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Acute Pulmonary Embolism due to Rupture of Pulmonary Valve Hydatid Cyst

Cardiac hydatid cyst is rare comprising 0.5–2% of all cases. A 20-year-old man was admitted for acute pulmonary embolism. Echocardiography and magnetic resonance imaging revealed hydatid cyst of pulmonary valve annulus. The cyst was drained surgically, and the patient was discharged with oral albendazole. For fatal complications of cardiac hydatid cyst, surgery is recommended in all patients.

Keywords: Echinococcosis, pulmonary embolism, magnetic resonance imaging, echocardiography

Introduction

Hydatid disease is a zoonosis mostly caused by *Echinococcus granulosus*. Human, sheep and ruminants are intermediate hosts, while dog or other carnivores are definitive hosts.¹ Cardiac hydatid cyst comprises only 0.5–2% of all cases of human hydatidosis.²

The most common location of cardiac hydatidosis is the left ventricle (75%), followed by the interventricular septum and right ventricle. Cysts in atria, pericardium and cardiac valves are very rare.²

Cardiac hydatid cyst may lead to congestive heart failure, valvular dysfunction or conduction disturbances.³ Herein, we reported on a patient with ruptured hydatid cyst of pulmonary valve annulus who developed acute pulmonary embolism.

Case Presentation

A 20-year-old man was admitted to our hospital with a history of exertional dyspnea and easy fatigability of two-month duration. He was a shepherd living in a rural area in the central part of Iran. He had hemoptysis and left-sided pleuritic chest pain since four days before admission.

On admission, he had an oral temperature of 38 °C and tachypnea. On physical examination, there was reduction of respiratory sounds in the base of the left lung. His electrocardiogram was normal at admission and during his course in the hospital. Chest roentgenogram revealed multiple pulmonary opacities and pleural effusion on the left side (Fig. 1). The patient was admitted with an impression of pulmonary emboli.

Lung perfusion scan showed multiple segmental defects throughout both lung fields with normal ventilation scan interpreted as high probability of pulmonary emboli (Figs. 2A and 2B).

The duplex scanning of pelvic and lower limb veins was negative for deep vein thrombosis.

Transthoracic in addition to transesophageal echocardiography revealed a

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Fig. 1. Opacities in bases of both lungs with left-sided pleural effusion.

multicystic mass with echogenic intracystic components originating from pulmonary valve extending to the right pulmonary artery, suspicious for complicated hydatid cyst (Fig. 3). There was a 20-mm Hg pressure gradient across the pulmonary valve.

Cardiac magnetic resonance imaging (MRI) demonstrated a cystic mass extending from pulmonary valve to the right and left branches of pulmonary artery (Figs. 4A and 4B).

Since there were neither clinical nor electrocardiographic signs of myocardial ischemia, coronary angiography was not performed.

The patient had eosinophilia (35%). Antibody titer against *Echinococcus* (ELISA) was 77 (positive > 20

AU. Abdominopelvic ultrasonography and computed tomography (CT) of brain were negative for hydatidosis.

After five days of therapy with 400 mg albendazole three times a day, cardiac surgery was planned.

The operation was performed through a median sternotomy. After induction of cardioplegia and dissection of the pericardium, the main pulmonary artery and its branches were dissected.

There was a large ruptured hydatid cyst originating from annulus of the pulmonary valve with extension to the pulmonary artery and its main branches.

To prevent more contamination of the surgical field, the cyst fluid was first aspirated and the cyst cavity was then sterilized by injection of 3% formaldehyde solution. The pericyst was then resected.

The patient was discharged from hospital on 14th day postoperative with oral albendazole, 400 mg, three times a day for six more weeks.

Histopathologic examination revealed a germinative membrane and scolices within a basophilic laminary structure consistent with hydatid cyst.

Discussion

Cardiac involvement in hydatid disease is either primary or secondary. Primary involvement of the heart occurs when the larva of *Echinococcus* reaches to the heart via coronary arteries.⁴ The larva invades the myocardium and reaches to adult form after one to five years.⁵

Primary cardiac hydatidosis comprises 0.5–2% of all infestations.³ The left ventricle—part of the heart which has the most blood supply—is most frequently involved (60–70%).^{2,4}

Fig. 2. Mismatch of ventilation (A) and perfusion (B) scans.

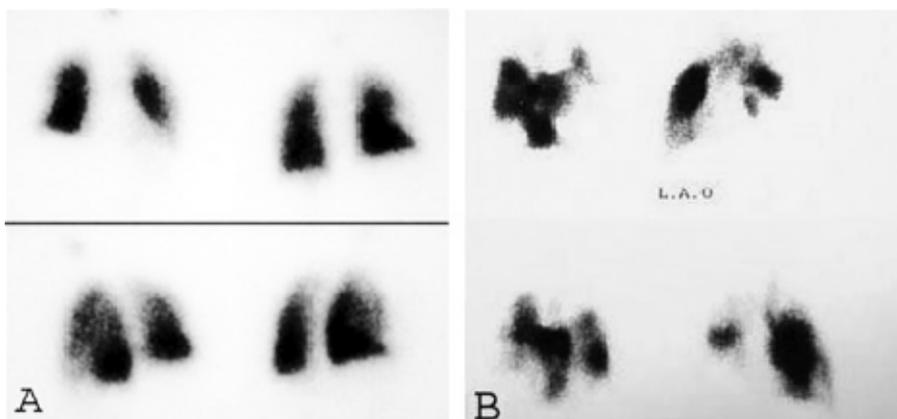




Fig. 3. 45° transesophageal echocardiography: Cystic mass with echogenic component in pulmonary artery with extension to the right branch.

Interventricular septum and right ventricle are less frequent locations. Involvement of atria and pericardium is very rare. In our patient, the origin of hydatid cyst was annulus of the pulmonary valve which, to the best of our knowledge, has not been described hitherto. Most of the patients are asymptomatic or have nonspecific symptoms.^{1,2} Specific symptoms depend on the location of the cyst: cysts of the left ventricle cause subepicardial ischemia and infarction, but cysts of the interventricular septum may cause conduction disturbances. Large cysts may cause obstruction

of cardiac chambers or valvular dysfunction. Rupture of the cyst to the pericardium may lead to tamponade, anaphylactic shock or pericarditis by daughter cysts. Constrictive pericarditis secondary to pericardial hydatid cyst has also been reported.⁶

Rupture of a cyst to cardiac chambers may result in anaphylactic shock, sudden death, pulmonary embolism or systemic dissemination of the organism.

The serologic test has high specificity. However, the test sensitivity is low in patients with intact cysts as the concentration of the antibody in serum is very low until the cyst leaks.

Transthoracic and transesophageal echocardiography constitute the most informative and efficient noninvasive diagnostic techniques. On echocardiography, a cyst with well-defined margins and internal trabeculation, corresponding to daughter cysts, is diagnostic of hydatid cyst.

Coronary angiography demonstrates displacement of the arteries around an avascular mass; there may be rim enhancement. CT and MRI are superior for evaluation of coexisting hydatid cyst of lungs or mediastinum. Calcifications are best seen on CT.

On MRI, hydatid cyst is usually characterized by a spherical lesion, which is hypointense on T1- and hyperintense on T2-W imaging. Low-signal rim in



Fig. 4. Coronal T1 MRI (turbo spin echo) black blood (A) and axial (turbo spin echo) white blood (B) images reveal daughter cyst in the hydatid cyst at the level of pulmonary valve with extension to the right pulmonary branch.

both T1- and T2-W imaging represents the pericyst and is characteristic of hydatid cyst.⁷ After contrast administration, ring enhancement may be detected. MRI is highly effective in evaluation of postoperative residual lesions, early discovery of recurrent lesions and detection of loculated or small pericardial effusion.⁸

Surgical excision remains the treatment of choice for cardiac hydatid disease. Oral albendazole therapy has been used to reduce the size of the cyst before operation and to prevent recurrence.⁶

Lahdhili has reported pulmonary embolism due to hydatid disease,⁹ but in that patient, the cyst was in the right ventricle not in the pulmonary valve. Arterial embolization has been reported from the left ventricular hydatid cyst as well.¹⁰

The primary intracardiac tumors are very rare with an incidence rate between 0.0017% and 0.19% in unselected autopsy series. Half of these tumors are myxomas. Rarely, cardiac hemangioma may appear as a cystic mass. These tumors could be considered as the differential diagnosis of cardiac hydatidosis.

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