Systemic embolism due to liver hydatic cyst: Case report

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Abstract: Nonthrombotic pulmonary embolisms are rare unlike pulmonary thromboembolisms. Hydatid cyst related complications such as anaphylactic shock, infection, and embolism might cause sudden deaths. Hydatid pulmonary embolism is a rare entity usually caused by rupture of a cardiac or a hepatic hydatid cyst. On the other hand, venous and arterial embolisms due to cyst hydatid are extremely rare. Here, we report a case of systemic hydatid embolism secondary to rupture of a hydatid cyst.

A 17-year-old male suddenly died following an immediate deterioration of health status. At autopsy, liver dissection revealed a ruptured haemorrhagic hydatid cyst. Vascular vesicle embolisms were observed in lungs, liver, hearth and kidneys.

Because of atypical clinical symptoms it is difficult to establish the diagnosis of pulmonary embolism of hydatid cyst. Histopathological examination plays a key role in establishing a definitive diagnosis for this cause of sudden death.

Key Words: non-thrombotic embolism, hydatid cyst, sudden death.

Hydatid cyst, a common disease that is endemic in rural areas, is mostly encountered in developing countries. Humans are intermediate hosts and accidentally infected by ingestion of food or water contaminated with Echinococcus species or by direct contact with infected animals [1]. Although living in a rural or farming area remains the most common risk factor, being connected to such areas by any means such as travelling, migration and trading may cause changes in the epidemiology of this uncommon disease [2]. Therefore, this disease might cause morbidity and mortality all over the world. In the affected individuals cysts might remain asymptomatic until identified during the autopsy as an incidental finding. However, it might cause sudden and unexpected death in some cases [1]. These sudden deaths are mostly secondary to hydatid cyst related complications such as anaphylactic shock, infection, and embolisms. Hydatid pulmonary embolism is a rare entity usually caused by rupture of a cardiac or a hepatic hydatid cyst. However, deaths caused by venous and arterial embolisms due to cyst hydatid are extremely rare. In this report, we present a case of sudden death due to systemic hydatid embolism secondary to rupture of a hydatid cyst.

CASE

A 17 year-old male with no previous history of trauma, symptom or disease collapsed, and suddenly died shortly after admission to hospital, before diagnosis could be reached. Therefore, a complete postmortem investigation was performed.

External examination did not reveal any findings specific to a disease or trauma. At internal examination; diffuse sub-pleural petechial haemorrhages were observed on the surface of the lungs. There was no fluid
or haemorrhagic material in the abdominal cavity. Liver was 2074 g in weight; the diaphragmatic face of liver was not solid and resembled two separate shrunken cyst of which one at upper subcapsular medial side of right liver lobe and the other at the middle of right and left liver lobes. Liver sections showed two neighbouring ruptured empty and shrunken giant hydatid cysts localized around vena cava inferior and its main hepatic branches. Giant cysts' dimensions were 22x18x14 cm and 8x10x6 cm, respectively (Fig. 1). At this localization diaphragm adhered to neighbouring liver part due to thickened fibrous adhesions. Macroscopic examination of remaining organs was completely unremarkable.

Histopathological examination of the liver at cyst site showed outer wall, laminated cuticula, and inner germinal layer, which was compatible with hydatid disease. Furthermore, there were scolices in the liver vessels (Fig. 2a-2b).

Echinococcus scolices were identified in the vessels of both lungs at microscopic examination, suggestive for a diagnosis of nonthrombotic hydatid embolism (Fig. 3). Scolices were also identified in cardiac vessels at myocardial sections (Fig. 4).

On the other hand, hydatid embolisms were identified in a renal venule (Fig. 4) and a coroner venule as a surprising finding (Fig. 5). There were no anaphylaxis related significant findings in external and internal examination and at histopathological examination. Detailed toxicological analyses were performed; no traces of any toxic agents were detected. Death was attributed to systemic hydatid embolism.

Figure 1. Giant hydatid cyst in the liver.

Figure 2 a, b. Hydatid cyst (a) (H&E, X40) and hydatid embolism (b) in liver (H&E, X200)
Hydatid cyst disease is a common entity in countries rich in farming facilities [3]. Humans are intermediate hosts and accidentally infected by ingestion of contaminated food or water or by contact with infected animals. Infected individuals are mostly asymptomatic [1]. However, the disease might cause a number of medical conditions including sudden death. Hydatid cyst is uncommon in young individuals [4]. Ben Khelil et al. reported that hydatid cyst is a potential cause of sudden unexpected death particularly among children and young adults [5]. Jediti et al. attributed this to an easier rupture of hydatid cysts in young individuals as a consequence of frequent traumas [6]. The presented case was a 17 year-old male who suddenly collapsed, and died soon after admission to the hospital. There was no trauma history, and external examination did not reveal findings related to trauma at the autopsy. Therefore, the rupture of the incidental giant hydatid cyst was thought to be spontaneous.

The reported mechanisms of sudden death due to hydatid disease include anaphylaxis, sepsis, multiorgan failure, fatal embolization of hydatid material. Furthermore, sudden death might be secondary to arrhythmias, cardiac tamponade, pericarditis, outflow obstruction, and coronary artery compression as complications of cardiac hydatid cysts [1, 7, 8]. Hydatid embolization to the lungs caused by hydatid rupture from the right side of the heart, while hydatid embolization into systemic circulation causes by hydatid rupture in the left. [9].

Systemic embolization is often found in cardiac hydatid disease. Unlike other cases cited in literature, no ruptured or intact hydatid cyst was detected in the hearth in our case. Pulmonary or systemic embolism caused by ruptured hydatid cysts into the venous or arterial system is an extremely rare complication of liver hydatid cyst, which may result in chronic pulmonary hypertension, subacute pulmonary hypertension, and sudden death [10]. In the presented case, histopathologic examination revealed hydatid embolism in pulmonary, cardiac, liver, and renal vessels. Although systemic embolism is mostly associated with cardiac hydatid disease, there were only liver hydatid cysts in the presented case. The exact mechanism of systemic hydatid embolism could not be fully explained in our case.

However, high intracystic pressure might be considered as a possible explanation for the systemic embolism in our case. Possibly higher intracystic pressure compared to portal and caval systems may result in reversed blood flow and systemic hydatid embolism. Previously conducted studies reported that intracystic pressure could rise up to 44-46 mmHg in intact cysts [11, 12]. On the other hand portal pressure is about 7 mmHg, while caval system pressure changes between 0 to 4 mmHg [13-15]. The rupture was possibly caused by a cough or Valsalva manoeuvre causing an acute increase of the abdominal pressure. High intracystic pressure and excessive amount of liquid, which force against circulation with lower pressure in portal and caval systems might cause reverse blood circulation and systemic hydatid embolism towards renal, cardiac and lung vessels.

DISCUSSION

Hydatid cyst disease is a common entity in countries rich on farming facilities [3]. Humans are intermediate hosts and accidentally infected by ingestion of contaminated food or water or by contact with infected animals. Infected individuals are mostly asymptomatic [1].
References