Cardiac Echinococcosis Associated with Other Organ Involvement: Report of Two Challenging Cases

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INTRODUCTION

Echinococcosis is an endemic parasitic disease in which a dog is typically a definitive host harboring the tapeworm. Echinococcus granulosus happens when a ruminant becomes an intermediate host carrying the cyst. Humans become infected after ingestion of food and water contaminated by animal feces.[1,2] The echinoccocal embryo, by passing through the intestinal venous system, may reach the liver and then the systemic circulation, after which it could be carried to any bodily organ.[2] Although hydatid cyst can affect any organ, the liver is the most commonly involved organ (60%). The second most commonly affected organs are the lungs (20%-30%), while cardiac and brain involvement are extremely rare, reported in 0.5%-2% and 2% of cases, respectively.[1-4] Since cardiac echinococcosis is a fatal disease leading to life-threatening complications, early diagnosis and prompt surgical and pharmacologic treatment are of paramount importance.[1,2,4-7]

Herein we present two cases with cardiac hydatid cysts associated with other organ involvement, the role of multimodality imaging in diagnosis, and the challenges regarding surgical excision from the heart and lung in the first case and from the heart and brain in the second case.

CASE 1 PRESENTATION

A 31-year-old male was admitted to our institution with complaints of dyspnea on exertion and hemoptysis for the preceding 2 months. His past medical history was remarkable for lung hydatidosis 20 years ago, and he had been treated with albendazole at that time. His physical examination and laboratory data were unremarkable. The electrocardiogram showed normal sinus rhythm with right-axis deviation. The chest x-ray showed two round well-defined mass lesions in the middle lobe of the right lung (Figure 1).

Transthoracic (TTE) and transesophageal echocardiography (TEE) revealed a large intramyocardial multilobulated encapsulated cystic mass (4.8 cm × 2 cm) with partial thickening and calcification located in the free wall of the right ventricle (RV) adjacent to the tricuspid valve (TV) annulus (Figure 2 and Video 1). The mass extended into the RV inflow and the right atrium (RA), resulting in mild RV inflow obstruction with a mean pressure gradient of 4 mm Hg detected by Doppler echocardiography. The RV was mildly enlarged with moderate systolic dysfunction. Moderate pulmonary hypertension was also detected. Based on the echocardiographic findings and the past history of hydatidosis, cardiac echinococcosis was highly suspected in this case.

Cardiac computed tomography (CT) angiography was performed for further evaluation, which revealed a calcified cystic mass in the right side of the heart involving the TV annulus with projection into the RA and RV (Figure 3A). The right upper pulmonary artery was cut off from its origin. Furthermore, segmental branches of right and left lower pulmonary artery were obliterated. Both aforementioned findings were consistent with mixed chronic pulmonary thromboembolism and hydatid cyst embolization. The chest CT also revealed a tubular shape cystic lesion of the mediastinal side of the right upper lobe (Figure 3B) as well as two cystic lesions with marginal calcification and well-defined border (about 27 and 20 mm) in the upper and lower lobes of the right lung (Figure 3C). Abdominopelvic CT scan was negative for involvement of other organs.

After consulting with the cardiac surgery, thoracic surgery, and pulmonology services, surgical excision of the cardiac and pulmonary lesions in combination with perioperative chemotherapy with albendazole was planned.

After a median sternotomy and under cardiopulmonary bypass using bicaval cannulation and clamping of aorta, the RV inflow was approached through a right atriotomy. A round mass was discovered in the midpart of the RV along with the course of the right coronary artery and adjacent to the TV annulus protruding into the RA. Palpation revealed partial hardening of the mass. The hydatid cyst was localized by gauzes irrigated with 0.5% silver nitrate solution, and after injection of hypertonic saline solution, it was completely excised. Postpump intraoperative TEE showed complete resection of the cysts with normal RV inflow Doppler flow pattern. After termination of the cardiac operation, an attempt to access the lung cysts was made by the thoracic surgeon by releasing the lung adhesions. However, surgery was prematurely stopped due to drop of O_2 saturation and pulmonary hemorrhage. Unfortunately, despite massive blood transfusion and insertion of an extracorporeal membrane oxygenation system, the patient expired on the third day after the operation.

CASE 2 PRESENTATION

A 32-year-old male with a history of brain hydatid cyst who had undergone surgery 10 years before presented with exacerbated right-sided hemiparesis. Brain CT scan and magnetic resonance imaging (MRI) showed a large, round, sharply demarcated encapsulated lesion in the left hemisphere with moderate mass effect (Figure 4A and 4B).

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VIDEO HIGHLIGHTS

Video 1: Apical four-chamber view of TTE shows a large calcified multilobulated encapsulated cystic mass located in the free wall of the RV adjacent to the TV annulus (*yellow arrows*). Mild enlargement with moderate dysfunction of the RV is also obvious.

Video 2: Surgical excision of cardiac hydatid cyst.

Video 3: Prepump intraoperative TEE in long-axis views showing a large multicystic mass attached to the interventricular septum (*arrowheads*) with multilocular appearance (*asterisk*) highly suggestive for cardiac hydatidosis. *Ao*, Aorta; *LA*, left atrium.

Video 4: Prepump intraoperative TEE in the transgasteric short-axis view at the level of mitral valve (MV) revealing the multilocular hydatid mass (*red arrowheads*), which occupies a large portion of the left ventricular cavity.

Video 5: Live three-dimensional imaging of the hydatide mass (*red arrowheads*) during prepump intraoperative TEE in the midesophageal view. *LA*, Left atrium.

Video 6: Postpump intraoperative TEE imaging in the long-axis view showing the successful removal of the majority of the hydatid mass. The large echo-free space in the interventricular septum (*asterisk*) suggests a remnant of the cyst wall, and this space was freely connected with the left ventricular cavity. *Ao*, Aorta; *LA*, left atrium.

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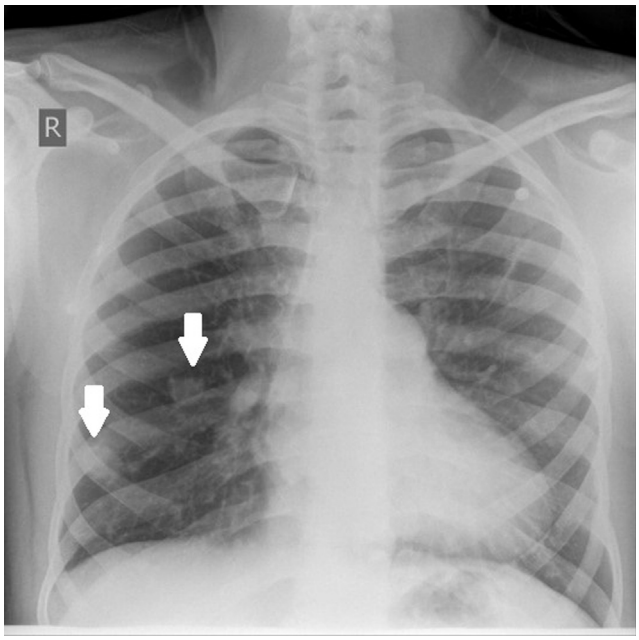

Figure 1 The chest x-ray of case 1 showing two round well-defined mass lesions in the middle lobe of the right lung (*white arrows*).

Abdominopelvic CT showed a cystic lesion in the great trochanter of femoral bone with periostial thinning and cortical disruption (Figure 4C). A multiloculate cyst was also noticed in the peritoneal cavity without calcification (Figure 4D). These cystic masses were suggestive of hydatid lesions. Seeking a cardiac source of the emboli, a TTE and TEE were performed, which were remarkable for severe left ventricle (LV) enlargement and dysfunction and also a large multicystic mass (8.5 cm × 6 cm in size) attached to the interventricular septum occupying a large space in the LV cavity with multilocular water lily appearance in at least two of the cysts, highly suggestive for cardiac hydatidosis mainly in active phases (Figure 5A and 5B). The tissue characterization by cardiac MRI was also in favor of this pathology (Figure 6). After presenting the case in the heart team meeting of our institution, it was decided to approach the cardiac involvement first as the potential source of embolization of the parasite to other organs. The patient underwent cardiopulmonary bypass with clamping of the aorta for surgical removal of the cardiac cysts (Video 2 and Figure 7). First, a bulging was noticed grossly on the anterior aspect of the RV in proximity to the left anterior descending coronary artery. After injection of hypertonic saline into the bulge, a right ventriculotomy was performed at the site of maximum bulging. The germinal layer was accessed, and many daughter cysts were removed. During the procedure, local invasion by the parasite was prevented by distributing sponges soaked with hypertonic saline solution throughout the pericardial cavity. Prepump and postpump intraoperative two- and three-dimensional TEE was performed (Videos 3-6). The overall general condition of the patient was markedly improved during the postop phase, and he was followed medically. The control TTE performed 2 months later showed no residual cyst. The patient was referred to the neurology service for further management of the brain cyst.

DISCUSSION

Cardiac echinococcosis is a rare manifestation of hydatid disease, which may result in catastrophic complications such as pericardial tamponade, pulmonary thromboembolism, anaphylactic shock, and even sudden cardiac death.[1,8-11]

Cardiac echinococcosis occurs more frequently in male patients, with a male-to-female ratio of 3:1.[8] The primary involvement of the heart usually occurs in two ways: the first route is defined as reaching the myocardium via the coronary circulation, and the second one is through the pulmonary veins carrying the ruptured pulmonary cyst.[3] The heart also can be secondarily involved by direct contact with the liver or lung hydatid cyst.[3] Any region of the heart could be involved. Considering the physiologic coronary distribution, the LV is involved more than the RV (60% vs 15%). Infestation of other parts of the heart is less common, including the pericardium (7%), pulmonary artery (6%), left atrial appendage (6%), and interventricular septum (4%).[1-3,6,8]

Right-sided versus left-sided hydatid cysts behave differently: right-sided cysts progress via intracavitary growth, whereas left-sided cysts tend to expand subepicardially.[3,8] Moreover, the rate of rupture into the RV is higher than into the LV. Consequently, RV hydatid cysts may be complicated with pulmonary embolism and anaphylactic shock. Rupture of the LV cysts into the pericardial cavity may lead to pericarditis or cardiac tamponade.[12]

Many cases of cardiac hydatidosis remain asymptomatic due to the prolonged latency period and slow growth of the cardiac cyst[3] but tend to present earlier than "classic" echinococcosis involving the

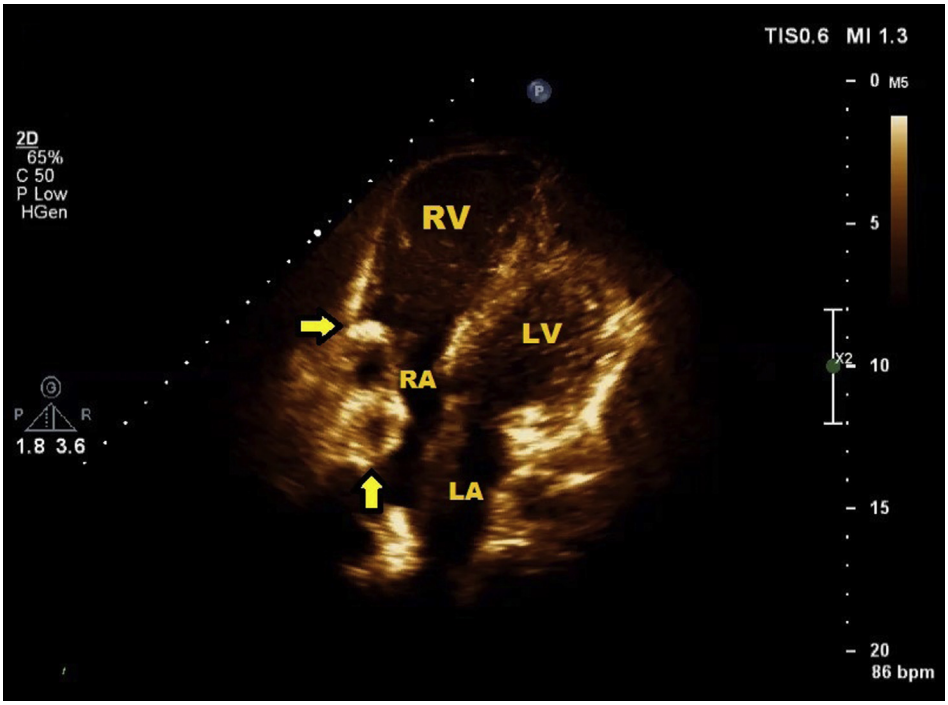

Figure 2 The apical four-chamber view of the TTE of case 1 showing a large partially calcified multilobulated encapsulated cystic mass located in the free wall of the RV adjacent to the TV annulus (*yellow arrows*). The more calcified parts suggest older lesions, and the less calcified parts suggest the more active ones; *LA*, Left atrium.

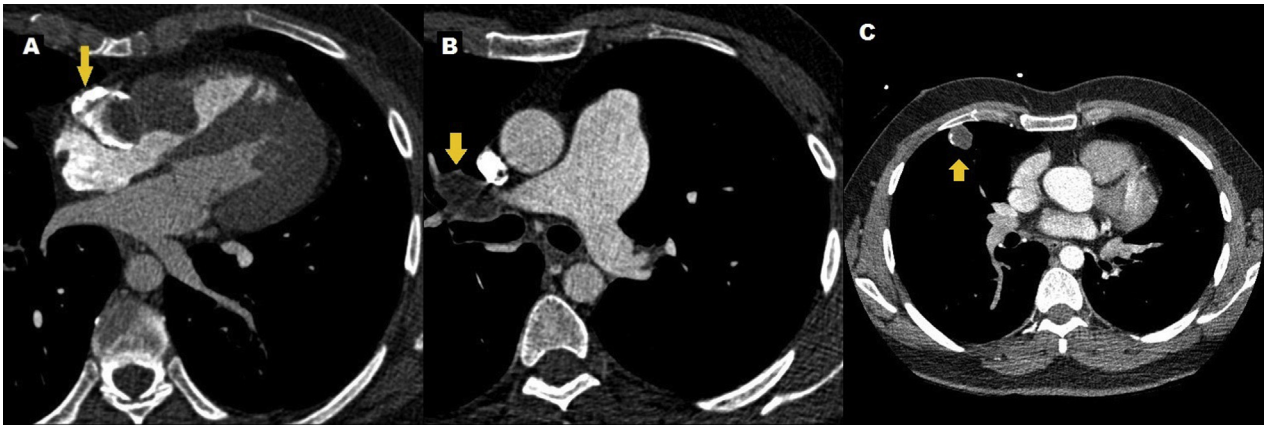

Figure 3 The pulmonary CT angiogram of case 1. **(A)** A calcified cystic lesion at the lateral aspect of the TV annulus with extension to the right atrioventricular groove (*yellow arrow*); **(B)** a tubular shaped cystic lesion in the right pulmonary artery (*yellow arrow*); **(C)** a unilocular and calcified cystic lesion in the periphery of the right middle lobe (*yellow arrow*).

liver/lung.[13] Some patients present with constitutional symptoms related to the infectious process such as fatigue, fever, and weight loss.[1] Cardiac symptoms are nonspecific and variable, based on the size and location of the cysts.[3] Chest pain, often atypical, is the most common cardiac symptom.[3] Rarely, a hydatid cyst may manifest with typical chest pain and ischemic electrocardiographic changes mimicking acute myocardial ischemia.[6] Other symptoms include dyspnea, palpitation, and heart failure.[3]

Signs and symptoms of cardiac echinococcosis may be related to its complications. The most life-threatening complication is cyst perforation, leading to systemic or pulmonary embolism, tamponade, anaphylactic shock, and late constrictive pericarditis. Acute pulmonary hypertension may occur following embolization of scoleces. Mechanical complications include ventricular outflow stenosis, valvular insufficiency or obstruction, and myocardial ischemia. Conduction disturbances or life-threatening arrhythmias are a consequence of interventricular septal masses. Sudden cardiac death has also been reported.[1,6]

Due to the potentially fatal complications of cardiac echinococcosis, in patients presenting with other organ involvement, screening

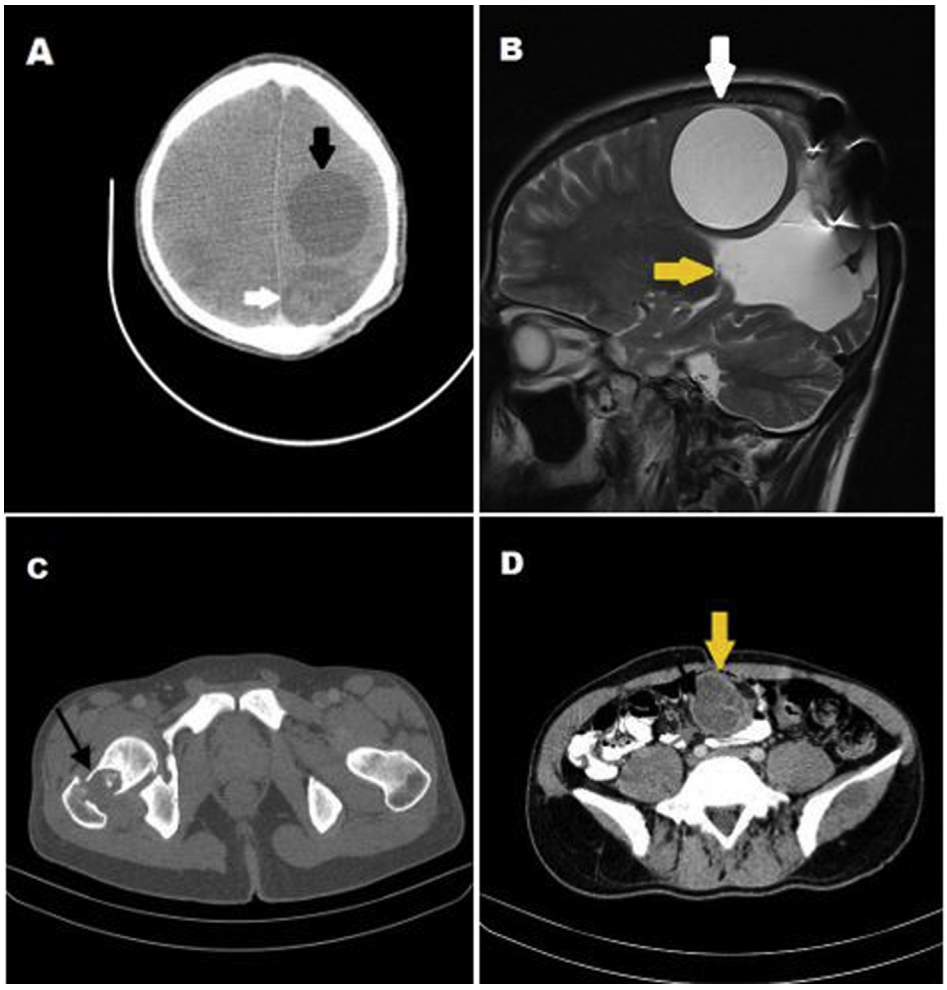

Figure 4 Extracardiac involvements in a patient with cardiac hydatidosis. **(A)** Brain CT showing a unilocular cystic lesion in the left parietal lobe (*black arrow*) and evidence of a previous craniotomy (*white arrow*); **(B)** brain MRI (a sagittal T2 weighted image) showing a unilocular and homogenous cystic lesion (*white arrow*) and brain malacia from previous surgery (*yellow arrow*); **(C)** abdominopelvic CT showing a cystic lesion in the great trochanter of the femoral bone with periostial thinning and cortical disruption (*black arrow*); **(D)** a multiloculated hydatid cyst in the peritoneal cavity without calcification (*yellow arrow*).

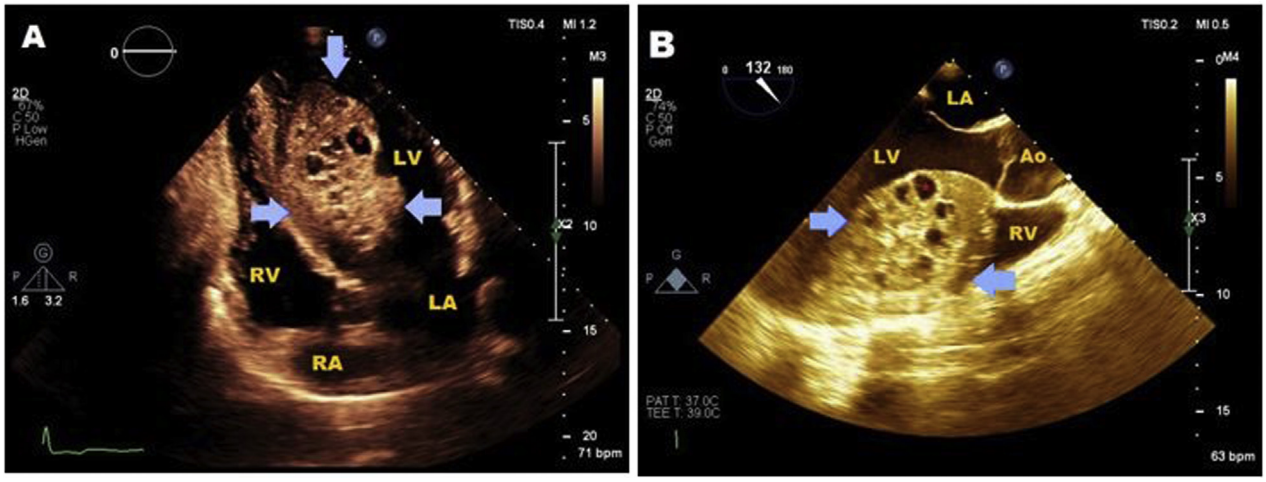

Figure 5 TTE **(A)** and TEE imaging **(B)** in apical and long-axis views, respectively, showing a large multicystic mass attached to the interventricular septum (*blue arrows*) with multilocular appearance (*red asterisks*) highly suggestive for cardiac hydatidosis mainly in active phases. *Ao*, Aorta; *LA*, left atrium.

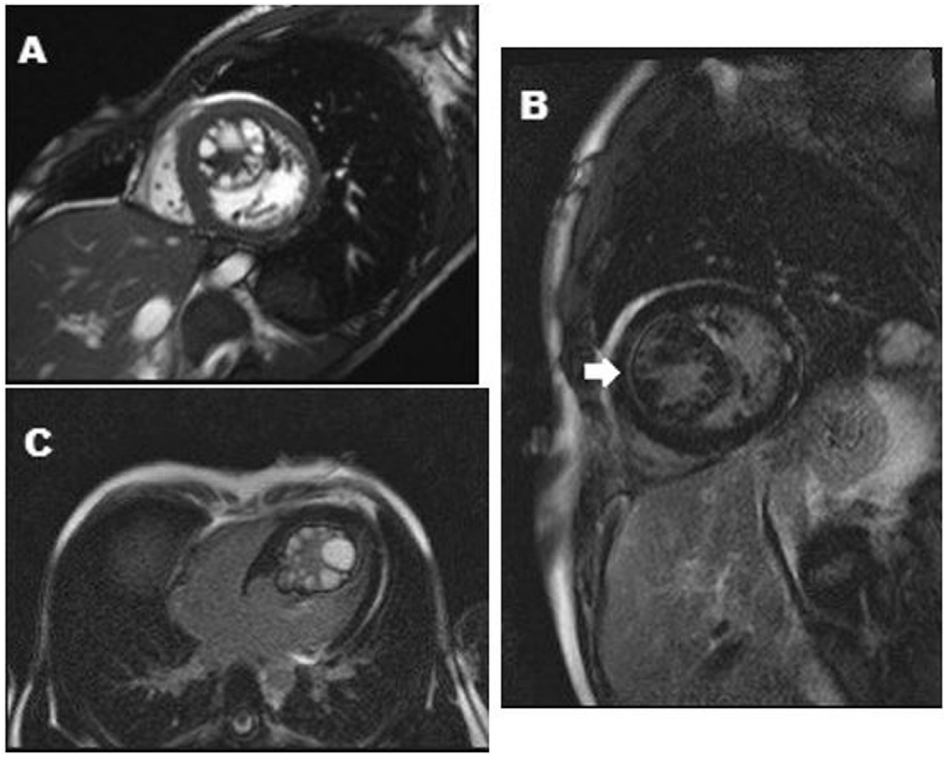

Figure 6 Cardiac hydatidosis demonstrated with cardiac magnetic resonance (CMR) imaging. **(A)** Short-axis SSFP sequence in CMR imaging shows multiloculated hydatid cysts; **(B)** late gadolinium enhancement sequence in CMR imaging shows pericystic enhancement (*white arrow*) in the septal wall and cystic lesion; **(C)** axial view T2 weighted image in CMR shows multiloculated hydatid cysts.

Figure 7 Excised hydatid cysts immediately after surgery.

for cardiac hydatidosis is mandatory.[1] Echocardiography, CT, and MRI can reveal the cystic nature of the mass and its relevance to the cardiac structures. Echocardiography is a simple, available, cost-effective, and highly sensitive method for detecting cardiac echinococcosis. Moreover, assessment of the hemodynamics of the lesion is feasible by echocardiography. Finally, compared with TTE, TEE has more accuracy.[1,8]

An echocardiographic classification system has been proposed for cardiac echinococcosis by Tufekcioglu *et al.*,[14] who define three types of lesions:

1. Active: characterized by a uni-/multilocular echolucent cyst with a double-layer wall.
2. Transitional: a shrunken cyst due to loss of intracystic pressure and showing the water lily sign.
3. Inactive: a completely degenerate cyst showing the ball-of-wool sign.

Assisting the differential diagnosis of cardiac cysts, CT and MRI are helpful imaging modalities and are needed in some cases in which echocardiography cannot establish the diagnosis.[6] "Double wall" is a specific sign of cardiac hydatidosis.[8] Wall calcification and presence of daughter cysts are also considered as specific signs.[3] In case of cardiac echinococcosis, MRI has a slight superiority to CT due to better determination of the ingredients of the liquid cysts. Conversely, calcified cysts can be reliably depicted using CT scan.[1] In our cases, we made the diagnosis by TTE, confirmed by TEE and CT.

Treatment options for cardiac echinococcosis include surgery and pharmacologic therapy with benzimidazole derivatives including albendazole and mebendazole.[1] The aforementioned drugs may slow down the cyst growth to a great extent[1] but cannot prevent rupture or embolization of cardiac cysts.[11] Hence, the treatment of choice for cardiac echinococcosis in both symptomatic and asymptomatic patients is curative surgery combined with pre- and/or postsurgery pharmacotherapy.[1,15] Recommendations on when to start pharmacotherapy before surgery are varied.[15] In our experience for the two presenting cases we used, the chemotherapy regimen before surgery included albendazole 400 mg daily for 14 days plus praziquantel 600 mg daily for 14 days.

CONCLUSION

Cardiac echinococcosis is a rare but potentially fatal manifestation of hydatid disease especially when complicated by brain or pulmonary involvement. Hence, it should be considered as a medical emergency. Antiparasitic agents with surgical cyst excision is the treatment of choice for cardiac echinococcosis. In our first case, cardiac surgery went very well, but pulmonary surgery was complicated by severe bleeding and massive transfusion. In the second case, cardiac surgery was performed first, and the patient was then referred to the tertiary care center for neurosurgery.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2020.09.008.

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