Surgical Treatment of Cardiac Hydatid Disease in 13 Patients

Cardiac hydatidosis is an uncommon but potentially fatal disease. It remains endemic in developed and developing countries. Its clinical presentation ranges from an absence of symptoms to congestive heart failure or other life-threatening sequelae. Herein, we report our clinical experience with the disease.

From 1991 through 2009, 7 male and 6 female patients (mean age, 36 ± 18.3 yr; age range, 10–68 yr) underwent surgical treatment at our hospital for cardiac hydatid disease. Cardiac hydatidosis was established as a component of multiorgan echinococcosis in 8 patients, and it was diagnosed upon echocardiographic examination in the other 5 during investigation of their nonspecific symptoms. Hydatid cysts were found in the left ventricle (in 5 patients); the right ventricle (in 3); the interventricular septum (in 2); and the interatrial septum, right atrium, and left atrium (in 1 each). All 13 patients underwent sternotomy and surgery under cardiopulmonary bypass.

No intraoperative rupture or operative death occurred. The only sequela was complete atrioventricular block that necessitated pacemaker implantation in a patient whose hydatid cyst had involved the basal interventricular septum. All patients underwent subsequent treatment with albendazole (400 mg/d). One patient experienced a recurrence 1 year postoperatively. We discuss our surgical approaches, the outcomes in our patients, and diagnostic and therapeutic recommendations. (Tex Heart Inst J 2010;37(2):189-93)

Hydatidosis, a parasitic disease that is caused by the larval form of Echinococcus granulosus, remains endemic in countries where farm animals are raised, particularly in the Mediterranean region. The incidence of hydatid disease in Turkey has been reported as 3.4 cases in 100,000 people.

Cardiac hydatidosis has been reported infrequently even in countries in which hydatid disease is endemic: 0.5% to 2% involvement, in comparison with the liver (65%) and the lung (25%). Here, we report on our surgical treatment of 13 patients who had cardiac hydatid disease, and we provide diagnostic and therapeutic recommendations.

Patients and Methods

From 1991 through 2009, 13 patients underwent surgery for cardiac hydatid disease at our clinic. The ages of the 7 males and 6 females ranged from 10 to 68 years (mean age, 36 ± 18.3 yr). All 13 had lived in a geographic area where hydatidosis was endemic, and 4 patients raised sheep. All showed a variety of symptoms, the most common of which was dyspnea, in 5 patients. Two individuals required preoperative intubation due to severe ventricular tachycardia and ventricular fibrillation. In 1 of these patients, these symptoms caused intractable angina and mimicked acute myocardial infarction and left ventricular (LV) aneurysm. Intraoperatively, the presumed aneurysmal sac was found to be a hydatid cyst. In 8 patients, cardiac hydatid disease was diagnosed as a component of multiorgan echinococcosis, and 3 of these had undergone surgery for the treatment of previous cysts. The diagnosis of cardiac hydatidosis was confirmed upon echocardiography in the other 5 patients during investigation of their nonspecific symptoms. Cardiac hydatid disease was found in the LV in 5 patients (38.5%) (Fig. 1), in the right ventricle (RV) in 3 patients (23%), in the interventricular septum (IVS) in 2 patients (15.4%), and in the interatrial septum, right atrium, and left atrium in 1 patient each (7.7%). Cystic masses caused mild LV and RV outflow tract obstruction locally within the IVS in 2 patients. Electrocardiograms showed nonspecific changes. Telecardiographic findings were normal or showed alterations.
of the cardiac silhouette due to cystic masses on the cardiac walls. In 4 patients, the hydatid cysts were seen on thoracic computed tomography (Fig. 2) or on magnetic resonance imaging as finely bordered cystic lesions that had cuticular membranes. Each patient’s diagnosis was confirmed by the positive results of hemagglutination titers for echinococcus antibodies. Serologic tests yielded a variety of results and were not effectively diagnostic. Table I shows the clinical data on the patients.

**Surgical Techniques**

All 13 patients underwent sternotomy and were placed under cardiopulmonary bypass (CPB). The surgical approaches to the cysts varied, depending upon their location (Table I).

Of the 8 patients with ventricular wall involvement, 3 had a subepicardial cyst on the LV (Patients 2, 8, and 13). The cysts in the other 5 patients were subendocardial—with LV wall involvement in Patients 5 and 12 and RV wall involvement in Patients 7, 10, and 11. In these patients and in the 2 with IVS involvement due to invasion (Patients 4 and 9), surgical correction required full-thickness ventriculotomies. Cysts were punctured, aspirated, and resected in all patients. Before puncture, each cyst was covered with wet sponges in order to avert embolism and possible introduction of free scolices to other cardiac structures. After sterilization with hypertonic saline solution and needle aspiration of the cystic contents, cystectomy was performed. The cystic cavity was carefully opened, and the remaining cystic contents and the germinative membrane were removed. After cystectomy, the residual fibrous cavity that was supported by the LV wall (Fig. 3) was plicated with pledgeted sutures and closed via capitonnage (Fig. 4) in patients who had subepicardial involvement. The ventriculotomy incisions in 5 patients were closed linearly in standard fashion.
TABLE I. Clinical and Surgical Data on the Patients

<table>
<thead>
<tr>
<th>Pt. No.</th>
<th>Age (yr)</th>
<th>Sex</th>
<th>Symptoms</th>
<th>Other Organs Involved</th>
<th>Location of Cardiac Cyst</th>
<th>Size (cm)</th>
<th>Surgical Procedure</th>
<th>Concomitant Procedure</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>10/F</td>
<td>Fatigue and fever</td>
<td>—</td>
<td>RA base</td>
<td>3 × 2</td>
<td>Right atriotomy</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>19/M</td>
<td>Cough</td>
<td>Left lung</td>
<td>LV anterior wall</td>
<td>4 × 3</td>
<td>Cystectomy</td>
<td>Left-lung cystectomy</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>41/M</td>
<td>Dyspnea</td>
<td>Liver (prior operation)</td>
<td>LA posterior wall</td>
<td>3 × 2</td>
<td>Left atriotomy</td>
<td>CABG and muscular bridge resection</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>17/M</td>
<td>Chest pain</td>
<td>—</td>
<td>Intervertricular septum</td>
<td>5 × 5</td>
<td>Right ventriculotomy</td>
<td>VSD (iatrogenic) repair with patch</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>51/F</td>
<td>Palpitation and syncope</td>
<td>Liver</td>
<td>LV anterolateral wall</td>
<td>8 × 8</td>
<td>Left ventriculotomy</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>46/M</td>
<td>Dyspnea and chest pain</td>
<td>Brain, kidney (prior nephrectomy), right lung</td>
<td>Interatrial septum</td>
<td>2 × 3</td>
<td>Biatriotomy</td>
<td>Right-lung upper-lobe cystectomy</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>13/F</td>
<td>Dyspnea</td>
<td>Bilateral lungs (prior operation)</td>
<td>RV mural anterior wall</td>
<td>4 × 5</td>
<td>Right ventriculotomy</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>53/M</td>
<td>Dyspnea</td>
<td>—</td>
<td>LV mural anterior wall</td>
<td>5 × 6</td>
<td>Cystectomy</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>32/M</td>
<td>Dyspnea</td>
<td>Liver</td>
<td>Intervertricular septum</td>
<td>5 × 5</td>
<td>Right ventriculotomy</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>41/F</td>
<td>Fatigue</td>
<td>—</td>
<td>RV mural wall</td>
<td>4 × 6</td>
<td>Right atriotomy and ventriculotomy</td>
<td>Secundum ASD repair</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>69/M</td>
<td>Chest pain</td>
<td>Liver</td>
<td>RV inferior mural wall</td>
<td>3 × 4</td>
<td>Right ventriculotomy</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>54/F</td>
<td>Palpitation</td>
<td>—</td>
<td>LV intracavitary apical wall</td>
<td>2 × 3</td>
<td>Left ventriculotomy</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>28/F</td>
<td>Palpitation</td>
<td>Liver</td>
<td>LV mural apical wall</td>
<td>5 × 6</td>
<td>Cystectomy</td>
<td>—</td>
<td></td>
</tr>
</tbody>
</table>

ASD = atrial septal defect; CABG = coronary artery bypass grafting; F = female; LA = left atrial; LV = left ventricular; M = male; RA = right atrial; RV = right ventricular; VSD = ventricular septal defect

Patients 1 and 10 underwent a right atriotomy (the latter for atrial septal defect closure). Patient 3 underwent a left atriotomy, and patient 6 a biatriotomy. After cystotomy and cystectomy, the atriotomies were closed primarily. Concomitant procedures—such as the atrial septal defect closure in patient 10, pulmonary hydatid cyst extirpation in patients 2 and 6, and coronary artery bypass grafting and muscular bridge resection in patient 3 (Table I)—resulted in no adverse sequelae.

Results

There were no operative or late deaths. Patient 9 experienced the only complication—complete atroventricular block that necessitated pacemaker implantation. One year after the initial surgery, Patient 6 underwent reoperation for recurrent cardiac hydatidosis, this time on the anterior RV wall. The cyst was removed, and the patient recovered uneventfully.

Discussion

Human beings are exposed to echinococcus through direct contact with hosts or by eating contaminated, insufficiently cooked food. Embryos enter the portal or lymphatic circulation from the human intestine. Hydatid disease results from the expansion of parasitic cysts in the visceral organs. Cardiac involvement is uncommon. Larvae reach the myocardium through the coronary circulation. Cardiac involvement through the pulmonary veins has also been reported. The LV—the part of the heart with the most abundant blood supply—is most frequently involved (in 55%–60% of reported cases). Next are the RV (15%), the IVS (9%),
artery involvement has been reported to be 7% to 8%, and of pericardial location, 5%. In 1 case series, the most common sites of occurrence were the LV and IVS (and not the RV), apparently due to the high myocardial mass and good perfusion in both regions. In our series, the most commonly involved sites were the LV, the RV, and the IVS. Once the larvae reach the myocardium via the coronary arteries, cysts form in 1 to 5 years. Due to the slow growth of the cysts, cardiovascular disease was reported to be more sensitive and specific for liver hydatid disease. Involvement of the IVS can cause outflow tract obstruction or a variety of conduction defects. Another clinical feature includes impairment of valvular mechanisms, which mimics mitral, pulmonary, aortic, or tricuspid valve stenosis or regurgitation. Our patients had nonspecific symptoms, such as dyspnea, chest pain, fever, fatigue, and palpitations.

Clinical signs and symptoms vary according to the number, size, site, and effect of the cysts. As cysts grow, they are pushed toward a weaker side of the cardiac wall, either the epicardium or the endocardium. Hydatid cysts of the LV are usually located subepicardially and rarely rupture into the pericardial space. However, when they do, rupture may be silent, or it may cause acute pericardial tamponade, secondary pericardial cysts, or constructive pericarditis. The rupture of subendocardial cysts, which can also be silent, may cause anaphylactic reaction and sudden death, peripheral systemic embolism, or pulmonary embolism. In the RV, local intracavitary rupture is more frequent than in the LV and can cause pulmonary embolization, pulmonary hypertension, and death.

Cysts growing toward the epicardium can compress the small coronary arteries, disturbing blood flow. These signs may lead to the misdiagnosis of coronary artery disease. Involvement of the IVS can cause outflow tract obstruction or a variety of conduction defects. Another clinical feature includes impairment of valvular mechanisms, which mimics mitral, pulmonary, aortic, or tricuspid valve stenosis or regurgitation. Our patients had nonspecific symptoms, such as dyspnea, chest pain, fever, fatigue, and palpitations.

On the basis of such clinical variety, diagnosis is difficult. It is essential to consider cardiac hydatid cysts in a differential diagnosis, especially when a patient has spent time in a geographic area where hydatidosis is endemic. Electrocardiograms and chest radiographs can show alterations that are not specific for the disease. Serologic tests are more sensitive and specific for liver hydatid disease than for cardiac hydatid disease. On the other hand, the immunoglobulin enzyme-linked immunoadsorbent assay (ELISA) was reported to be the most sensitive (94%) and specific (99%) test for most cyst locations. In the same study, immunoelectrophoresis was positive in only 73% of cases. The immunoglobulin ELISA and indirect hemagglutination have proved to be the best tests for use in postsurgical follow-up.

Echocardiography remains the most reliable imaging method in the diagnosis of cardiac involvement and in locating cysts within the cardiovascular system. Patients with other systemic hydatid cysts should undergo echocardiographic evaluation for possible cardiac involvement. Indeed, most of our diagnoses of cardiac hydatidosis were made as a component of investigating multiorgan echinococcosis. Computed tomography and magnetic resonance imaging are other valuable diagnostic tools.

Early operative therapy is the treatment of choice for cardiac hydatid cysts. It is important to consider the location, number, and size of the cysts when choosing the operative approach and deciding whether to use CPB or to perform surgery on the beating heart. According to Birincioglu and colleagues, subepicardial cysts, regardless of their extent, can be resected without CPB with satisfactory results, provided that the cysts are not connected with the ventricular cavity. In contrast, we consider it safer to perform surgery with the use of CPB. Extracorporeal circulation and cross-clamping of the aorta prevent embolization into the systemic circulation and enable a direct view of the hydatid cyst and the cardiac structures. In operations for cysts that are located in the right side of the heart, the pulmonary artery can be clamped in order to avoid pulmonary embolism. As a rule, the heart should not be manipulated before applying the cross-clamp. It is crucial to minimize the possibility of contamination before cystectomy by first performing puncture and needle aspiration of the cystic contents and washing out the fibrous cavity with scolicidal agents (such as hypertonic solution, iodine solution, or alcohol). We use hypertonic saline solution for this purpose.

There are concerns regarding how to deal with residual cavities after removing the cystic membrane. Birincioglu and colleagues concluded that all residual cavities should be left open for self-closure via secondary healing because closure by suture may cause regional contractility and relaxation abnormalities, or even tears in the myocardium. We prefer cap tonsion after cystectomy in patients who have subepicardial ventricular involvement, and we observed none of the above adverse consequences in our patients who were thus treated. Postoperative sequelae include myocardial tearing, atrioventricular block that requires pacemaker implantation, ventricular arrhythmias, and sudden death due to ventricular arrhythmias caused by the scar. In an Anatolian literature review, Akar and colleagues revealed an overall mortality rate of 4.8%. All of our patients survived, and only 1 experienced an adverse sequela (atrioventricular block).

After surgical treatment, benzimidazoles are widely used to prevent the recurrence of cysts. These antiparasitic agents are also given to patients before surgery, in order to limit dissemination of the parasite during enu-
cleration. The main mechanism of benzimidazoles is reduction of parasitic growth.\(^8\) We prefer to administer albendazole at a dosage of 400 mg twice daily, starting at least 1 week before surgery and continuing postoperatively for at least 4 cycles of 28 days of treatment and 14 days of respite. Medical therapy may be extended beyond 4 cycles if the preoperative therapy was shorter than 1 week, if the disease is extensive, or if living parasites were encountered during the operation.\(^15\)

Despite the well-established efficacy of surgery in the treatment of cardiac hydatidosis, medical therapy may be the only option in some circumstances. Bozbuga and co-authors\(^21\) reported that a patient with cardiac hydatidosis refused surgery and was successfully treated with albendazole alone. Medical treatment might be recommended for small or heavily calcified cysts in elderly patients, or when surgery is otherwise contraindicated or is declined by the patient.

**Conclusion**

Patients who have cardiac hydatid disease may remain asymptomatic for years or experience minor symptoms, but the risk of fatal complications increases when the hydatidosis remains undiagnosed and untreated. Diagnosis is difficult on the basis of clinical symptoms alone. It is essential to consider cardiac hydatidosis in the differential diagnosis, especially when a patient has spent time in geographic areas where the disease is endemic. Early surgery is the treatment of choice even in asymptomatic patients. Typically, surgery for cardiac hydatid disease is safe and the results are satisfactory. Serial follow-up examinations by echocardiography and other imaging methods should be considered in order to detect recurrences.

**References**