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Case report

Multiorgan *Echinococcus* infection: Treatment of an immigrant in the United States

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ABSTRACT

We discuss the clinical presentation and management of a 36-year-old woman presenting with a 12-year history of recurrent hydatid disease, a disease caused by *Echinococcus granulosus* and other *Echinococcal* species, which is rarely seen in the contiguous United States. She has had multiple procedures since her diagnosis in 2005 and is still closely being followed for possible disease recurrence.

Introduction

Echinococcus is a parasite non-endemic to the United States. It is a difficult infection to manage and treat due to its potential for extensive involvement and high rate of recurrence. This is further complicated by physicians' lack of familiarity with hydatid disease, making it difficult for patients to receive proper and quality management of their disease.

Case report

The patient was born in Iraq with a history of traveling to Syria, Saudi Arabia, Turkey, and Mali. The exact origin of her infection has not been established. She recalls commonly coming in contact and playing with dogs as a child in Iraq and recounts her complete immersion into the lifestyle of the natives of Mali, including consuming their food and drink, which may have been a risk factor for her condition.

The diagnosis was made in 2005 after arriving to the emergency department at Oakwood Hospital in Dearborn, MI due to gradually progressive dyspnea and pleuritic chest pain. On arrival, her oxygen saturation was alarmingly low. Chest radiographs revealed a large right pleural effusion with atelectasis. Hydatid cysts were found in her liver and spleen on CT scan. CT scan demonstrated no hydatid cysts in the chest, so CT-guided thoracentesis was indicated at this time. Thoracocentesis revealed eosinophilic exudative fluid with no evidence of hydatid disease in the chest. The pleural effusion was determined to be sympathetic to diaphragmatic irritation from liver involvement. It was determined that the patient's infection was due to *E. granulosus* after she was found to have an elevated *Echinococcus* titer. The patient began a six-month course of 400 mg albendazole twice daily, which she took regularly. She declined surgery at that point.

In 2006, the patient did not have significant improvement and was recommended surgery again, but she declined. Within a year, she presented to the ED complaining of severe abdominal pain due to hydatid cyst rupture, confirmed by CT. CT also revealed extensive hydatid disease in the omentum, spleen, liver, and retroperitoneum. Due to the severity of the hydatid disease (Fig. 1A), she agreed to surgery at this time. A surgeon from Lebanon with experience with hydatid disease performed the surgery. Over 100 cysts were discovered within the abdomen. Omentectomy was performed, splenic cysts were aspirated and injected with 5% saline, and multiple cysts in the liver were drained and partially excised. There were also retroperitoneal hydatid cysts on the right that were excised. She was placed on the same regimen of 400 mg albendazole twice daily for 60 days postoperatively.

On CT, it was found that her hydatid cysts had recurred in the spleen (Fig. 1B) and pelvis seven years later during a follow-up appointment. The surgeon once again attempted to preserve the spleen by aspirating the cysts and injecting hypertonic saline again. At this time, pelvic cyst removal and splenic cyst drainage was performed. Within the year, the patient's hydatid cysts recurred in the pelvis, spleen, and diaphragm. She was recommended to consult a gynecologist oncologist due to pelvic involvement of cysts who recommended a total hysterectomy and bilateral salpingoophorectomy (TAH-BSO) (he informed her that he was unfamiliar with the disease). She returned to her previous surgeon and was told that a TAH-BSO may be unnecessary. At this time, the patient underwent a laparotomy with lysis of adhesions with splenectomy and drainage of multiple perisplenic, pelvic, and diaphragmatic cysts in addition to a partial gastrectomy due to extensive involvement. The patient was given appropriate vaccinations and prophylactic penicillin.

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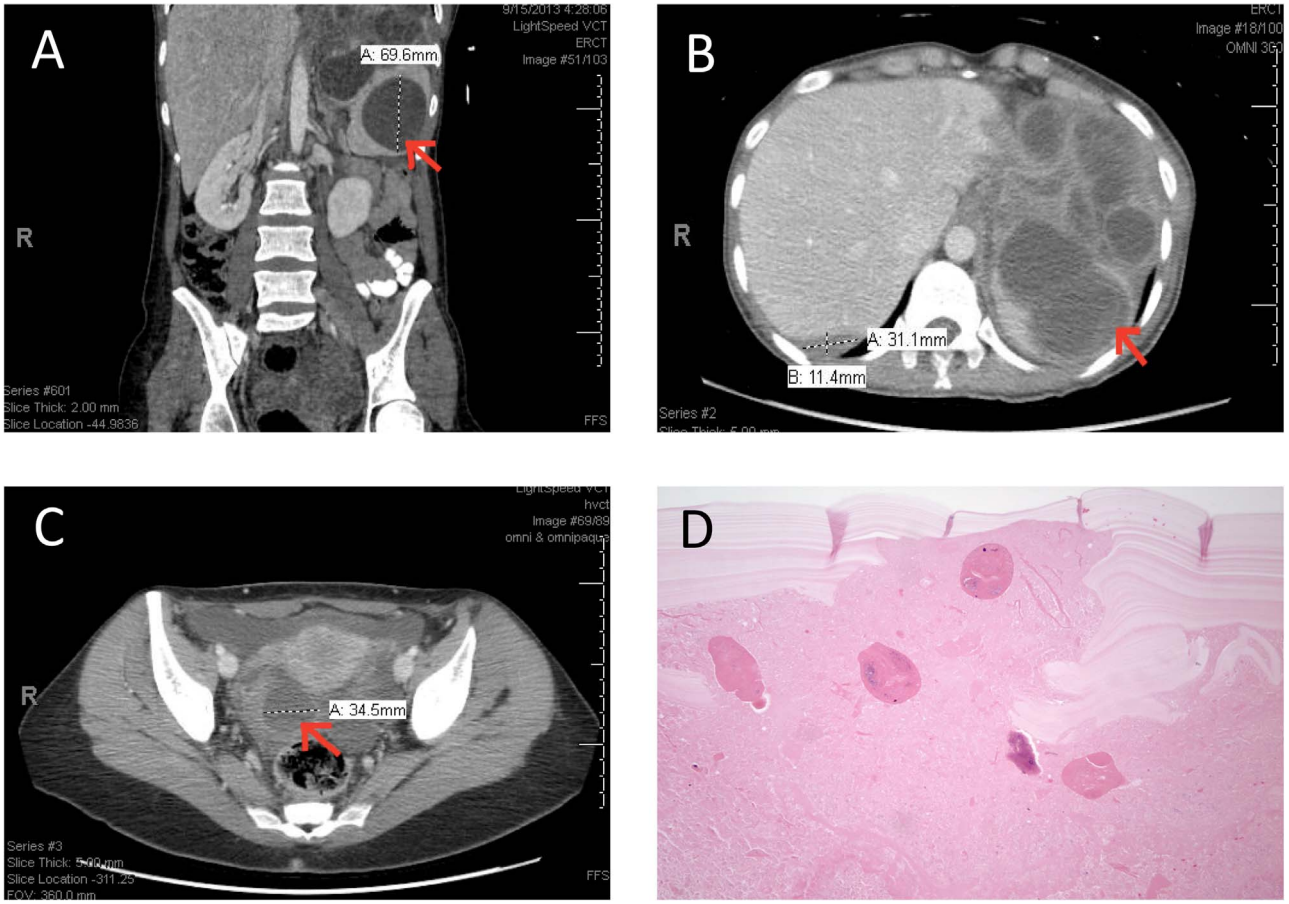

Fig. 1. Radiographic and pathologic findings of *Echinococcus* granulosus.
Panel A – Coronal CT demonstrating splenic hydatid cysts.
Panel B – Transverse CT demonstrating splenic hydatid cysts.
Panel C – Transverse CT demonstrating right ovarian hydatid cyst.
Panel D – H & E stain of pelvic cyst biopsy at 100× demonstrating the laminated membranes and scolices.

She developed cholecystitis in 2014 in addition to recurrence of the pelvic, gastric, hepatic, and diaphragmatic hydatid cysts found on CT after she presented with elevated antibody titers for echinococcosis on follow-up. She underwent an exploratory laparotomy, excision of pelvic and gastrohepatic cysts, drainage and excision of hepatic and diaphragmatic cysts, and appendectomy. In 2015, she was found to have rising antibody titers and another recurrence of hydatid disease limited to the pelvis with a 12 cm adnexal cyst (Fig. 1C). Interventional Radiology (IR) was consulted and they evaluated the patient but ultimately declined to intervene due to unfamiliarity with the disease and concern about spillage of hydatid cysts contents into the peritoneal cavity. In addition, she consulted many gynecologists to perform surgery on her but they declined as well due to unfamiliarity with the disease.

In 2016, her previous surgeon and a gynecological oncologist decided to perform a combined surgery for exploratory laparotomy with possible TAH-BSO for the recurrence discovered in 2015. The patient was given a perioperative regimen of albendazole. At laparotomy, there was no recurrence of the cysts in the abdomen. She was found to have multiple large simple pelvic cysts that were not related to hydatid disease. A small hydatid cyst on the right ovary was drained and removed. A TAH-BSO was deemed unnecessary. Pathology noted that these cysts consisted of clots with surrounding fibrosis and lymphocytosis, suggesting that these cysts were the result of a focal inflammatory reaction (related to prior hydatid disease) as opposed to being hydatid cysts. This pathological finding was in contrast to her prior confirmed hydatid cysts that were described as having broad cystic palisading granulomas with an eosinophilic avascular refractive laminated membrane, occasionally surrounded by multinucleated giant cells (Fig. 1D). In addition, brood capsules and protoscolices were appreciated in the right ovary. The patient has recovered well and continues to be followed disease free as of February 2017.

Discussion

Echinococcus granulosus and its related species are a genus of zoonotic cestodes that cause cystic and alveolar echinococcosis. Their definitive hosts are carnivores, most commonly dogs. Definitive hosts are those species that can facilitate the growth of these parasites up to their mature form. Their intermediate hosts are typically domestic animals such a pig, goat, and sheep; humans are incidental hosts (Fig. 2). Intermediates hosts differ from definitive hosts in that the parasite cannot fulfill its entire life cycle and can only create cysts and eggs in these species. Typical infection occurs through ingestion of meats, fruits, or vegetables contaminated by feces of infected foxes or other canines [1]. Once eggs hatch and release oncospheres in the gastrointestinal tract, these *Echinococcus* oncospheres can penetrate the mucosa and spread through the circulatory system to organs such as the liver (60%), lungs (20%), spleen (3%), kidneys (4%), central nervous system (3%), and bone (2%) [2,3].

Upon reaching these organs, oncospheres develop into cysts containing protoscolices. This is *Echinococcus*'s larval stage and may develop into adult worms in the intestines of definitive hosts. Cysts are slow growing, so the disease is typically diagnosed in adults presenting with symptoms from mass effect. Cysts possess an outer cuticular layer

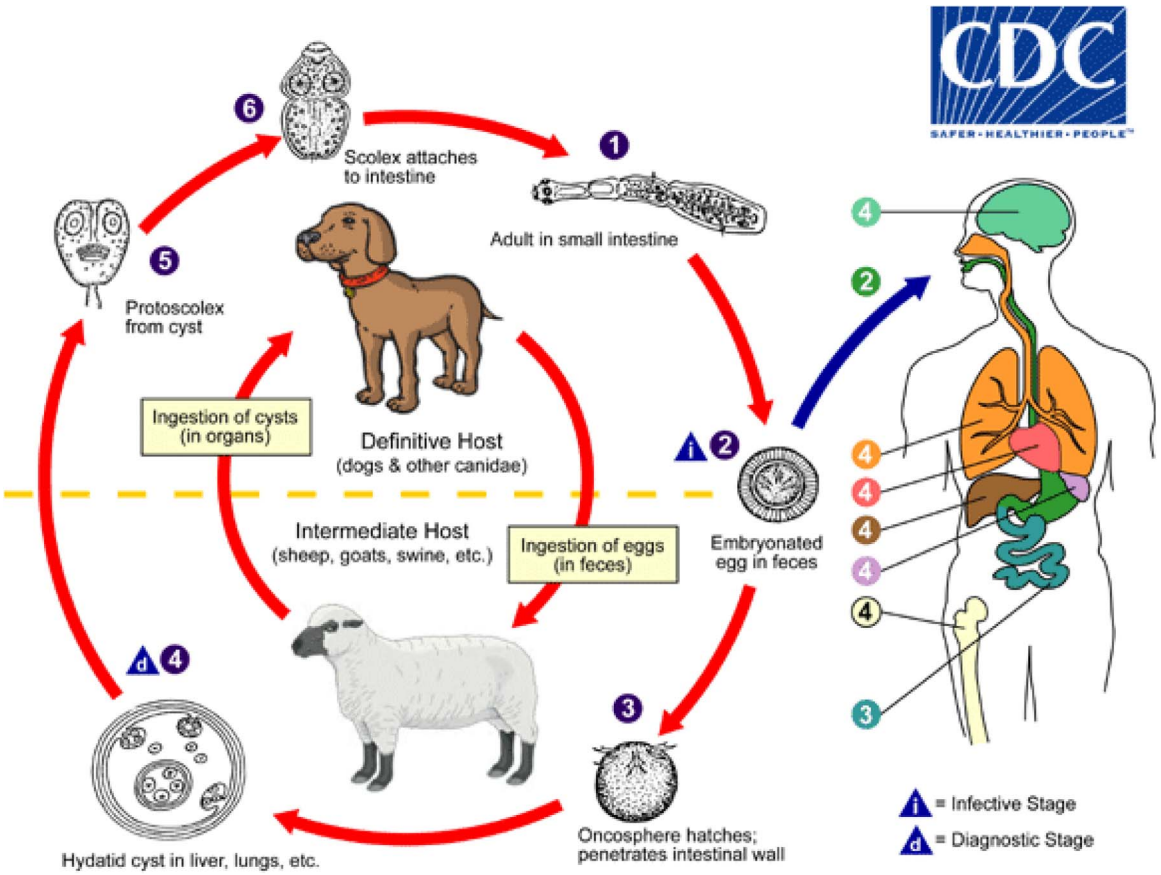

Fig. 2. Diagram of life cycle of *Echinococcus* granulosus.

(pericyst) and an inner germinal layer (endocyst). The cuticular layer is entirely composed of host cells. The cuticular layer is also described as a laminated, hyaline membrane, whereas the germinal layer secretes hydatid fluid into the cysts [4]. Within these hydatid cysts, daughter cysts can form that also contain protoscolices. Cyst rupture can lead to diffuse "metastatic" involvement of hydatid cysts in the peritoneal cavity as well as fatal anaphylactic shock. Thus the need for cyst injection with hypertonic saline or hydrogen peroxide before cyst removal is required to kill the protoscolices and prevent this outcome. Peritoneal seeding can form grape-like clusters along structures such as the greater omentum [5]. This disease is not contagious between humans.

Endemically contracted cases of echinococcosis are relatively rare in the United States. There have been reported cases of echinococcosis in California, Arizona, New Mexico, Utah, and Alaska. Over an 18-year period from 1990 to 2007, there were 41 reported cases of *Echinococcus*-related deaths in the United States [6]. The most recent formal incidence report of *Echinococcus* in the United States is from 1958, when an average of five cases were reported each year [7]. It can be found endemically in South America, select regions of Canada, the Middle East and eastern Mediterranean areas, Africa, western China, and the former Soviet Union. In South America, echinococcosis can be an important problem in countries such as Brazil, Peru, Chile, and Argentina. In regions of Argentina, surgical incidence rates for echinococcosis reach as high as 404 per 100,000 people [8]. It should be noted that this patient was seen at Beaumont Hospitals-Dearborn, a hospital located in Southeastern Michigan, an area with the highest Middle-Eastern population in the United States.

Due to the rare prevalence of *Echinococcus* in the United States, most physicians are unfamiliar with its management. In this case, this is evident in the reluctance of gynecologists and IR to intervene. Typical treatment involves removal of cysts followed by a regimen of albendazole for remission. A continuous regimen of albendazole for two to three months is ideal for preventing recurrence [9]. However, poor compliance to albendazole can be attributed to its adverse effects including nausea, vomiting, hair loss, and weakness, as is the case with many microtubule-inhibiting drugs. Cost is another factor. In the United States, a single dose is over 200USD (whereas it is 0.06USD in most developing nations). In addition to clearing the parasite, albendazole can help clear smaller cysts. Because of this effect, the patient was given albendazole preoperatively to clear any small cysts not grossly visible. Another method of treatment is PAIR (puncture, aspiration, injection, and re-aspiration), a procedure whose guidelines are published by the WHO [10]. This method was used to deal with the splenic cysts but failed, leading to recurrence of splenic cysts that ultimately led to splenectomy. After surgery, albendazole should be given to clear any residual disease that cannot be detected by imaging or during surgery. The high rate of recurrence of hydatid disease is attributable to these small cysts that can insidiously grow, preventing true remission. This patient's high rate of recurrence could also be related to her hydatid cyst rupture in 2006 (in addition to the nature of the disease). Recurrence can be detected through antibody titers. Antibody titers greater than 1:160 are considered positive. Sensitivity for serological testing ranges from 60% for pulmonary cysts to 90% for liver cysts [11].

This case demonstrates the potential difficulties with the treatment of *Echinococcus* due to the complex nature of the disease as well as the high rate of recurrence. Physicians need to be familiar with this condition and its management. Lack of familiarity may lead to misdiagnosis as well as inappropriate management. In addition to highlighting the value of proper treatment with experts of non-endemic diseases, this case illustrates the need for a high index of suspicion for rare conditions when managing a patient who is either an immigrant or has a travel history to high-risk areas. This is especially important in lieu of the increasing numbers of refugees coming into the US from the Middle

East that may carry this disease.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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