Anaphylactic shock as a single presentation of Echinococcus cyst

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Abstract

We describe the case of a 14-year-old boy of Turkish origin, presenting with anaphylactic shock after a minor abdominal trauma. Further investigations revealed a hepatic Echinococcal cyst without evidence of rupture. Anti-helminthic therapy was administered. Because of aggravating symptoms and recurrent anaphylaxis, surgical excision was performed. Intra-operative, a rupture into the biliary tree was seen. After surgery, the anaphylactic symptoms disappeared and the patient recovered. This case-report supports the fact that anaphylactic shock can be the only presentation of a hydatid cyst. Microscopic spillage can possibly be sufficient to cause major anaphylaxis. (Acta gastroenterol. belg., **2011**, 74, **462-464**).

Introduction

Human hydatid disease occurs by infestation of *Echinococcus granulosus* (cystic hydatid disease) and less frequently by the more malignant form *Echinococcus multilocularis* (alveolar hydatid disease). In the normal life cycle of Echinococcus species, the adult tapeworms inhabit the small intestine of definitive hosts such as dogs, wolves and other canines. The different echinococcal cyst stages are found in the intermediate hosts. Most often the tapeworm eggs are passed by the faeces of infected dogs and ingested by sheep. The larve stage penetrates the intestinal wall, and is then disseminated by blood to major organs. The problem arises when humans act as an accidental intermediate host and ingest viable eggs which have been shed in the faeces of the definitive host. As the liver acts as the first filter for hydatid larvae, it becomes the most commonly affected organ followed by the lung (1).

Case report

A 14-year-old boy of Turkish origin presented with syncope after a minor abdominal trauma during ice skating. After admission to the hospital, he developed an anaphylactic shock with hypotension, Quincke oedema and rash in the emergency department. There was no medication used or food ingestion reported that could have explained a possible allergic reaction. After stabilization, a computed tomography of the brain was obtained because of sustained lethargy. No abnormalities were detected. Toxicology was negative. Laboratory results showed an elevated total IgE (353 kU/L), leukocytosis (24,17 103/μL) with eosinophilia (1021/μL), an elevated total bilirubin (7,0 mg/dL : both conjugated and unconjugated), a slightly elevated AST (60 U/L), and a high LDH (1301 U/L). Inflammatory parameters were also raised such as C-reactive protein (6 mg/dL) and erythrocyte sedimentation rate (31 mm/h first hour). Nevertheless, a specific RAST for Latex was less than 0,35 kU/L. During the hospital stay both total IgE (798 kU/L), C-reactive protein (9,9 mg/dl), eosinophilia (1842/μL) and gamma-GT (136U/L) continued to rise. Blood cultures remained negative.

Subsequently, he developed fever, a skin rash and an icteric sclerae. Because of suspicion of cholangitis, an ultrasound of the abdomen was requested which revealed a liver cyst with septae, without intra-abdominal free fluid. A computed tomography was performed to determine the exact localisation of the cyst. The images were pathognomonic for an Ecchinococcus cyst and showed the detachment of the germinal layer from the cyst wall, the typical "water-lily sign" (Fig. 1, Fig. 2). Specific enzyme-linked immunosorbent assay (ELISA) confirmed the formation of antibodies against *Echinococcus granulosus*. Albendazole (15 mg/kg per day) and Praziquantel (35-40 mg/kg), both anti-helmintic drugs, were administered.

Because of aggravating symptoms such as dissemination of the skin rash, swollen joints, fainting, tachypnoe with desaturation and intermittent hypotension, he was transferred to the intensive care department. Radiologic imaging of the chest revealed diffuse reticulo-nodular opacities consistent with an acute respiratory distress syndrome. Because of the systemic inflammatory syndrome, the decision was made to remove the cyst surgically.

During surgery there was no evidence of rupture of the cyst into the peritoneal space. However, the cyst had a shrunken appearance. When the cyst was punctured, biliary stained fluid was aspirated (Fig. 3). After closed aspiration and instillation and reaspiration of hypertonic saline (NaCl 3%), the cyst was opened. An undulating

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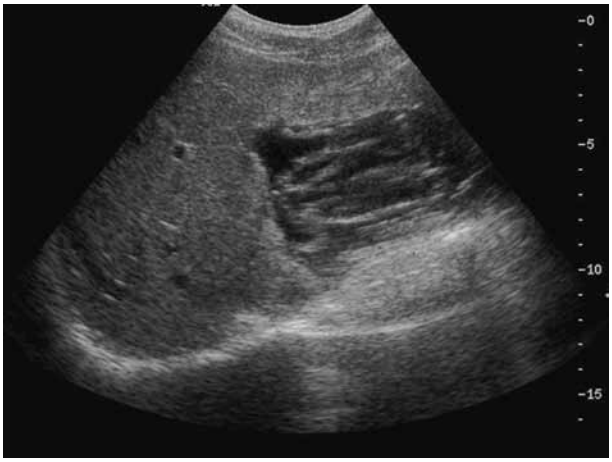

Fig. 1. — Echography of liver : Liver cyst with detachment of germinal layer from cyst wall "water lily sign", pathognomonic for E. granulosis cyst or Echinococcosis.

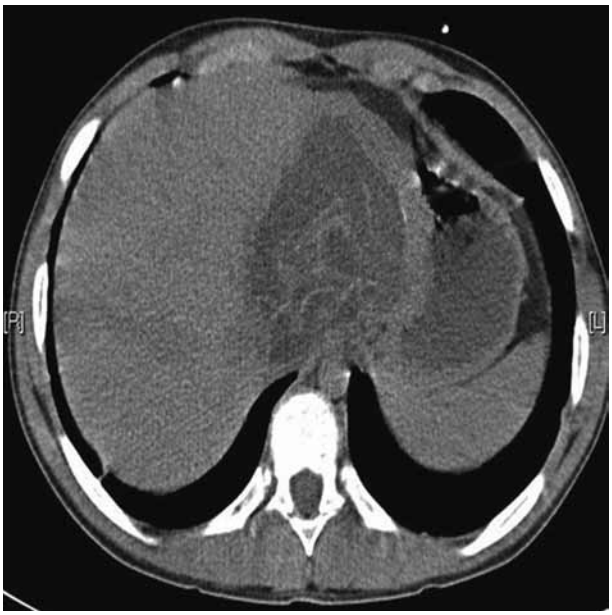

Fig. 2. — Computed tomography of liver : Solitary hydatid cyst with calcification (arrow) 8-9-7,5 cm in liver segment I-II.

endocyst was removed (Fig. 4). A part of the pericyst, which was impregnated with bile, was resected. Cholangiography via the gallbladder demonstrated two biliary tract erosions. These were over sewn. An omental flap was mobilised to cover the former cyst cavity.

Microscopic investigation of the cyst revealed an Echinococcus species. To reduce the risk of secondary hydatidosis, the anti-helminthic therapy was continued for six month after surgery. After the surgery, the anaphylactic symptoms disappeared and the patient recovered.

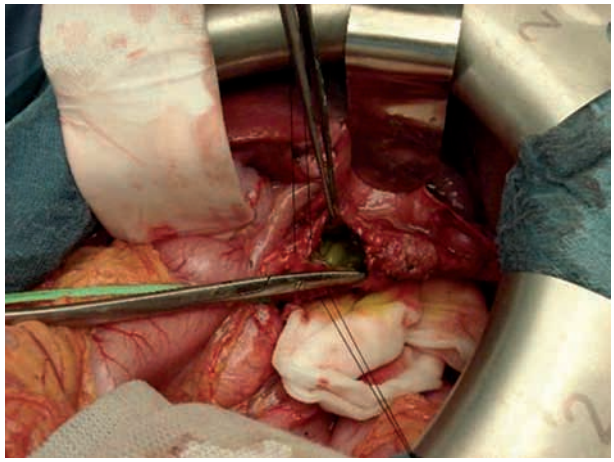

Fig. 3. — Left liver segment with cyst containing bile, suggestive for rupture into biliary tree.

Fig. 4. — Removed endocyst from hydatid cyst after aspiration, instillation of NaCl 3% and reaspiration.

Microscopic investigation of the cyst reveals an Echinococcus species. Specific enzyme-linked immunosorbent assay (ELISA) is positive for Echinococcus.

Discussion

Hydatid cysts remain often asymptomatic during many years. Symptoms are mainly due to the mass effect of the enlarging cyst in a confined space. Other serious complications are rupture and secondary bacterial infection. The causes of rapid clinical decline involve a wide range of mechanisms including anaphylaxis, with or without cyst rupture (2).

Diagnosis is made by imaging and serologic testing, but false negative serologic testing can occur (3). Several treatment options exist but evidence on their efficacy lacks. Most authors agree on the subsequent therapy : albendazole, sometimes combined with Percutaneus Aspiration and Instillation of a protoscolicidal agent and Reaspiration (PAIR procedure), or surgery. To reduce the risk of secondary hydatidosis, therapy should be continued at least one month after surgery (4).

In this case, the patient lived in a rural area in Turkey till the age of 6. He still visits his homeland frequently during the holidays. During the weeks before presentation, he experienced a decreased appetite. The presentation with anaphylaxis occurred after a mild abdominal trauma. Release of antigens after cyst spillage or rupture caused a secondary immunologic response.

In the literature, anaphylaxis is described when evident rupture of a pulmonary or hepatic hydatid cyst occurs, e.g. after a blunt trauma (5,6,7). Seldom, anaphylactic shock was reported in patients without macroscopic rupture of the hydatid cyst (8,9). In this case there was leakage into the biliary tree. Possible spillage of cyst fluid with intra-vascular spread caused the serious anaphylactic symptoms. One case-report described the sudden death of a patient due to unexpected anaphylactic shock. Autopsy revealed laryngeal oedema with inflammatory infiltration composed of mast cells in the larynx. A non-ruptured hydatid liver cyst was detected with the presence of scolices in the pulmonary artery (10).

In conclusion, we stress the importance of maintaining a high level of clinical awareness for Echinococcosis, especially in patients from endemic regions presenting with anaphylaxis without an obvious cause. This is of great importance because missing the diagnosis can have serious complications and can lead to sudden death.

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