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Hydatid cyst – a rare etiology of sudden death. Case report and literature review

Silviu Morar[1], Horaţiu Dura[1,], Adrian Cristian[1], Dan Perju-Dumbravă[2], Adrian Boicean[3], Mihaela Cernuşcă-Miţariu[4], Romeo Mihăilă[3]*

Abstract: Hydatid cyst is an extremely rare cause of sudden death, with only a handful cases being reported in the English speaking scientific literature. The purpose of this article is to present a case of an unexpected death of a young woman in full health, whose death was sudden and was proved to be caused by the rupture of a hydatid cyst.
The case, a 22 years old woman, was found in cardiac arrest by the Ambulance, and, based on the family statements, was suspected a mushroom poisoning. She was admitted in the ICU, where she died about three hours after. The autopsy showed no characteristic aspects of mushroom poisoning, but revealed the presence of multiple hydatid cysts of various sizes. The largest (7 cm in diameter) was devoid of content and presented a 0.5 cm long tear, amid a capsular parietal lesion (wall thinning). Adjacent to the tear, liver tissue presented inhomogeneous necrotic areas, situated on generalized dystrophic background. Toxicological examination invalidated toxic etiology and histopathology confirmed the diagnosis of hydatid cyst.
Key Words: sudden death, forensic autopsy, hydatid cyst, mushroom poisoning.

Echinococcosis is a parasitic disease caused by the larval stage of the cestode Echinococcus granulosus. The mature worm inhabits the intestines of the dog, and humans are accidental hosts in their life cycle [1]. The echinococcal cyst contains a inner germinal layer, an acidophilic, acellular membrane, and a host produced layer of granulomatous reaction. The germinal layer produces brood capsules that bud internally producing protoscolices through asexual division [2]. After ingestion of Echinococcus eggs, they hatch and release embryos in the small intestine, that they penetrate, reach the portal circulation, and from there can go in practically every organ, with the liver being however the most frequently involved [2].

Hydatid cyst is an extremely rare cause of sudden death, with only a handful cases being reported in the English speaking scientific literature [3-10]. The purpose of this article is to present a case of an unexpected death of a young woman in full health, whose death was sudden and was proved to be caused by the rupture of a hydatid cyst.

CASE REPORT

A 22-year-old female was found, by the Ambulance, in cardio-respiratory arrest. She was

1) "Lucian Blaga" University of Sibiu, Faculty of Medicine, Preclinical Department, Sibiu, Romania
** Corresponding author: Assist. Prof., Lucian Blaga no. 2A, 550169, Sibiu, Romania, Tel. 0040727238616, Fax. 0040269212320, Email: horatiudura@yahoo.com*
2) "Iuliu Haţieganu" University of Medicine and Pharmacy Cluj-Napoca, Forensic Pathology Department, Cluj, Romania
3) "Lucian Blaga" University of Sibiu, Faculty of Medicine, Medical Clinical Department, Sibiu, Romania
4) "Lucian Blaga" University of Sibiu, Faculty of Medicine, Dentistry and Nursing Department, Sibiu, Romania

unconscious, cyanotic, with, fixed mydriasis and sphincter relaxation; no injuries were detected. The medical history suggested a possible acute mushroom poisoning.

On arrival at the hospital she was hemodinamically stable, in a first degree coma, with cyanotic extremities, normal thoracic conformation, normal tactile fremitus, no rales, rhythmic heart sounds (heart rate 81 beats / minute), BP 128/54 mm Hg, no abdomen outline painful to palpation, normal abdominal transit.

She had overall increased clotting times (prothrombin time PT increased to 39.9 seconds, low prothrombin activity - 21.9%, an increased INR up to 3.55, APTT - partial activated prothrombin time - increased to 126.3 seconds), hepatic cytolysis syndrome (SGOT 247 U / L, SGPT 256 U / L), azotemia (creatinine 1.58 mg / dL), severe metabolic acidosis (pH 6.5 - 6.9 even when bicarbonate was administered). Due to these findings and the patient's history, the suspicion of mushroom poisoning persisted and the medical staff proceeded to sample gastric content for the toxicological examination.

The neurological consultation showed no signs of meningeal irritation, a surpassed coma flaccid quadriplegia presenting bone tendon and plantar areflexia. She died about three hours after the admission, in cardiopulmonary arrest.

Autopsy findings

External examination found no signs of violence, nail bed cyanosis, generalized edema (especially at the level of the face), pink aerial foam around the nostrils.

Internal examination revealed a nonspecific pathology, bearing a semblance of hyper hydration, bleeding syndrome and an acute pulmonary edema. The liver had a normal volume, brittle, presenting capsule integrity and a smooth shiny surface; it had a reddish-brown section coloring with vague yellow spots and unevenly scattered blackish red dots, in some places having a confluence tendency. The section of the right hepatic lobe revealed a number of cystic formations. Their diameters ranged between 0.5 and 7 cm, displaying a fibrous wall and containing a clear fluid kept under pressure. One of these formations (about 7 cm in diameter), which was visible on the diaphragmatic hepatic surface, subcapsular, was devoid of content and presented an unevenly thinned wall area (of about 1 cm in diameter) on the opposite of the diaphragmatic hepatic face. This area was centered by a solution of continuity with slightly irregular borders, about 0.5 cm in length. The liver tissue adjacent to this area had a lower consistency and a decrepit aspect, with grey-yellow millimetric areas, bearing the tendency of confluence on areas of up to 3/2 cm.

Kidneys showed cortical intumescence and paler pyramidal centers, given the grey-purple background of the parenchyma.

The toxicological examination performed by GC-MS for the blood sample taken on admission was negative.

The histopathology of the fragments sampled from the liver revealed the following: at the edge of the histological slide, the presence of collagenous connective structures suggesting the presence of a cystic wall, with a lamellar aspect; the adjacent hepatic tissue displayed a rich, inflammatory infiltrate of a chronic, lymphocytic type; further away from the conjunctive wall hepatic tissue with large areas of necrosis (erasing of the cellular limits, nuclear alterations) and cells presenting various dystrophic aspects have been discovered.

The kidneys showed dystrophies and tubular epithelial necrosis. The stomach fragments revealed hematic infiltrates in the chorionic stomach lining.

Analyzing clinical and autopsy data we concluded that death was nonviolent, due to hepatic-renal failure within an anaphylactic reaction caused by the spontaneous rupture of a hepatic hydatid cyst.

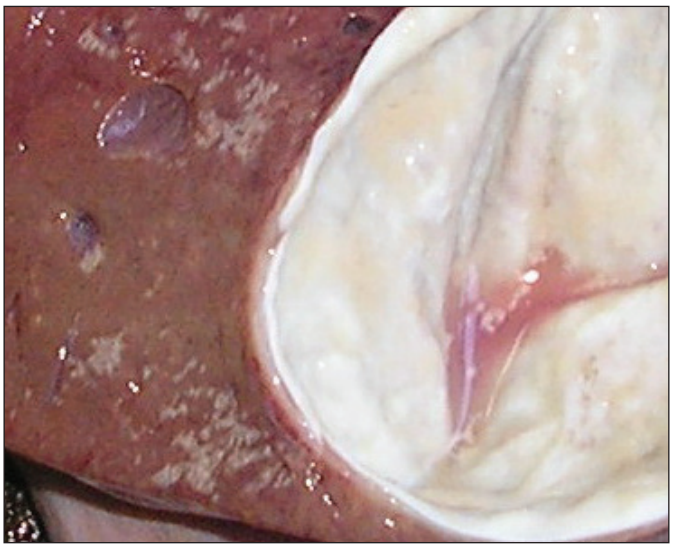

Figure 1. The 7 cm diameter intrahepatic cystic formation; the adjacent hepatic parenchyma with areas of severe dystrophy.

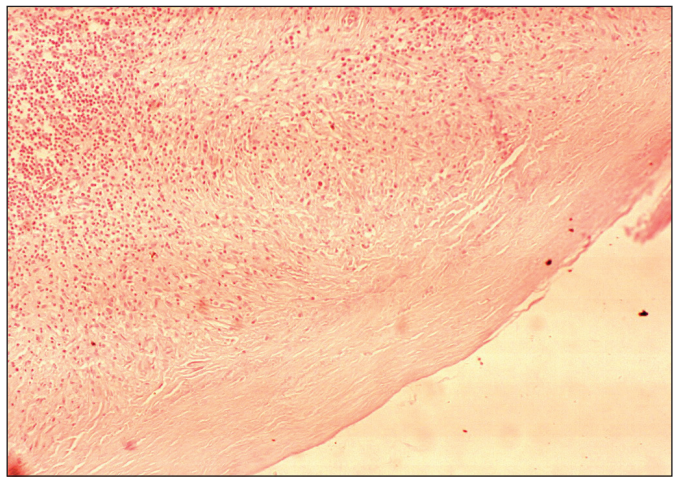

Figure 2. Microscopic appearance of liver tissue: various dystrophic aspects can be distinguished, even necrosis; cystic wall with lamellar structure; chronic inflammatory infiltrate of lymphocytic type (col HE 10x).

DISCUSSIONS

It is estimated that about 2-3 million people around the world suffer from echinococcosis. A review of the specialized literature highlighted that the prevalence of echinococcosis varies between 1-7% in community studies, while the incidence of hospitalized cases ranges between 0-32/100.000. This is more common in women and its frequency increases with age [1]. The disease is endemic in the Middle East, in countries around the Mediterranean and in South America [11]. The incidence is lower in North America, its maximum incidence being in Alaska (1:100.000 inhabitants). The disease is uncommon in Northern Europe, but has a high endemicity in Eastern Europe and in the Mediterranean countries. A recent study in Romania showed the need to implement new preventive measures, due to the disease rate of 5:100,000 inhabitants [12]. In the North-East and Southern Romania, the presence of echinococcosis was studied in a number of 8.569 animals. It was discovered in 49.87% of the sheep and in 32.34% of the cattle analyzed, establishing that the disease in these areas is hyperendemic. The disease has been isolated in 13 animals and a human being by analyzing the Echinococus granulosus sensu stricto molecular biology [13].

Morbidity in echinococcosis is caused by the mass effect of the cyst, or by anaphylactic reactions in case of cyst rupture. In theory, the disease can affect any organ. The most affected organ is the liver (63%), followed by the lungs (25%), muscles (5%), bones (3%), kidney (2%), brain and spleen (1%). Hydatid cyst may occur in different locations: the heart cavities [14], the central nervous system, heart, kidneys, muscles, or mammary glands, ovary, appendix, pancreas, spleen, salivary and adrenal glands, thyroid [15], scrotum [16], gallbladder [17], bones (e.g., the proximal tibial metaphysis and diaphysis) [18] etc. In a group of 64 patients from Samarkand suffering from hydatid cysts it was found that the most frequent location of cysts in the liver were the segments 7 and 8. Hepatic functional status depends on the number and size of hydatid cysts [19].

Urticaria may sometimes be the first clinical manifestation of echinococcosis and it could be a sign of warning prior to severe anaphylactic manifestations.

Clinical manifestations caused by the presence of the cyst and its associated compression effects relate to the affected organ. Complications may arise due to internal cyst superinfection and to the potential rupture of the cyst. A retrospective study of an endemic area of Greece showed that a group of 227 patients have been operated for a total number of 322 liver cysts. The study concluded that over half of these (53.7%) were complicated. The most common complications were: biliary breach - present in 34.4% of the cases (which may sometimes cause acute, even recurrent, pancreatitis), cyst infection - in 32.7% of the cases, while 24.5% of the cases even presented both complications [20].

The independent predictors of liver hydatid cyst breaches within the biliary ways would be: leukocytosis (over 9.000/mm^3), increased direct bilirubin (above 0.7 mg/dL), increased serum levels of alkaline phosphatase (over 120 U/L) and a cyst diameter of over 8.2 cm [21]. The fissures in the cyst lead to an increase in pain and to minor allergic reactions characterized by urticaria and flushing. Massive rupture of the cyst leads to severe anaphylactic reaction, potentially being fatal - as in the analyzed case. The breach of a hydatid cyst, including the hepatic one, can cause different allergic symptoms up to anaphylactic shock, when its content flows into the peritoneum or reaches the bloodstream. In such a patient presenting hydatid cysts, admitted to intensive care, the disease was complicated by an ischemic stroke [22].

Sometimes vesicles leaving a hepatic hydatid cyst can migrate through the veins and can cause asymptomatic or symptomatic pulmonary arterial embolism (even leading to sudden death, in massive forms) [23].

The breaches of hydatid cysts mostly appear after trauma or during surgical intervention. But this breach can also appear spontaneously, as in the presented case. In a similar recently published case [24] a breach of hydatid cysts in the spleen also led to a reaction of anaphylactic shock. The literature also reports of hydatid cyst fistulization within the duodenum [25], in the colon[26] or percutaneous [27]. Anaphylactic reactions may occur following the surgical percutaneous aspiration of the hepatic hydatid cyst, usually after a few hours; these patients should be carefully monitored after surgery [28]. A case of anaphylactic shock in the operating room was also published. The shock occurred even before the intervention started and showed a transitory oscillation of the EKG ST segment, without the breach of the hepatic hydatid cyst. Thus the cyst could surgically be removed after the patient was stable [29].

Sometimes hepatic hydatid cysts may have impressive dimensions. Moreover, it can even extend within the thoracic cavity. One such case was recently published: the cyst invaded the diaphragm and extended to the posterior mediastinum, compressing the pericardium and esophagus. Moreover, it produced a secondary Budd-Chiari syndrome by occluding the suprahepatic left vein near its outfall in the vena cava and near the inferior vena cava [30].

In an autopsy study conducted in Northern Tunisia, over a period of 6 years, 26 hydatidosis deaths were analyzed. The deaths most commonly occurred at home, sometimes after exercise and, rarely, after trauma, sepsis or due to acute respiratory failure; anaphylaxis was the cause of 17 deaths [31]. Most cases of sudden death associated with hydatid cysts involve a cardiac cyst [3, 5, 6, 8, 9], as it can cause anaphylaxis, embolism, obstruction of the heart chambers, pericarditis, or cardiac tamponade [8].

Hepatic cysts causing sudden death are rare, but have been cited as well, usually through anaphylactic processes [10].

CONCLUSION

Even if rare, sudden deaths caused by complications of Echinococcus infections due occur, and they myst be suspected alwaya when finding a cystic mass with clear (river mountain fluid). In these cases, the autopsy must be conducted with extreme caution, as the personnel involved may become infected.

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