Introduction

Hydatid cyst in the heart is an uncommon pathology. The reported incidence ranges between 0.5% and 2% of all cases of echinococcosis in human (1). The cysts are commonly localized in the myocardium and pericardium (2). But pericardial hydatid cyst is seen excessively rare (3).

Clinically sudden death, valvular dysfunction, heart block, and more often chest pain have been described (4). We report a case of a 14-year-old boy with multiple hydatid cysts of pericardium and epicardium who died due to cerebral herniation caused by a large cerebral infarct after operation.

Case Report

A 14-year–old boy with chest pain, high fever, dyspnea, fatigue was referred to our department with a possible diagnosis of pericarditis. He had been hospitalized in a different centre with the diagnosis of pericarditis and had undergone percutaneous pericardiocentesis for diagnostic evaluation of pericardial effusion five years ago. During pericardiocentesis approximately 800 ml of fluid had been removed and he had been treated for tuberculous pericarditis.

On physical examination temperature was 38 °C, his heart rate was 116 bpm, arterial blood pressure was 110/60 mmHg. On auscultation of heart pericardial rub was heard. There were no jugular vein pulsations. Over lung fields respiratory sounds were coarse and diminished on the right lung. Laboratory analysis showed increased sedimentation rate (55 mm/h) and white blood cell count (13500/mm³). The result of an indirect haemagglutination test for echinococcus was positive (1/8192). A chest roentgenogram revealed cardiomegaly. Two dimensional echocardiography showed multiple cysts arising inside the pericardial cavity and pericardial effusion of 5-6 mm behind the left ventricular posterior wall. Computed tomography (CT) of the chest and abdomen showed multiple cysts in the anterior mediastinum, paratracheal and subcarinal regions, inside pericardial cavity and in the left kidney (Fig.1A-B). Brain CT was normal. The patient was given albendazole treatment (20 mg/kg/day) and was submitted to open heart surgery after ten days. Median sternotomy was performed, and multiple cysts surrounding his heart were demonstrated. Enucleation of cysts was planned without cardiopulmonary bypass (CPB) due to the concomitant pericardial and epicardial involvement. After sterilization with 30% hypertonic saline solution, many cysts were aspirated and daughter cysts with their germinative membranes were extracted (Fig 2). During postoperative period, mechanical ventilation was started in the intensive care unit. Left hemiparesis and agitation were observed in the patient on postoperative day one. Tonic-clonic convulsions were also observed on the 2nd day. Convulsions and agitation were controlled with clonazepam infusion and sedation. Inappropriate antidiuretic hormone (ADH) release syndrome was diagnosed on the day five and was managed with intravenous desmopressin. Repeat cranial CT revealed a large infarct in the right temporoparietal region. The patient died on the 12th day after surgery due to cerebral herniation.

Discussion

Echinococcosis is a widely known zoonosis caused by Echinococcus granulosus. The infection is more frequent in some areas of the world where sheep raising is common (5). In humans the most frequent locations of hydatid cysts are the liver (> 65% of cases), lungs (25 %) and renal (1-5 %), but they can be seen in any location including pericardium (6,7). Most of the patients with cardiac hydatid cyst may remain asymptomatic for many years or have minor nonspecific complaints (1,4,8). The clinical manifestation of cardiac echinococcosis depends on the location of the cyst. Although hydatid cysts are more fre-
quently located in the left ventricle, right ventricular hydatid cysts more frequently prone to rupture, leading to pulmonary embolisms, an anaphylactic reaction, or sudden death (9). Pericardial involvement in the cardiac hydatidosis is rare. Clinically, these lesions manifest by chest pain or signs of cardiac compression. If hydatid cysts rupture into the pericardial cavity, this may lead to effusion or cardiac tamponade and formation of secondary cysts (1,2,9).

In the presented case pericarditis was thought to be the initial diagnosis since chest pain and dypsnea were the main complaints. But, the diagnosis of cardiac hydatidosis was established by two-dimensional echocardiography. On echocardiographic examination multiple cysts were apparent inside the pericardial cavity. Then, we thought that probably a pericardial hydatid cyst was ruptured during the pericardiocentesis procedure done five years ago leading to formation of multiple daughter cysts surrounding his heart. Two-dimensional echocardiographic examination done five years ago revealed only pericardial effusion in this patient but not multiple pericardial hydatid cysts.

Transthoracic echocardiography, computed tomography and nuclear magnetic resonance imaging are successfully used in the diagnosis of cardiac hydatidosis (10). Transesophageal echocardiography (TEE), is a very helpful diagnostic procedure and can also be used to decide technique of operation in the cases of multiple hydatid cysts of epicardium and pericardium (11). Invasive techniques and interventional radiological procedure carry the risk of puncturing cyst (10). As a consequence, this can lead to anaphylactic reaction, sudden death and, as reported in our case, formation of secondary cysts. In the presence of pericardial effusion, possibility of a hydatid cyst should be kept in mind, because it may rupture during the pericardiocentesis.

Although effectiveness of high dose and prolonged use of benzimidazoles (mebendazole, albendazole) in the treatment of cardiac and non-cardiac hydatidosis has been advocated, this can not prevent serious complications (11). Therefore, surgical intervention is the definitive therapy for cardiac hydatidosis. In choosing the technique of operation the localization, number and size of cysts are important. Surgical treatment of pericardial hydatid cysts have been reported to be safe and efficient and usually performed using cardiopulmonary bypass (CPB) (4,11). However, pericardial and epicardial hydatid cysts can be removed without CPB (12). Birincioglu et al. (13) reported that cysts protruding towards the pericardial cavity and ventricular myocardial echinococcosis without relation with cardiac chamber can be opera-

Figure 1A: Computed tomographic scan of the chest shows multiple hydatid cysts surrounding the heart inside the pericardial cavity.

Figure 1B: Computed tomographic scan of the chest shows multiple hydatid cysts surrounding the heart inside the pericardial cavity.

Figure 2: Germinative membranes extracted during operation.
ted by the off-pump surgery with the aid of TEE and intraoperative surface echocardiography. In our case epicardial and pericardial hydatid cysts were also enucleated without CPB. But we could not have chance to use TEE and intraoperative echocardiography due to technical insufficiency. So, evaluation of sub-endocardial cysts or small cysts could not be performed. As a result, these unevaluated cysts may rupture during surgical manipulation and particles of these cysts drain into systemic circulation, causing cerebral embolic infarct.

Regarding this case, besides the selection of the technique of resection and type of operation; TEE and intraoperative surface echocardiography are also helpful in prevention of the possible surgical complications.

References


