Cardiac echinococcosis with intra-atrial localization

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Echinococcosis is a frequently encountered parasitic disease in the Mediterranean region, including Turkey. Cardiac disease is seen in 0.5-2% of patients. Usually the cysts are located within the pericardium or intramyocardially; intracavitary localization of the cyst is rarely seen. We herein report a patient who initially presented with hemoptysis and was echocardiographically diagnosed to have an intra-atrial hydatid cyst. Although patients with cardiac hydatid cysts may present with cardiac symptoms, symptoms typically involve other organ systems, following dissemination of the organism. Therefore, echocardiographic screening of patients who are diagnosed with echinococcosis, even if they have no cardiac symptoms, may ensure early diagnosis and prevent development of lethal complications, such as cyst rupture or embolization.

Key words: cardiac, hydatid cyst, children.

The larval stage of Echinococcus granulosus causes echinococcosis infection in humans. The liver and the lungs are the most common sites of the infection. Cardiac involvement in echinococcosis is infrequent. Clinical manifestations of cardiac echinococcosis are extremely variable depending on the size, number and localization of cysts, making the diagnosis problematic and challenging. In the pediatric age group, cardiac echinococcosis rarely becomes symptomatic due to the slow growth of the organism[1]. However, early diagnosis and definitive treatment are important to prevent lethal complications of the cyst, like rupture into the circulation or embolization to vital organs.

Case Report

A five-year-old girl (T.A.) was admitted with a five-month history of cough, fever and hemoptysis. In the prior two months she had been hospitalized and treated in two different hospitals with intravenous antibiotics for presumptive diagnosis of pneumonia. Following antibiotic treatment her symptoms had resolved partially but consolidations on the chest X-ray had remained unchanged. At this time the computed tomography scan of the thorax had demonstrated bilateral multiple nodular lesions in the lungs (Fig. 1). She was referred to our hospital for further evaluation. On admission her physical examination revealed a regular pulse with normal heart sounds and no murmurs. The only positive finding was the presence of clubbing. Remainder of the examination was unremarkable. Complete blood count, and renal and liver function tests were within normal limits. The peripheral blood smear demonstrated (25%) eosinophilia without any indications for malignancy. Erythrocyte sedimentation rate was 108 mm/hr. Nodular lesions in the lungs were interpreted as possible hydatid disease or metastatic lesions of a primary malignancy. However, serial examinations to find a primary site of malignancy failed. Bone marrow aspiration and biopsy were also negative. Echocardiography was performed to rule out cardiac involvement and revealed a non-homogeneous mass (259x214 mm) located on the posterior leaflet of the tricuspid valve (Fig. 2). The mass had a to-and-fro motion

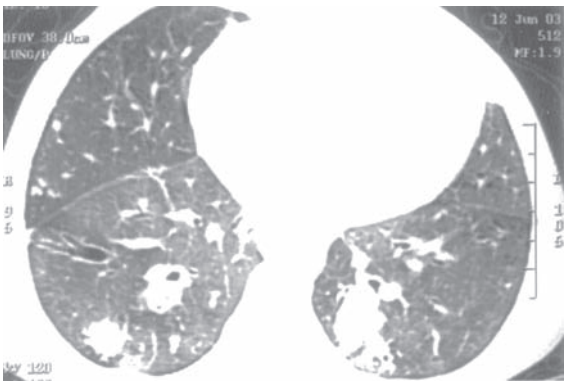

Fig. 1. Computed tomography scan of the thorax.

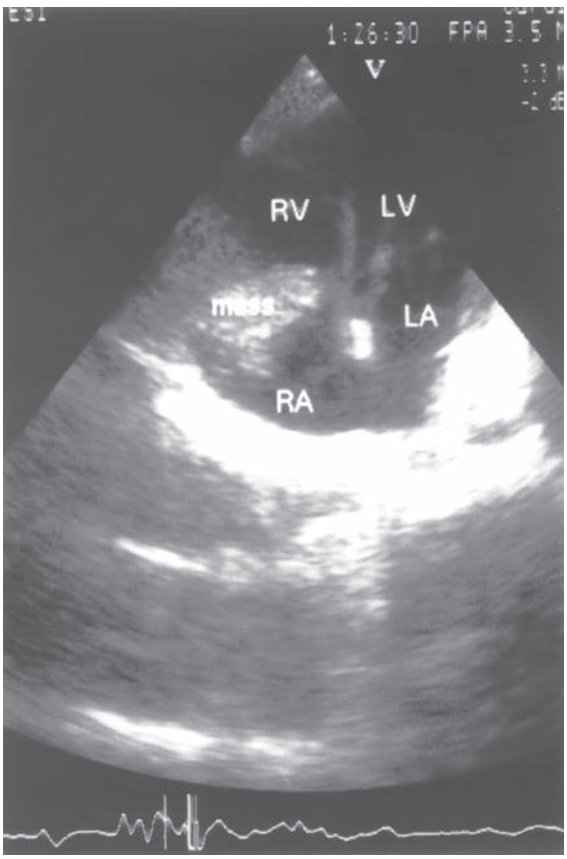

Fig. 2. Echocardiographic appearance of the intra-atrial mass.

through the tricuspid valve into the right ventricle. Electrocardiography of the patient did not disclose any abnormality. On further questioning, the patient was found to be from a region of the country endemic for echinococcosis. An indirect hemagglutination test for Echinococcus was strongly positive. Extensive imaging with computed tomography did not show involvement of any other abdominal or retroperitoneal organ.

Treatment with mebendazole was initiated and the patient underwent surgery for removal of the cardiac mass. Pathological examination confirmed the preoperative diagnosis of cardiac hydatid cyst (Fig. 3). Currently the patient is still on oral mebendazole for her lung lesions. She is asymptomatic and follow-up echochardiographic examinations are within normal limits.

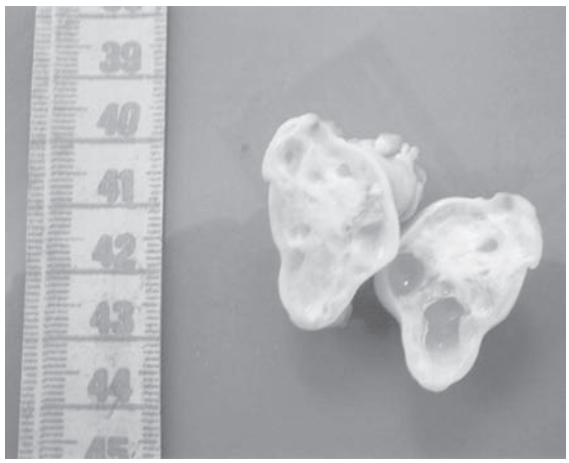

Fig. 3. Macroscopic examination of the cardiac hydatid cyst.

Discussion

Echinococcosis is endemic in sheep-raising areas of the world, particularly Argentina, Uruguay, Australia, New Zeland, Iceland and the Mediterranean basin[2]. The incidence of hydatid disease in Turkey is reported to be 1 in 2,000 children[3,4]. Cardiac hydatid cyst is an uncommon disorder, representing only 0.5 to 2% of all cases of hydatidosis[5,6].

Dogs that pass eggs in their feces are the definitive hosts of the Echinococcus. After ingestion of eggs by sheep, cattle, human and horses, cysts develop in these intermediate hosts. The organism is most commonly encountered in the liver or the lung. However, once in the systemic circulation, they can reach any organ, including retroperitoneal organs, pericardial cavity and the central nervous system[7,8]. Echinococcal cysts grow slowly and generally remain silent for years. They can cause symptoms in the affected organ by mass

effect. Anaphylactoid reaction can occur if the cyst ruptures into the circulation. Rupture of the ecchinoccoccal cysts can also cause further dissemination of the disease. Echinococcus embryos from coronary circulation or pulmonary veins can invade the myocardium[1,5,6,8,9]. The laminated membrane of the cyst becomes surrounded by the adventitial pericyst layer. The actual cyst develops in 1-5 years[1,5]. The density and configuration of the myocardium can sometimes limit the growth of the cyst. Hemodynamic and conducting function of the heart becomes progressively affected by the mass effect of the cyst, leading to myocardial ischemia, arrythmias and obstruction of the ventricular and atrial outflow tracts[5,6]. At this stage, rupture of the cyst can lead to embolization to vital organs. Localization of the cardiac cyst hydatid is most commonly intrapericardiac or intramyocardial[10,11]. Intracavitary localization of the cyst as seen in our case is very rare.

In our patient it is difficult if not impossible to determine the site of the primary localization of the disease. It is possible that the intra-atrial lesion could have been the primary site, the embolization of which resulted in pulmonary involvement. However, this is difficult to prove retrospectively. The presence of fever, cough and hemoptysis in the patient's history may support the theory of embolization from a cardiac source, although primary pulmonary disease with rupturing cysts may have caused these symptoms as well, especially since they apparently persisted for some time.

Atrial myxoma was the other disease considered in the differential diagnosis of our patient because of systemic symptoms and elevated acute phase reactants. However, eosinophilia, presence of pulmonary lesions and positive serology were indicative for echinococcosis. After surgery, pathological examination confirmed the diagnosis.

Nearly 90% of hydatidosis is clinically silent. Cardiac hydatid cysts should be suspected in patients from sheep-raising areas with signs of cardiac tumors or any mass close to the heart on chest X-ray[6]. Physical examination of the patient may be completely normal, although murmurs, arrhythmias or signs of heart failure, depending on the localization of the cyst inside the pericardial cavity, may be present. There may also be other systemic and organ-specific findings if the disease has disseminated. There is no specific electrocardiographic finding for echinococcosis. Echocardiography, magnetic resonance imaging and computed tomography scans are used for localization of the cyst. Echocardiography is superior to magnetic resonance imaging for demonstration of the cardiac cysts inside the dynamic structure of the heart. It can be more informative in regard to movement of the cyst, its relation to the beating heart and for impact of the mass on cardiac functions[12-14].

Because of the risk of rupture and embolization of cardiac hydatid cysts, medical treatment with mebendazole or albendazole alone without surgery is not acceptable. Some authors advocate the use of routine medical therapy as a supplement to surgery, and albendazole therapy is considered useful for reducing the incidence of recurrences[15,16]. Surgery has been proven as the best treatment for hydatid cysts of the heart[6,8,17,18]. Cardiac echinococcosis is usually seen as a component of disseminated disease involving more than one organ system[19]. Some authors suspect that the reported prevalence of cardiac echinococcosis would be higher if cardiac structures were routinely explored in patients with multiple hydatid cyst disease[6]. We also recommend routine investigation of all hydatid disease patients with two-dimensional echocardiography.

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