common in those 20 to 40 years of age. In our study the female/male ratio was 1.6/1 and the mean age of the patients was 36 years. The clinical presentation varies, depending on the location, size and integrity of the cardiac cysts. In previous studies, the most common locations of cardiac echinococci cysts were the left ventricle (60%), and the ventricular septum (9% to 20%), but the right ventricle and right atrium can also be involved (4% to 17%).^{18,19} But in our study the most common sites of cardiac involvement were the interventricular septum (46%), followed by the right atrium (15.3%), LV free wall (15.3%), pericardium (7.7%), RV free wall (7.7%), and left atrium (7.7%). Isolated cardiac involvement was seen in 84.6% of our patients, but this may be due to the fact that we are a referral unit. The most frequent symptom was dyspnea, followed by dysrhythmia. Cardiac hydatid cysts may be fatal due to complications

such as cardiac failure, cyst rupture, embolization, etc. Early diagnosis and surgical treatment are of utmost importance. In our series, a negative serology test can not rule out the diagnosis. Eosinophilia is uncommon except after cyst rupture. In this study, echocardiogram provided definitive diagnosis in 100% of the cases.

Standard median sternotomy and cardiopulmonary bypass are well established modalities for surgical treatment of cardiac hydatid disease. However subepicardial cysts can be operated with off-pump technique. The basic steps of the procedure are eradication of the parasite by mechanical removal, sterilization of the cyst cavity by injection of a scolicalid agent, and protection of the surrounding tissues and cavities. Scolicalid agents include formaline, hydrogen peroxide, hypertonic saline, chlorhexidine, absolute alcohol, and cetrimide. A variety of complications have been described with all scolicalid agents, but in our experience, 10-20% hypertonic saline solution provides the best protection with the least complications. At surgery, the surrounding tissues are protected by covering them with scolicalid agent-soaked pads. The cyst is then evacuated using a strong suction device, and hypertonic saline is injected into the cavity. This procedure is repeated until the return is completely clear. This ensures both mechanical and chemical evacuation and destruction of all cyst contents. During this process, care is taken to ensure no spillage occurs to prevent seeding and secondary infestation. We had 1 death due to rupture of hadayid cyst and anaphylactic shock before use of cardiopulmonary bypass. We had no postoperative mortality. Postoperative progress is usually satisfactory and uncomplicated but involvement of the interventricular septum may result in complete heart block and need for a permanent pacemaker as it was the case in

one of our patients. Concomitant treatment with benzimidazoles (albendazole or mebendazole) has been reported to reduce the risk of secondary echinococcosis. Treatment is started 4 days preoperatively and lasts for 1 month. In CE, prognosis is generally good, with complete cure with total surgical excision without spillage. Spillage occurs in 2-25% of cases (depends on location and surgeon's experience), and the operative mortality rate varies from 0.5-4% for the same reasons.¹

Conclusion

Iran is an endemic area for hydatid disease; cardiac involvement should be included in the differential diagnosis of any cardiovascular symptoms. This study suggests that surgical resection of cardiac hydatid cysts offers a good chance of cure with acceptable operative mortality. Early treatment with drug like albendazole can prevent extension of disease. Serial echocardiographic examinations or other imaging modalities in the follow-up should be considered to detect recurrences.

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