Giant Cardiac Hydatid Cyst with Rare Adhesions

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We present a 29-year-old woman who was admitted to the emergency department with shortness of breath. Using echocardiography, a giant multi-cystic mass was detected in the right ventricle, attached to the septal leaflet of the tricuspid valve and basal portion of the interventricular septum. Serologic tests (hydatid cyst antibody) confirmed *Echinococcus* infection. Lung computed tomography with intravenous contrast showed involvement of the pulmonary vasculature. The patient underwent cardiac surgery and the large cardiac cyst and the one in the right pulmonary artery branch were both removed. The tricuspid valve was also replaced by a bioprosthetic one. Albendazole was started preoperatively and was continued for six months after surgery. The patient recovered uneventfully and was followed up for one year. This is a report of a rare case of a very large cardiac hydatid cyst complicated by pulmonary embolism with attachments to both the tricuspid valve and interventricular septum.

Echinococcosis an infection that occurs in humans and is caused by the larval stage of *Echinococcus granulosus*, *E. multilocularis*, or *E. vogeli*. Echinococcosis is a severe health issue in cattle-raising geographical regions of the world. Cardiac hydatid cysts are found in fewer than 2% of cases of hydatidosis. In 50% of such cardiac cases, there is multiple organ involvement.1 The most common cardiac localizations are in the myocardium of the left ventricle wall. The endocardial infection occurs infrequently in comparison to the myocardial involvement. On the other hand, hydatid cysts may rarely develop within pulmonary arteries following ruptured cardiac or hepatic cysts.3

The clinical presentation in cardiac hydatid cyst is usually insidious, but early diagnosis and management are very important given the lethal hazard of cyst perforation. The importance of this case was the rare endocardial infection and the associated involvement of the pulmonary artery vasculature.

Case presentation

A 29-year-old woman was admitted to our hospital with a 4-month history of progressive dyspnea, precipitated by minimal exertion. She had lived in a rural community all her life, never smoked, and had no past medical history of note. On examination, she had sinus rhythm at 80 beats/min, blood pressure 110/70 mmHg, and respiratory rate 15 breaths/min at rest. Diastolic rumble (intensity: II/VI) was heard in the left lower sternal border. Other systemic physical examinations were unremarkable. Routine hematological and biochemical investigations were normal. The electrocardiogram showed sinus rhythm with complete right bundle branch block.

Echocardiography (Figure 1) showed a large, well defined and multi-cystic mass (5 × 4.7 cm) that had attachments to the...
septal leaflet of the tricuspid valve and the basal portion of the interventricular septum. It protruded into the tricuspid valve inflow, causing (Figure 1B-C) right ventricular inflow obstruction (mean pressure gradient: 4.6 mmHg). The right ventricle was severely enlarged, which raised the suspicion of associated pulmonary emboli; however, no masses were discovered in the main pulmonary artery or the proximal parts of its branches on the transesophageal echocardiogram. Estimated pulmonary artery pressure was 40 mmHg. On the chest X-ray (posterior-anterior), the cardiothoracic ratio was normal. The right hilum was prominent with a round, well circumscribed mass seen within it.

Lung CT with intravenous contrast showed a 40 mm filling defect within the heart in the right atrioventricular junction and the right atrium was enlarged (Figure 2A). In the right pulmonary artery an intraluminal fluid-density filling defect (average density=5 Hu) was seen, causing occlusion and expansion in the right pulmonary artery extending into the middle and inferior branches (Figure 2B-C). There were also multiple smaller fluid-density filling defects (average density=1 Hu) in the distal inferior branches of the left pulmonary artery. A small, focal sub-segmental atelectasis was seen in the right lower lobe. A 5 mm cyst was present in the posteriobasal segment of the left lower lobe (Figure 2D).

**Figure 1.** Transthoracic and transesophageal echocardiographic images. A. Large polycystic mass with attachment to the septal leaflet of the tricuspid valve and the upper part of the interventricular septum, protruding into the right ventricular (RV) inflow. B. Doppler study showing RV inflow obstruction. C-D. Transesophageal images showing the cyst with partially thickened capsule.
The serum level of *Echinococcus* IgG by ELISA was 144 RU/mL, which was considered as a positive serologic test for *Echinococcus* infection. Oral albendazole (400 mg/day) was administered and the patient was referred to the cardiac surgery unit for surgery under cardiopulmonary bypass. A right atriotomy was performed and the large cyst was seen. (Figure 3A). An infusion of 20% hypertonic saline was delivered into the cyst through a closed circuit. After a few minutes, the cyst content was aspirated. Hydatid sands (scolices) were seen in the fluid aspirated from the cyst. Once the cyst was opened, the germinal inner layer was removed, and finally the outer capsule was separated from its adhesion sites to the interventricular septum and the septal leaflet. (Figure 3B) There was no residual interventricular septal defect, but it was necessary to replace the tricuspid valve with a bioprosthetic one. The surgical field was then irrigated with a hypertonic saline.

Transesophageal echocardiography was applied intraoperatively to rule out any remaining cysts in the cardiac cavities. Subsequently, an incision was made in the right pulmonary artery branch and a cyst was removed under direct vision. (Figure 3C-D) Pathol-
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Histology confirmed the diagnosis. The patient had a complete and uneventful recovery, with no symptom recurrence during a one-year follow up. Albendazole was continued for six months after surgery.

Discussion

Cardiac echinococcosis is a rare entity, accounting for 0.01% to 2% of all hydatid infestations. The reason for such rarity could be the contractions of the heart, which provide a natural resistance to the presence of viable hydatid cysts.5

Although any part of the heart may be affected, the most common cardiac locations are the left ventricular wall (60%), followed by the right ventricle (10%), pericardium (7%), left atrium (6-8%), right atrium (3-4%), and the interventricular septum (4%).2 Endocardial cysts are extremely rare in comparison to myocardial and pericardial involvement.6-8

The disease can remain asymptomatic (90%), but it may cause displacement of the coronary vessels, rhythm disturbances and mechanical interference with the atrioventricular valves and ventricular function.6,7,9 Pulmonary embolism could complicate the

Figure 3. Intraoperative images. A. Gross view of the cyst after right atriotomy. B. Excision of the outer capsule with attachments to the septal leaflet of the tricuspid valve and interventricular septum. C. The white arrow indicates the incision made in the pulmonary artery branch. D. The black arrow shows the cyst removed from the proximal part of the pulmonary artery branch.
It may result from rupture of cardiac or hepatic cysts and may only be diagnosed postmortem. Transthoracic echocardiography showing the cyst with echo-negative contents and smooth contours is the most efficient method of diagnosing the hydatid cyst. Other diagnostic steps to be taken subsequently include CT scan and MRI, and the performance of serologic tests. In our patient, the diagnosis was suggested by transthoracic echocardiography, and confirmed by serologic tests and CT scan.

The increase in the right ventricular size and pulmonary artery pressure could not be explained simply by the presence of a right ventricular mass, though it had obstructive effects at the level of the tricuspid valve. The CT scan located the associated pulmonary embolism, whose detection was important because it could have affect the selected management strategies.

Surgery has traditionally been the principal definitive method of treatment in cardiac hydatidosis, even in asymptomatic cases, and the safest method is under cardiopulmonary bypass. Preventing cyst contamination of the surgical field is crucial. Multiple agents have been proposed as helminthicides: 2% formalin, 0.5% silver nitrate solution, 20% hypertonic saline solution, 1% iodine solution, or 5% cetrimonium bromide solution. There is no obvious superiority among these substances. It has been suggested that hypertonic saline solution-soaked pads be distributed within the pericardial cavity in order to prevent dissemination of the infection intraoperatively. Albenzazole, an active agent against *Echinococcus* infection, should be administered adjunctively. It is prudent to begin albenzazole several days before resection and continue it for several weeks after that.

**Conclusion**

Endocardial hydatid cysts are very rare. The clinical presentation is usually insidious. Echocardiography is an easy and safe method for the diagnosis. A CT scan could provide additional information about the involvement of other organs, including the pulmonary parenchyma and vasculature. The size, location and manifestations are the most important determinants of the management. Combined surgery and albenzazole therapy is the safest approach in cardiac hydatidosis.

**References**