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CASE REPORT

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Sudden Death Due to an Unrecognized Cardiac Hydatid Cyst: Three Medicolegal Autopsy Cases

ABSTRACT: Echinococcosis is a human infection caused by the larval stage of *Echinococcocus granulosus*. The most common sites of infection are the liver and the lungs. Cardiac hydatid cysts are very rare, even in regions where hydatic cysts are endemic (the Mediterranean, South America, Africa, and Australia). It has been reported that cardiac involvement is seen in about 0.5–3% of human echinococcosis cases. Three cases of cardiac hydatid disease that caused sudden death and which were histopathologically diagnosed are reported. Cardiac echinococcosis is rare, but due to its insidious presentation and affinity to cause sudden death, it is important that it be identified in the histopathological examination.

KEYWORDS: forensic science, forensic pathology, heart, hydatid cyst, sudden death

Echinococcosis, also known as hydatid disease, is common in several regions of the world, e.g., the Mediterranean countries, the Middle East, South America, East Africa, as well as some areas in Canada and Australia (1,2). The incidence and prevalence of this disease are estimated to be high in countries where sheep farming is widespread such as Turkey. Echinococcosis is a human infection caused by the larval stage of *Echinococcus granulosus*. The liver and the lungs are the most common sites of infection (3,4). Cardiac hydatid disease is very rare, even in endemic regions.

Cardiac involvement has been reported in approximately 0.5–3% human hydatid disease cases (2). Although cardiac echinococcosis is rare, its importance should be stressed, because of its insidious presentation and the possibility of sudden death.

Case Reports

Three sudden and unexpected deaths because of the cardiac involvement of hydatid disease are included in this study. The autopsies were performed between 1998 and 2004 at the Council of Forensic Medicine in Istanbul.

Case 1

A 16-year-old woman became ill at home and was taken to the hospital. Even though cardio-pulmonary resuscitation was carried out, the patient died. Her medical history did not reveal any prior significant health problems. At autopsy, the heart was found to be grossly normal, weighing 280 g (*N*: 247–327). Cross-sections of the heart and liver revealed a cyst in each, measuring $4.5 \times 4 \times 3$ cm and $6 \times 4 \times 2$ cm in diameter, respectively. The cysts contained a small quantity of clear fluid, and they were surrounded by thick hyalinized fibrous tissue. Loose and white germinative membranes were seen in the inner wall of the cyst. Microscopic examination of the cysts showed germinal layers and numerous scolices that confirmed the diagnosis of hydatid disease. Examination of the other organs did not reveal any remarkable morphological changes. No cysts were identified in other organs. There were no toxicological findings.

Case 2

The autopsy of a 39-year-old woman, who died suddenly at home, revealed multiple cysts throughout the atria and ventricles of the heart. The cysts measured between $2 \times 1.5 \times 1$ cm and $7 \times 5 \times 4$ cm in diameter. The heart was found to be enlarged, weighing 1150 g (*N*: 315–378). The right lung contained cysts with diameters between 0.5 and 1.5 cm. All the cysts were surrounded by thick fibrous tissue, and they contained clear fluid and germinative membranes, except for the largest cyst in the left ventricle. The largest cyst was surrounded by a thick fibrous tissue and had a colliquative center and large amount of necrotic debris and a degenerative germinal layer, surrounding the thick fibrous tissue with mononuclear cell infiltration (Fig. 1). Examination of the other organs did not reveal any remarkable morphological changes. No cysts were identified in other organs. There were no toxicological findings.

Case 3

The autopsy of a 28-year-old woman, who died suddenly, revealed a large cyst in the interventricular septum of the heart, with a diameter of $4 \times 4 \times 3$ cm (Fig. 2). The heart was found to be enlarged, weighing 500 g (*N*: 270–324). The cyst was filled with a

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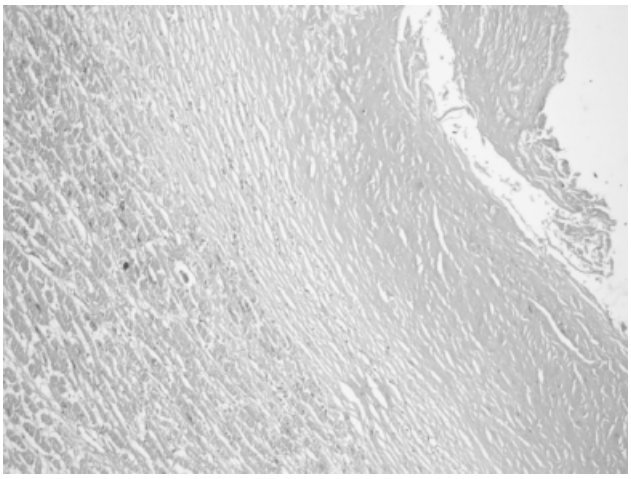

FIG. 1—*Myocardium, thick fibrous tissue, and colliquative center (HEX200).*

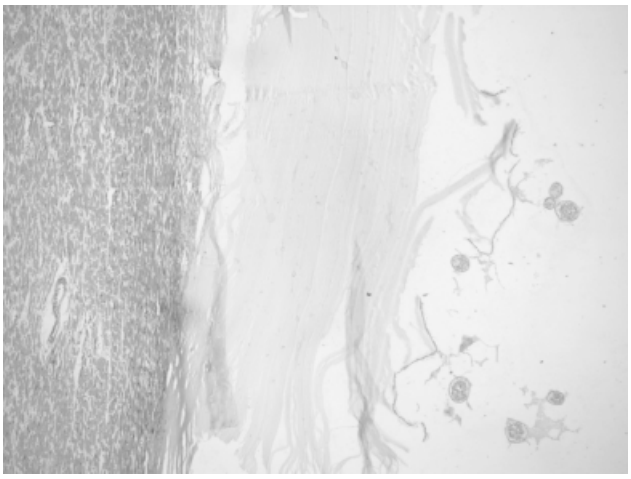

FIG. 3—*Myocardium, germinative membrane, scolices (HEX200).*

clear fluid, covered by a germinative membrane, and surrounded by a thick fibrous tissue. Microscopic examination of the cyst showed a germinal layer and numerous scolices that confirmed the diagnosis of hydatid disease (Figs. 3 and 4). Examination of the other organs did not reveal any remarkable morphological changes. No cysts were identified in other organs. There were no toxicological findings.

Discussion

Hydatid disease is a parasitic infection most frequently caused by the larval form of the tapeworm *E. granulosus*, which uses the dog as the definitive host (2,3). After ingestion, the larvae go through the duodenal wall to the portal blood system and into the liver, where they are found in about 60% of cases (5). Some of them (ca. 20%) may escape hepatic filtration and continue to the pulmonary circulation. A small percentage may reach the systemic circulation, resulting in infection and cyst formation in any organ (2,3). Isolated cardiac involvement by echinococcosis at any age is very rare and occurs in 0.5–3% of the cases (2). Isolated cardiac involvement was detected only in Case 3 of the present study. In the other cases, pulmonary and liver involvement was observed.

There is a strong supply of blood to the myocardium via coronary circulation. Cardiac cysts are commonly situated in the left ventricular free wall and interventricular septum because of their thickness and rich blood supply, although less frequent involvement of the right ventricle, atria, and pericardium have also been reported (6). Consistent with the literature, cysts were localized to the left ventricular wall and interventricular septum in Cases 1 and 3. Multiple cysts throughout the myocardium were found in Case 2.

Cardiac hydatid cysts are usually asymptomatic. They can display nonspecific symptoms such as chest pain, palpitations, and cough. The cysts usually grow slowly (1–5 cm a year) without causing symptoms (2). Myocardial atrophy may occur because of gradual expansion of the cyst. Probably only 10% of patients, especially those with large hydatid cysts, have clinical manifestations. Signs and symptoms of cardiac hydatic cysts are extremely variable and directly related to the location and the size of the cysts (7). No clinical signs or symptoms were present in the cases reported here. All deaths were sudden and unexpected.

The major causes of sudden cardiac death are coronary artery diseases, valvular diseases, cardiomyopathies, myocarditis,

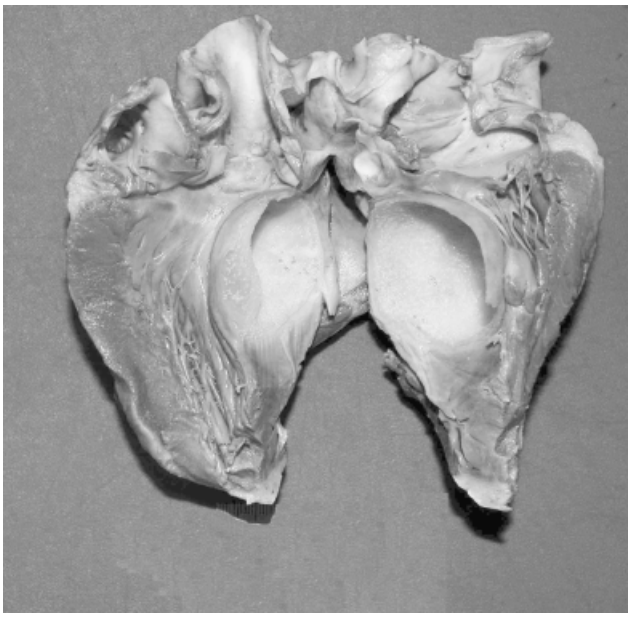

FIG. 2—*Solitary hydatid cyst in the interventricular septum.*

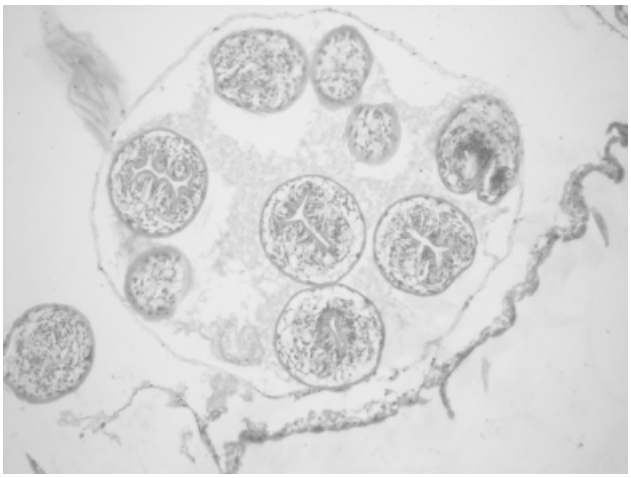

FIG. 4—*Germinative membrane, scolices (HEX400).*

idiopathic arrhythmia, mechanical causes such as aorta–ventricle ruptures, and commotio cordis (8,9). However, hydatid disease with cardiac localization also may be the cause of a sudden cardiac death, especially in developing countries.

Hydatid cysts grow slowly, and after 3–6 years they can reach the size of a chicken egg (2). Tamponade, acute pulmonary hypertension by embolization of several scolices, systemic arterial embolization, severe anaphylactic shock, and acute myocardial infarction are the major life-threatening complications of ruptured intra-cardiac cysts. Atrial and ventricular arrhythmias and complete heart block have also been reported (7). Cases of sudden death caused by cardiac echinococcosis are rarely reported in the literature, and in these, death has usually been attributed to cyst rupture (6,10). The complications of right ventricular hydatid cysts include fissuration or rupture with pulmonary emboli. Pansard et al. (11) reported a case of progressive fissure of a hydatid cyst of the right ventricle that led to a chronic pulmonary hypertension. This is probably secondary to microemboli migration in small vessels of the lung.

Malamou-Mitsi et al. (2) reported a case in which the cyst was found intact. They suggested that sudden death seems to be because of left ventricular fatal arrhythmias. None of the cases in the present study had macroscopic ruptures. For this reason, we hypothesized that fatal arrhythmias might be the cause of the sudden deaths.

However, anaphylactic shock was also reported in cases where there was no macroscopic rupture of the hydatid cysts. Therefore, in the management of this kind of case, the possibility of anaphylaxis should not be excluded (12,13).

If not diagnosed early, a cardiac hydatid cyst may prove fatal. Therefore, especially in developing countries, cardiac hydatid disease should be considered as a potential cause of a sudden cardiac death.

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