Lone, Mobile Left Atrial Hydatid Cyst

Echinococcosis is endemic in various regions of Turkey. Cardiac involvement in echinococcosis is rare, and lone cardiac hydatid cysts are even more unusual. Because cardiac hydatid disease can be fatal, even asymptomatic patients are optimally referred for surgical treatment. We present a rare case of a lone, primary, mobile hydatid cyst in the left atrium of a 62-year-old woman. The cyst caused dyspnea from left ventricular inflow obstruction. In addition to reporting the patient’s fatal case, we discuss cardiac hydatid cysts in terms of the scant medical literature. (Tex Heart Inst J 2016;43(3):261-3)

Hydatid disease is a parasitic infection caused by *Echinococcus granulosus*; human beings function as the intermediate host.¹ Echinococcosis, most prevalent in developing countries where sheep are raised, is endemic in Turkey.² In human beings, the parasite is most often found in the liver (in 65% of cases) and lungs (25% of cases).³ Because of myocardial contractions, the heart is rarely a target organ in which the echinococcal cyst will nest,⁴ and indeed cardiac involvement is observed in only 0.5% to 2% of patients who have hydatid disease. The typically affected cardiac sites are the left ventricular free wall (50%–77% of cases) and interventricular septum. Most cases have involved a single cardiac cyst. Most patients with cardiac echinococcosis also have multiorgan involvement, chiefly the liver and lungs. Symptoms depend on the location and size of the cyst. Advances in imaging and surgical techniques have contributed to remarkable progress in early diagnosis and treatment. We report the case of a woman in whom we found a lone hydatid cyst in a highly unusual cardiac location.

Case Report

In August 2014, a 62-year-old woman presented at our cardiology outpatient clinic with palpitations and severe dyspnea that had worsened during the last month. Her only other known medical condition was hypertension. Auscultation revealed an apical grade 1/4 diastolic murmur. Transthoracic echocardiograms (TTE) showed a unilocular, thin-walled, 90 × 60-mm cystic lesion attached to the interatrial septum at the left atrium. The patient’s eosinophil-to-leukocyte ratio was 1.9:1. Results of transesophageal echocardiography (TEE) and computed tomography (CT) confirmed the left atrial mass (Fig. 1). On TEE, the cyst was seen moving toward the mitral valve during atrial systole, leading to obstruction of left ventricular inflow (Fig. 2). Hemagglutination test results for echinococcosis were positive (1/3,200 IU). A peripheral-blood smear yielded predominant eosinophils. Abdominal ultrasonography and abdominal and cerebral CT were performed to detect the primary focus, but nothing unusual was reported. A CT scan of the thorax showed no cystic lesion other than that in the left atrium. Therefore, we concluded that there was no extracardiac primary focus, and the patient underwent surgical excision of the cardiac hydatid cyst. The cyst ruptured intraoperatively, and the patient died of anaphylactic shock. Histopathologic analysis revealed scolecies inside the cyst (Fig. 3).

Discussion

Cardiac echinococcosis was first reported at autopsy in a case by Williams in 1836,⁵ and the first antemortem diagnosis was made in 1905 by Cerné.⁶ Cardiac echinococcosis typically occurs as a component of multivisceral involvement. Of note, our patient had only one hydatid cyst, in an atypical cardiac site.
The age and size of a cardiac hydatid cyst and its degree of interior calcification determine the clinical presentation, which otherwise is not distinctive. Such cysts usually cause subjective symptoms such as difficulty in breathing, palpitations, and atypical chest pain. Our patient had palpitations and exertional dyspnea caused by left ventricular inflow obstruction. Cardiac hydatid cysts can lead to myocardial infarction, ventricular arrhythmias, pulmonary edema, or sudden cardiac death, because the enlarging cyst compresses the coronary arteries, cardiac conduction system, and intracardiac cavities. Rupture of the cyst can result in pulmonary or...
systemic emboli, anaphylactic shock, or sudden death. Rarely, pericardial tamponade can develop when the cyst ruptures into the pericardial space.

The Casoni test, Weinberg reaction, and measurement of eosinophils in the peripheral blood have been used in the diagnosis of echinococcosis; however, these have high false-negative rates. Although elevated eosinophil levels are themselves not a sufficient marker for the diagnosis of echinococcosis, they might indicate cyst rupture. Eosinophilia has no diagnostic value in patients who have no other clinical and radiologic signs of cardiac echinococcosis. The enzyme-linked immunosorbent assay and indirect hemagglutination tests, which have higher diagnostic value, have been in use together with Echinococcus-specific immune tests. The greatest progress in diagnosing cardiac hydatid disease has been enabled by TTE: the cysts are seen surrounded by a thin membrane, containing several septae and vesicles, and sometimes with daughter cysts. These features easily distinguish this formation from other cardiac masses.

Albendazole can be administered beneficially as therapy for cardiac echinococcosis. However, medical therapy cannot prevent cyst rupture and emboli, so it should be reserved for patients who decline surgical treatment or who are at high risk for open-heart surgery. The optimal treatment for cardiac hydatid cysts is surgical excision, especially of mobile cysts that might rupture.

Although echinococcosis is currently rare in developed countries, world travelers might facilitate its migration. Clinicians should be aware of the typical signs of cardiac echinococcosis and take advantage of advanced imaging techniques for diagnosis and treatment.

Acknowledgment

We thank Sonya E. Fogg, MLS, Manager of the Library and Learning Resource Center, Texas Heart Institute, for her research of our historical references.

References

5. Williams WH. Hydatiden in dem Herzen eines Kindes [in German]. In: Schmidt CC, editor. Jarsücher der in- und aus-