

Recurrent Anaphylaxis due to Non-Ruptured Hepatic Hydatid Cysts

Asli Gelincik^a Ferhan Özşeker^a Suna Büyüköztürk^a Bahattin Çolakoğlu^a
Murat Dal^a Aydin Alper^b

^aDivision of Allergy, Department of Internal Medicine, and ^bDepartment of Hepatobiliary Surgery, Faculty of Medicine, Istanbul University, Istanbul, Turkey

Key Words

Anaphylaxis · Hydatid disease · Liver

Abstract

Background: Hydatid disease, a parasitic infestation of humans, is endemic in the Mediterranean region, Australia, New Zealand and the Middle East, and mostly involves the liver. Anaphylactic reactions, which sometimes are the first manifestations of the disease, frequently occur due to cyst rupture after a minor/major trauma, though they may also be spontaneously seen on rare occasions. In extremely few studies, anaphylactic shock has been reported in patients without macroscopic rupture of the hydatid cysts. **Case Report:** Our patient had recurrent anaphylactic episodes without any trauma and had been misdiagnosed for several years even though the patient was living in a region endemic for hydatid disease. **Conclusion:** We emphasize that physicians should be highly aware of hydatid disease as a possible etiology for seemingly idiopathic anaphylactic reactions, especially in endemic regions.

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Introduction

Hydatid disease (cystic echinococcosis) is a parasitic infestation of humans caused by the larval stage of *Echinococcus granulosus* [1]. It is highly endemic in Australia,

New Zealand, the Middle East and the Mediterranean region including Turkey and Greece [2, 3].

The infestation mostly involves the liver and is clinically not apparent in most patients. Anaphylactic reactions, which sometimes are the first manifestations of the disease, frequently occur due to rupture of the cysts after a minor or a major trauma, though they may also rarely be seen spontaneously [4, 5]. These reactions result from hypersensitivity to allergens found in the echinococcal cysts. However, in extremely few reports anaphylactic shock has also been reported in patients in the absence of macroscopic ruptures of the hydatid cysts [6–8].

Our patient had recurrent anaphylactic episodes without any trauma and had been misdiagnosed for several years even though the patient was living in a region endemic for hydatid disease.

Case Report

In October 2005, a 48-year-old woman was admitted to our Allergy Outpatient Clinic 1 month after an episode of emesis, abdominal discomfort, facial flushing, generalized pruritus, palpitations, dizziness and fainting following the ingestion of an ordinary meal. During this episode, she received emergency treatment in a public hospital with the diagnosis of anaphylactic shock and was referred to our clinic for further evaluation. A detailed medical history revealed that the patient had had her first anaphylactic episode in 1996 which had resolved spontaneously. The second episode, occurring 2 years later, was much more severe and required resuscitation; the etiology of this anaphylactic attack was

not determined. She was started on daily antihistamine in order to prevent a subsequent attack. The third episode in 2000 occurred during an abdominal ultrasonography for abdominal discomfort of unknown etiology. Again she went into cardiopulmonary arrest but was successfully resuscitated. Ultrasound and subsequent computed tomography revealed two hydatid cysts, a large (6 × 5 cm) cyst in the left lobe of the liver and a smaller one in the right lobe of the liver. Surgery was performed in 2000 for the hydatid cysts; cystotomy, unroofing, drainage and partial cystectomy were done for the large cyst in the left lobe and unroofing only for the smaller cyst. The patient was started on albendazole for 6 months, and she was not called for follow-up controls. During the 5 years following the operation, she complained of mild symptoms of facial flushing but did not seek any further medical attention.

On admission to our clinic in October 2005, her clinical examination and routine blood tests including blood eosinophil counts, hepatic transaminase values, alkaline phosphatases and γ -glutamyltransferases were normal. No kind of parasitic infestation was found in her fecal examination. Her thyroid function tests and electrocardiography were normal. Skin prick tests with common aeroallergens and food allergens evaluating her atopic status were normal. Total serum IgE was 16,800 IU/ml and the *E. granulosus* indirect hemagglutination assay was 1/1,000.

In T₂-weighted axial MRI sequences, a 3 × 3.5 cm cyst with hypointense capsule in the posterior segment of the right lobe and a 4 × 6 cm cyst with septations in the lateral segment of the left lobe, consistent with the diagnosis of hepatic hydatid cysts, were observed (fig. 1). The thoracic computed tomography was normal.

Relapsing hydatid disease was diagnosed, and the patient was operated on in the Hepatobiliary Surgery Department of our clinic on October 19, 2005. The most definitive solution for the cyst was planned to eradicate all the live elements of the cysts. The cyst in the left lobe was treated with cystotomy, drainage and partial cystectomy, whereas cyst aspiration and irrigation with salty water and alcohol under ultrasonographic guidance was performed for the cyst in the posterior segment of the right lobe. The patient was admitted to the intensive care unit for the following 15 days and recovered without any complications. The diagnosis of hydatid cyst was confirmed by the pathologic examination of the resected material. Postoperative prophylactic treatment with two courses of albendazole (each lasting 15 days with a drug-free interval of 2 weeks) was started.

Neither recurrence of the hydatid disease nor anaphylactic reactions were observed during the 6-month follow-up of the patient, and the total IgE level in blood gradually declined to 4,350 IU/ml.

Discussion

Hydatid disease is mostly recognized in the liver, followed by the lungs, spleen, kidneys, and occasionally some unusual organs [9]. Usually, hepatic hydatid cysts are clinically unapparent and diagnosed incidentally when abdominal ultrasonography is applied for other reasons [10]. Symptomatology, when present, is usually

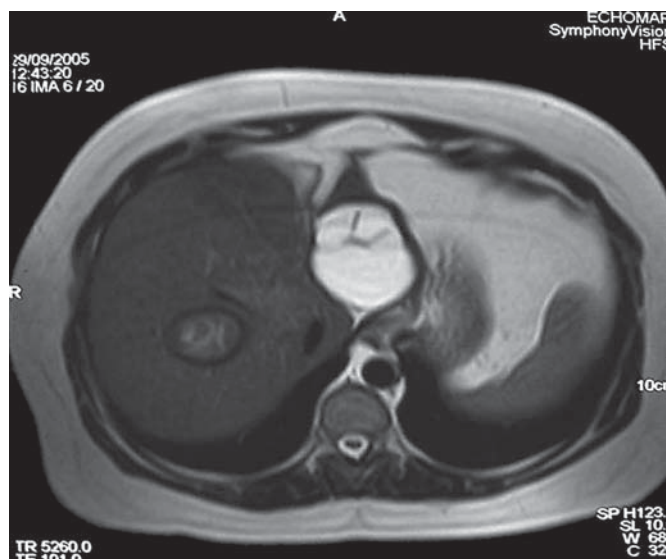


Fig. 1. T₂-weighted axial MRI of the patient.

due to the rupture or infection of the echinococcal cyst, or dysfunction of the affected organ [6].

Hydatid cysts can rupture as a result of trauma or sometimes spontaneously [9], and anaphylaxis can be a complication. However, several case reports indicate that anaphylaxis can also occur without macroscopic hydatid cyst rupture. At the autopsies of 2 patients who had died suddenly from severe anaphylaxis, it was concluded that non-ruptured cysts had caused anaphylaxis due to the spread of the cyst content into the systemic circulation [6, 11]. In another case report, a patient with recurrent anaphylaxis was operated on with the diagnosis of hydatid cyst, and small, incomplete ruptures of liver cysts were observed [8], which were suspected to be the cause of the anaphylactic attacks.

Our patient first had a mild anaphylactic attack in 1996 and again in 1998 and had to be resuscitated for severe anaphylactic shock. Although Turkey is in the endemic region for hydatid disease, this was not taken into consideration and the reason for the attacks remained uncertain until another severe attack occurred in 2000. After the hydatid cysts had been operated on and a 6-month therapy with albendazole had been started, the patient was not given appropriate information regarding disease follow-up. The recurrence of spontaneous anaphylactic reactions 5 years after hydatid cyst operation did not lead the primary care physicians to investigate hydatid cyst relapse as a most probable diagnosis. Although no cyst ruptures were observed at the operation,

we concluded that anaphylactic episodes of the patient were due to spontaneous spills of the cyst contents into the systemic circulation. This may be explained by the failure of the technique used in the initial operation and the prophylactic treatment given in the postoperative period. According to the report of this operation, no data concerning the usage of protoscolicidal agents during the operation was mentioned. This may be another reason for the relapse of the cysts. Therefore, surgeons operating on hydatid cysts should favor the most definitive surgery and remove all parasitic material using the new surgical techniques which make such procedures easier. Complementary treatment of albendazole and in some cases praziquantel with albendazole must be considered. A careful follow-up of all cases operated on for cystic echinococcosis should be ensured for at least 2 years. During this period, the success of the treatment can be judged by monitoring serum-specific IgE, total IgE and IgG serol-

ogy. High levels of these immunoglobulins after surgery must remind us that the patients may develop allergic reactions in the postoperative period, whereas low levels may prove the success of the treatment.

Taking into consideration such unusual presentations as allergic/anaphylactic reactions in otherwise asymptomatic patients of hydatid cysts, we emphasize that physicians should be highly aware of hydatid disease as a possible etiology for seemingly idiopathic anaphylactic reactions, especially in endemic regions. Therefore, for patients presenting with recurrent anaphylaxis episodes in these regions, liver ultrasound examination and further echinococcosis serology must be done. Whenever hydatid disease is suspected, radiologists should be cautioned not to prolong radiologic and ultrasonographic procedures which may lead to a minor trauma and thereby trigger an episode of anaphylactic reactions as in our case.

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