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CASE REPORT

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Sudden Death Due to an Unrecognized Cardiac Hydatid Cyst*

ABSTRACT: Echinococcosis is an endemic disease, most common in sheep-raising communities, usually caused by the larval or cyst stage of the tapeworm *Echinococcus granulosus.* Isolated cardiac hydatid cyst is uncommon at any age, occurs through the coronary circulation, and accounts for less than 3% of all hydatid disease.

We describe a case of an 18-year-old female, who died suddenly. The post-mortem examination revealed an isolated cyst in the left ventricle of the heart with intact wall. The cytologic examination of the cyst fluid demonstrated the presence of the characteristic scolices and hooklets and established the diagnosis of cardiac hydatid cyst.

The present case is of special interest because of the rare primary localization and the onset of sudden death in a young person as the initial manifestation of the disease.

KEYWORDS: forensic science, forensic cytopathology, heart, echinococcosis, hydatid cyst, sudden death

Echinococcosis, also known as hydatid disease, is common in several regions all over the world, including the Mediterranean countries, the Middle East, South America, East Africa, as well as some areas in Canada and Australia (1,2). It is a human parasitic disease, usually caused by the larval or cyst stage of the tapeworm *Echinococcus granulosus* and to some extent by *E. Multicularis* and *E. Vogeli* (3,4). Humans are infected by ingesting canine tapeworm eggs. The cysts are primarily localized in the liver and lung, but they can also be found in almost any organ (5,6). Primary cardiac involvement in echinococcosis is extremely rare, with a prevalence of 0.5 to 3% (6–8). The clinical presentation may be remarkably silent since the majority of these cysts are slow growing and may be without any symptoms for many years (9). We report the case of a primary cardiac hydatid cyst that caused sudden death in a young female and in which the diagnosis was established cytologically.

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Case Report

An 18-year-old female was found dead in her bedroom by her family and brought to the University of Ioannina for medico legal investigation. She was a student in high school and lived in the Greek countryside in a sheep-growing family. Her medical history did not reveal any prior significant health problem.

At autopsy, the body was that of a young female, 18 years old, 168 cm tall, and weighed 70 kg. The heart was found to be enlarged, weighing 420 g ($N \cong 233$ to 322 g), and the left ventricle wall was replaced by smooth, whitish tissue (Fig. 1). The left ventricular chamber was almost filled by a cyst (7 cm diameter) containing 120 cc of turbid fluid. The cyst wall was composed of an external layer of smooth whitish tissue and an internal one with a granular appearance. Microscopic examination of the cyst fluid showed several typical scolices with hooklets, as well as detached hooklets arranged in crowns or scattered in the background of the smears (Fig. 2), resulting in a definitive cytologic diagnosis of echinococcosis. Examination of the other organs did not reveal any remarkable morphological changes. No cysts were identified in any other tissues. Additionally, the toxicological examination was negative.

Discussion

Echinococcosis is a parasitic disease most frequently caused by the larval form of the tapeworm *E. granulosus*, which uses dogs as definitive hosts. The parasite lives its adult life in the intestinal tract of dogs. Ova are excreted and feces-contaminated material are ingested by sheep, pigs, and humans. Humans are accidental inter-

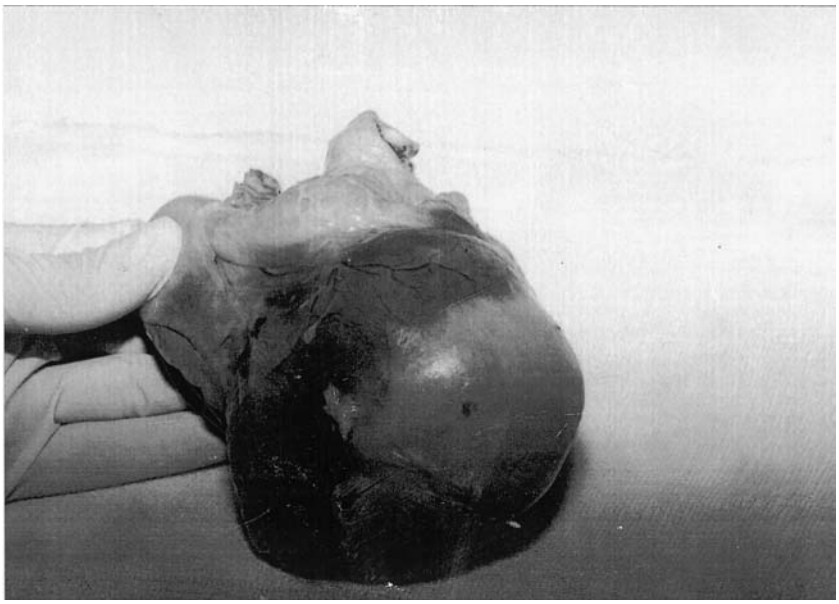

FIG. 1—*Autopsy findings: enlarged left heart ventricle. The wall is almost totally replaced by smooth whitish tissue.*

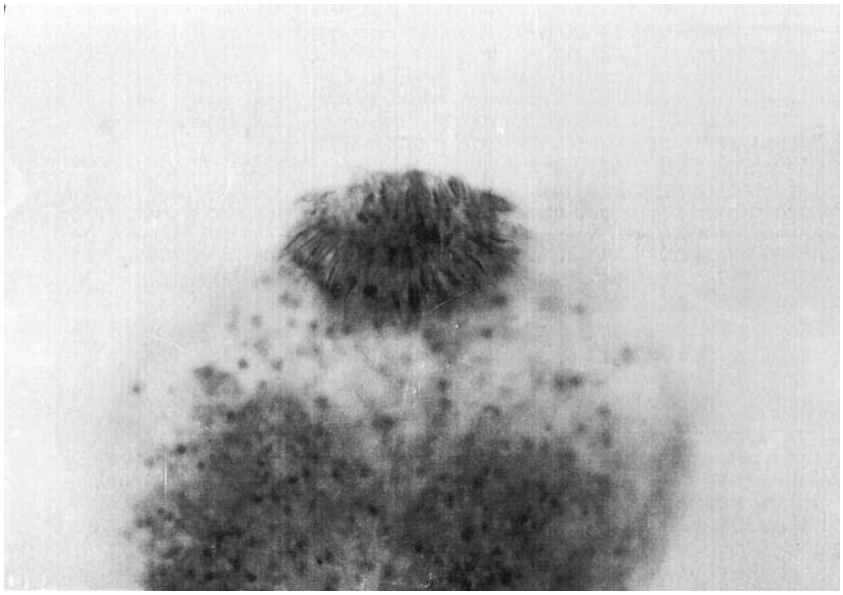

FIG. 2—*Cytologic smear: a typical scolex with a crown of hooklets at the anterior extremity (Papanicolaou stain ×1320).*

mediate hosts. After hatching in the duodenum, larvae penetrate the duodenum wall and enter the portal blood system through which they reach and infect the liver (60% of cases) (3). Some of them may escape hepatic filtration and continue to the pulmonary circulation (20%). A small percentage may reach the systemic circulation, resulting in infection and cyst formation in any organ (4,10).

Hydatid cysts grow slowly, and after three to six years they can reach the size of a chicken egg (11). For many years they may not cause any symptoms (12). Accidental rupture of the cyst may result in further dissemination of infection or trigger an anaphylactic reaction (13). Rarely, the cysts may undergo necrosis (14).

Cardiac involvement by echinococcosis at any age is very rare and occurs in 0.5 to 3% of the cases (6–8). The left ventricle is most often affected (50 to 60%). The cysts usually grow slowly (1 to 5 cm a year) without causing symptoms (7,11). Myocardial atrophy may occur due to gradual expansion of the cyst. Major complications, which are almost always fatal, result from rupture of the cyst and include anaphylactic shock, pulmonary or systemic embolization, pulmonary hypertension, or when rupture occurs into the pericardium, cardiac tamponade or pericarditis (15–17).

To our knowledge, the reported cases in the literature of cardiac echinococcosis and sudden death are extremely rare, and death has usually been attributed to cyst rupture (18,19). The present case is the first one in which the cyst was found intact and sudden cardiac death seems to be due to left ventricular malignant arrhythmias and/or pumping obduration (20).

The wall of a mature cyst consists of three layers: the pericyst, which derives from host fibrous tissue; the intermediate laminated membrane; and the inner, germinal layer, which produces daughter cysts (11). The cyst fluid contains scolices of *E. granulosus* and hooklets that can be identified in cytologic material and are pathognomonic of hydatidosis (6,21). Scolices are seen as polychromatic, oval structures bearing a crown of hooklets, which are sickle-shaped, refractile entities that can also be seen scattered in the background (5). In addition, it has been proposed that a diagnosis of echinococcosis may be considered when only laminated membranes are identified (22).

In our case, a definitive diagnosis was established by identifying in cytologic preparation of the cyst fluid several scolices and many hooklets arranged in crowns and scattered in the background.

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