

Unusual Presentation of Cardio-Pericardial Hydatid Disease

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Abstract

Objectives: To present here our experience in the management of cardiac hydatidosis in the form of five cases we admitted for management.

Methods: we included in this study five cases admitted to our center with unusual odd presentation of pericardio-cardiac Hydatid disease in the last eight years. The diagnosis of echinococcosis in heart was achieved through serological reactions, echocardiography, X-ray, computerized tomography (CT). These cases were managed both medically and surgically.

Results: 3 cases were managed surgically and 1 case was managed interventional and 1 case was managed only medically. 1 case died as a complication of ruptured intracardiac hydatid cyst and the other 4 cases cured and survived without recurrence within 1 year follow up.

Conclusion: *Echinococcus granulosus* can affect any organ in the body, and spillage during surgery can lead to a significant morbidity and recurrence. Cardiac HC is associated with a high risk of potentially lethal complications. Clinical manifestations and complications vary according to cyst's location and size.

Surgical treatment is highly recommended in the management of cardiac echinococcosis and considered in most of the cases as a medical emergency due to its high morbidity and mortality, but albendazole and/or praziquantel can be alternative option if the patient rejects surgery or if small hydatid cyst is present or if it is inoperable.

Medical treatment should precede and follow the surgical intervention and preventive measures are necessary to avoid recurrence.

Keywords: Cardiac Hydatid; Pericardial Cysts; Surgical Management

Introduction

Hydatid disease is caused by infection with the larval stage of the tapeworm *Echinococcus granulosus* or less frequently *Echinococcus multilocularis*. The adult worm is usually found in natural host dogs, cats or other canines; the tapeworm eggs are discharged in the animal's feces if ingested by humans through eating uncooked meat or via pets he will be infected [1].

The infection is most common in many parts in the world especially sheep-raising areas like Middle East countries, Turkey, Iran, India, Australia, South America and Africa [2].

Cardiac hydatidosis is extremely rare, occurs in 0.5-2% of cases. The most common sites of hydatid cysts are the liver (in 50% - 70% of cases), lungs (5% - 30%), muscles (5%), bones (3%), kidneys (2%), spleen (1%) and brain (1%) [3].

Morbidity from heart echinococcosis in men is three times higher than that in women [4]. Cardiac solitary cysts occur in almost 60% of the cases; the most frequent location is the ventricular myocardium and they are usually subepicardially located, this why they rarely rupture in the pericardial space [5].

It can present at any age and can be presented as completely asymptomatic or can cause obstruction in outflow tract, valves, and chambers of the heart, or great vessels, and can induce conduction disturbances such as atrioventricular nodal blocks, ventricular tachycardia and fibrillation, or cardiac tamponade [6]. Pulmonary embolism, anaphylactic shock, and systemic metastasis are some more important and catastrophic complications of cardiac hydatid cysts [7].

The diagnosis of echinococcosis in heart can be achieved through serological reactions, echocardiography, X-ray, computerized tomography (CT), and/or magnetic resonance imaging(MRI) [8].

Despite cardiac hydatidosis is very rare but it is potentially fatal. The most dangerous complication of cardiac echinococcosis is cyst perforation and rupture and if this happens; 75% of the patients die from anaphylactic shock, septic shock or embolic complications [9].

Once cardiac hydatid cyst is diagnosed must be considered as a medical emergency necessitates rapid investigation and urgent surgical management [10]. The risks during surgery is leakage of fluid from the cyst cavity leading to anaphylaxis and dissemination of infected scolices, which can be minimized by using scolicedal solutions such as iodine, hypertonic saline, methylene blue, or ethanol [11].

We present here our experience in the management of cardiac hydatidosis in the form of five cases admitted to our center over the last eight years.

Cases Presentation

Case 1

A case of 24-year-old man who presented with non-specific precordial chest pain and shortness of breath, right chest tightness of 5 months duration due to intrathoracic infestation with multiple hydatid cysts. The cysts were found in the anterior mediastinum, right lung, all aspects of right pleura.

His past history 9 months ago revealed that he had right thoracotomy for ruptured huge single pulmonary hydatid cyst after acute onset of cough with expectoration of salty dirty fluid, followed by progressive chest pain and SOB. His previous operation was complicated by prolonged air leak due to bronchopleural fistula which was managed finally with surgical closure via redo thoracotomy operation. He didn't receive postoperative chemotherapy.

On his admission to our hospital, clinical examination showed diminished air entry and restriction of the right hemithorax movement. The laboratory investigations were essentially normal, but the IgG for hydatid cyst HC was positive.

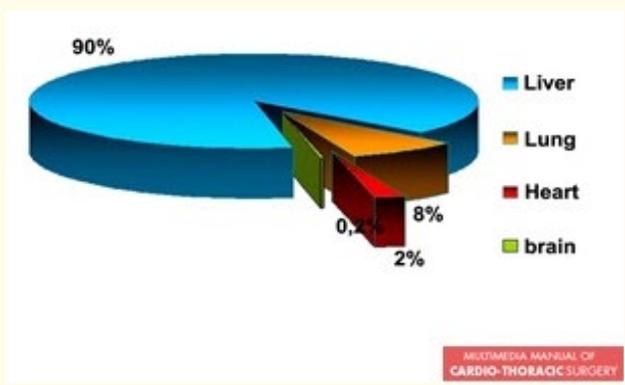


Figure 1: Showing percentage distribution of hydatidosis in different body organs.

CT scan chest showed multiple cystic lesions (Figure 2) varying in size, distributed in the mediastinum, right pleura, and lung. The pericardial HC are located on the right side (Figure 3).



Figure 2: Showing a disseminated thoracic HC.

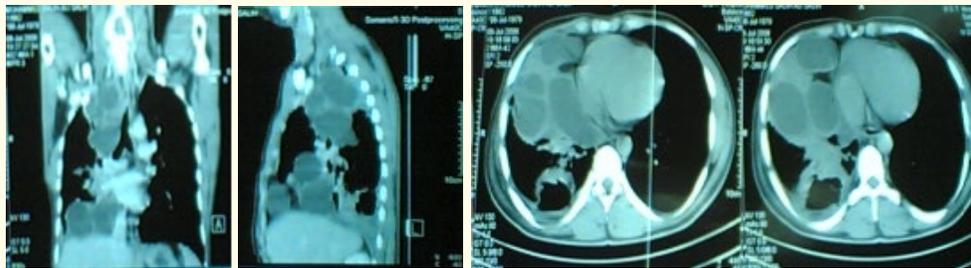


Figure 3: CT-scan coronal and sagittal images showing: epicardial and right hemithorax multiple hydatid cysts.

Albendazole 10 mg/Kg/day was initiated 4 - 7 days preoperatively. We approached via a median sternotomy. Operative exploration and careful dissection of mediastinal and pericardial adhesions showed pericardial and right pleural hydatidosis with average size 5 - 8 cm. Cystectomy of most of the HCs were performed. For large and adherent cysts we continued mobilization of the most prominent cysts, then puncture and vacuum suction was done. The Hydatid fluid and membrane (Figure 4) were removed, and the residual cavities washed with a 10% hypertonic solution and a 1% iodine solution several times.



Figure 4: (A) The cyst After opening and drainage. (B) The Germinal layer after enucleation.

Post-operative course was uneventful and he was discharged on oral Albendazole for 3 months.

The patient did not present with complications or recurrences after one year of follow up (Figure 5).



Figure 5: Chest X-rays one year after operation and 6 courses of chemotherapy.

Case 2

25 yr old male referred to us from the urology department after the incidental finding of asymptomatic cardiac mass on the CT examination of the abdomen which revealed right para-renal cystic lesion which was aspirated by CT guided needle.

Transthoracic echocardiography was performed and revealed Intracardiac cystic lesion (3.39 × 2.87cm), round, thin walled, lying at the base of the anterio-lateral papillary muscle of the heart, it contained some small cysts (Figure 6). It projects into the LV cavity and LV function was normal and there was no mitral regurgitation or flow obstruction. Although the echocardiography was mostly diagnostic for hydatid cyst, ELISA serum test was also positive for echinococcosis.



Figure 6: Showed hydatid cyst in the LV wall.

The patient refused surgical treatment so he was prescribed oral albendazole (10 mg/Kg/day) and instructed clearly about the risk of cyst rupture then was scheduled for follow-up with echocardiography. Upon 6 months of follow-up: trans-thoracic echocardiography showed disappearance of HC with residual thickness at its site suggested healed HC in comparison to the previous echo (Figure 7) and the 12 months "follow-up echo" showed complete disappearance of the cyst without residual abnormalities.



Figure 7: Follow-up Transthoracic echocardiograph (A) disappearance of HC (B) the previous echo with obvious intraventricular HC.

Case 3

A 65years old lady working as farmer complained of occasional atypical chest pain, palpitation, and vague symptoms as malaise, and fatigue with shortness of breath that started since 1 year. The patient had no history of previous cardiac disease or any risk factors leading to heart disease. There was history of contact with sheep and dogs. The chest radiograph showed a localized bulge at the apex of heart. The ECG was normal, Cardiac enzymes were within normal range. Transthoracic echocardiography was performed and revealed one large and well defined non-calcified cysts and one of them has intra-myocardial extension in the left ventricular wall while the other is a 3 × 5 cm cystic structure adjacent to the inferolateral wall of the left ventricle. The cystic mass was thin-walled and contained some small daughter cyst (Figure 8-A) without significant hemodynamic compromise. The serologic test (ELISA for IgG) was positive for echinococcal infestation.



Figure 8: Follow-up TTE shows hydatid cyst in the LV inferior wall (A)with evidence of size regression (B).

Further examinations with cardiac Multi-slice computed tomography (MSCT) scan confirmed the echocardiographic findings and location of the cyst, it demonstrated a hypodense lesion (5.8 x 5.4 x 6 mm) on the inferolateral region of the left ventricle. The lesion was located inside the pericardium and inside the myocardium of the apico-inferolateral wall of the left ventricle (Supplementary Figure 9).

Other works up showed no involvement of other organ system. The patient refused surgery so Albendazole 600 mg/day was prescribed for 12 weeks.

The follow-up echocardiography 8 weeks and 5months later showed that the cyst showed regression 2 x 1.5 cm in dimension, consolidation of the daughter cysts produce echogenic solid lesions and demonstrated albendazole-induced subtle echocardiographic changes

(Figure 8-B). But because the patient still complaining of chest pain and palpitation although she had good general improvement on medical treatment, she finally agreed for surgery. The cyst excised successfully and completely through median sternotomy without cardiopulmonary bypass, the base of the capsule which is attached to the myocardium is sutured to reinforce the myocardium at the cyst site and then she remained symptoms free during the 12-months period of follow up.



Figure 9: Contrast-enhanced CT sections showing giant multiloculated hydatid cyst in the inferolateral myocardium compressing the LV cavity and bulging into the pericardium.

Case 4

A rural 32 years-old woman, pregnant in 2nd trimester who was admitted to the emergency department due to cough with blood-tinged sputum for the last two weeks before admission, and complained of typical chest pain, numbness of the left upper limb, malaise, and fatigue with shortness of breath that had been exaggerating for last 3 months. There was history of sheep contact. The chest X-ray showed a localized bulge at the hilar level of the left lung and extends to the apex of heart. The ECG showed T wave inversion and ST depression in the lateral leads, Cardiac enzymes were within normal range. Transthoracic echocardiography was performed and revealed one large and well defined non-calcified cyst (3 × 5 cm) adherent to the lateral pericardial aspect. The cystic mass was thin-walled and contained some small daughter cyst and floating germinal layer (Supplementary Figure 10) moving with each cardiac beat but without hemodynamic compromise. The serologic test (ELISA for IgG) was positive for echinococcal infection. To avoid the radiation risk to the fetus, the cardiac CT scan was not performed.

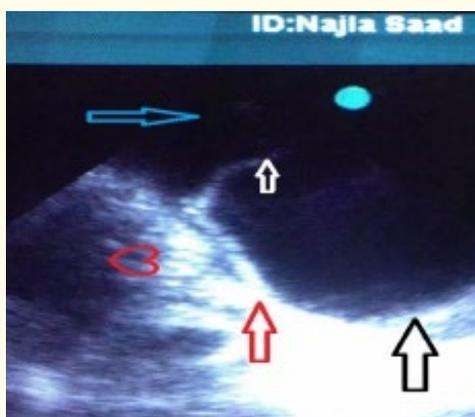


Figure 10: Two-D echocardiography: Showing the cyst (the black arrow) is attached to the pericardium (the red arrow), the floating germinal layer inside the cyst (the white arrow) was detached from the cyst wall (the blue arrow). The heart symbol indicate the LV.

After full discussion with the patient and her family regarding the potential teratogenic effect, Albendazole at a dose of 10 mg/kg for 7 days was administered preoperatively to decrease the adverse effect of cyst rupture as it has thin wall, moves with each cardiac cycle. The lesion was approached through a left posterolateral thoracotomy. The pericardial cyst was injected with hypertonic saline 20% and sterilized. The cyst wall was completely excised with the attached pericardium. The patient was doing well postoperatively and delivered a normal full term baby.

Case 5

23 years old male presented to Emergency Room in a hypotensive shock and sudden collapse preceded by vomiting. By examination: acute ischemia of both lower limbs could be detected. Echocardiography study revealed that the left ventricle cavity communicating with a cystic cavity in the lateral wall which was very thin. After resuscitation, chest and abdominal CT scan was done which showed a huge cystic lesion in LV wall communicating directly with LV cavity, obstruction of the distal abdominal aorta at its iliac bifurcation by a mass and embolic celiac artery trunk occlusion (Supplementary Figure 11).

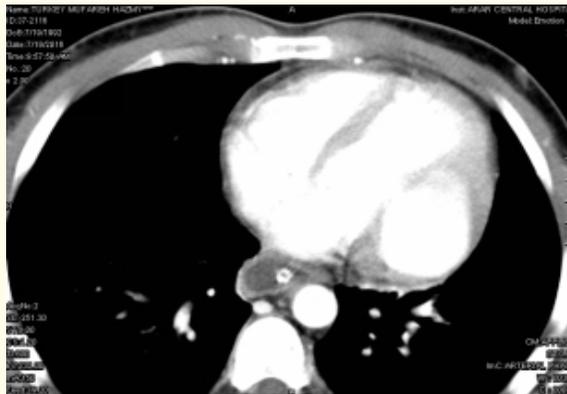


Figure 11: Showing huge hydatid Cyst communicating to left ventricle.

The case was diagnosed primarily as ruptured left ventricular hydatid cyst to the LV cavity, led to severe anaphylactic shock, visceral arterial embolization by the cyst contents and total embolic occlusion of the distal abdominal aorta by the laminated membrane of the cyst (ruptured germinal layer) which lead to acute ischemia of both lower limbs (Supplementary Figure 12).



Figure 12: CT showing the complete occlusion of distal aorta by the HC membrane.

After maintained stabilization of the hemodynamic status, embolectomy done through bi-femoral approaches and after many trails by endovascular snare and Fogarty catheter we could able to remove the obstructing tissue (i.e. the laminated membrane) in pieces (Supplementary Figure 13) but unfortunately, no back flow was coming from both femoral arteries and the lower limbs ischemia worsened and also the intestinal ischemia manifested and finally septic shock developed then the patient was declared dead after 24 hours from the diagnosis.



Figure 13: Showing the fragmented laminated member extracted from aorta by Fogarty Catheter.

Discussion

Although cardiac hydatid cysts can be fatal, they are rare, and often asymptomatic in their early stages [1,12] (like our case 1 who was asymptomatic and accidentally discovered). The growth of hydatid cyst is usually slow and just about 10% of patients with cardiac hydatid cyst are symptomatic therefore, clinical suspicion is important for a correct diagnosis [13]. Patients who have cardiac echinococcosis can present with a variety of clinical manifestations including atypical or typical angina (as in case 3, 4), conduction disturbances, arrhythmias, cardiac tamponade if the cyst ruptures in the pericardial sac, systemic embolic symptoms and anaphylactic shock if it ruptures inside left ventricular cavity (as what happened in case 5) [14].

Cardiac hydatid cyst should be considered and suspicious, in the differential diagnosis of patients with cardiac mass or with chest pain specially those who don't have risk factors for ischemic heart diseases, even for those who do not have a history of hydatid disease [15,16]. Furthermore, it should be noted that serological test is false-negative in 10% to 20% of patients with hepatic hydatid cysts, 40% with pulmonary cysts, and 50% with cardiac cysts [1].

Hydatid cyst with LV involvement can mimic left ventricular aneurysm and it should be one of differential diagnosis of cystic cardiac lesions especially in endemic areas [17] and this was the typical picture of case 5.

Hydatidosis involving the pericardium usually is part of disseminated hydatidosis and it occurs usually as a result of rupture of a primitive pulmonary hydatid cyst [17,18] as was in our cases 1 and 4.

Echocardiography is accurate for diagnosis and determining the number, location, and size of the cysts. CT and magnetic resonance imaging are used for diagnosis [18,19]. Coronary studies should be done in symptomatic patients or when electrocardiographic abnormalities are present [19]. Both Echo and CT were the main tools that defined the hydatidosis and concomitant extracardiac hydatid disease in our cases.

Some serological tests as indirect haemagglutination (IHA) test, ELISA and latex agglutination test can be done for diagnosis, screening and post-operative follow up for recurrence. The ELISA test for hydatid disease has a sensitivity of 91% and specificity of 82% [1,19]. All our 5 cases were serologically positive.

The treatment based mainly on surgical management with or without heart-lung machine and with or without aortic cross clamp. In epicardial and pericardial cysts, it is possible to carry out the operation under beating heart conditions [20].

Surgical excision was performed in case 1, 3, 4 combined with chemotherapy preoperatively and postoperatively aiming to decrease the recurrences. Enucleation (or extrusion) of solitary intact cysts remains our surgical treatment of choice because it completely obviates the possibility of dissemination through spillage. This can usually be performed safely and successfully in the case of epicardial cysts [21].

Risks at surgery from leakage of fluid include anaphylaxis and dissemination of the infected scolices can be minimized by clamping the main pulmonary artery and local instillation of scolicidal solutions like hypertonic saline or ethanol [7,21]. Common local scolicidal agents are solutions of hypertonic saline (10% - 20%), chlorhexidine, alcohol (80%), silver nitrate (5%), and iodine (1%) [22]. In our patients, after aspiration, the cyst was injected first with 10% NaCl solution to kill the daughter cysts and then the cyst was opened and the whole contents were sucked out and iodine was instilled into the cyst cavity. After removal of the cyst, the whole pericardial cavity was washed with hypertonic saline.

Cystectomy and pericystectomy remain the most useful surgical techniques. Pre and post-operative 10 days to 1-month courses of Albendazole and 2-weeks of praziquantel should be considered in order to sterilize the cyst, decrease the chance of anaphylaxis, decrease the tension in the cyst wall and to reduce the recurrence rate post-operatively [9,23]. However some authors don't prefer preoperative albendazole claiming that it enhances cysts rupture due to death of the larvae and thinning out of the germinal layer and the cyst wall [3,23].

The recurrence rate of this disease is still relatively high, accounting for about 10% [19,23]. The recurrence encountered in case 1 of our cases was "from our opinion" due to dissemination from ruptured old cyst and lacked of pre & postoperative albendazole. No recurrences 1 year after surgery and albendazole in cases 2, 3 and 4. Recurrence 2 years after treatment is uncommon [21,24].

The second option is medical therapy alone for a long time (minimum of two years) use of Albendazole, Praziquantel or combination of both and it is indicated in inoperable cases or after incomplete resection of lesions [25].

Two of our cases refused surgery so Albendazole was started and cardiac cyst disappeared after 6 cycles of chemical treatment, in case 2 and in case 3 there was decrease in the size and alteration of the cyst content showed by Echo and good improvement of the symptoms.

Continuous treatment is preferred and has been administered for periods of up to 2 years without significant side effects. During treatment, we should monitor aminotransferases, WBC, RBC, and hemoglobin monthly [26].

Conclusion

Echinococcus granulosus can affect any organ in the body, and spillage during surgery can lead to a significant morbidity and recurrence. Cardiac HC is associated with a high risk of potentially lethal complications. Clinical manifestations and complications vary according to cyst's location and size.

Surgical treatment is highly recommended in the management of cardiac echinococcosis and considered in most of the cases as a medical emergency due to its high morbidity and mortality, but albendazole and/or praziquantel can be alternative option if the patient rejects surgery or if small hydatid cyst is present or if it is inoperable.

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