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Pancreatic echinococcosis in a 5-year-old girl

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ABSTRACT

Pancreatic echinococcosis accounts for 0.2–0.6% of cases, with the pediatric population being at a higher risk. Most commonly, pancreatic lesions occur in the head of the pancreas (50–58%); and in the body and tail in 24–34% and 19% of cases, respectively. Given the potential complications, surgery is usually performed. Albendazole is recommended before and after the surgery due to the risks for rupture and dissemination of protoscolices. Here we describe the case of a 5-year-old girl with progressive abdominal pain and cystic lesion in the pancreas compatible with echinococcosis in the ultrasound. The computed tomography showed bile duct compression. Indirect hemagglutination was negative. She had elevated total bilirubin, with a clear predominance of direct bilirubin, and high liver enzymes. Exploratory laparotomy, cholecystectomy, and unroofing of the cyst were performed. The patient had a favorable course and continued with albendazole for 3 months after the surgery.

Key words: *echinococcosis, pancreas, treatment, pediatrics.*

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INTRODUCTION

Echinococcosis or hydatid cyst is a zoonotic disease that is widely spread worldwide caused by *Echinococcus granulosus*. The most affected organs are the liver (67–89%) and lung (10–15%).[1] Location in the pancreas is extremely unusual.[2] Symptoms depend on the location and size of the cyst; imaging tests may be nonspecific and serology has low sensitivity, making etiologic diagnosis difficult when it is exclusively located in the pancreas. In Argentina, echinococcosis is distributed across the national territory and is a notifiable zoonotic disease of mandatory reporting. Echinococcosis is more prevalent in rural areas, especially in sheep and goat farming areas, with poor sanitary infrastructure, poor knowledge of the disease, and a dog population without veterinary care.[1] A pathological study is required to make an established diagnosis.[3] Given their location and potential complications, pancreatic cysts are not usually treated exclusively with medications.[4] Wide surgical resection, such as a pancreaticoduodenectomy or distal pancreatectomy, is often necessary.[5]

Figure 1. Ultrasound image showing a well-defined anechoic lesion with thick double-lined hyperechoic wall and echogenic internal structures arising from the head of the pancreas

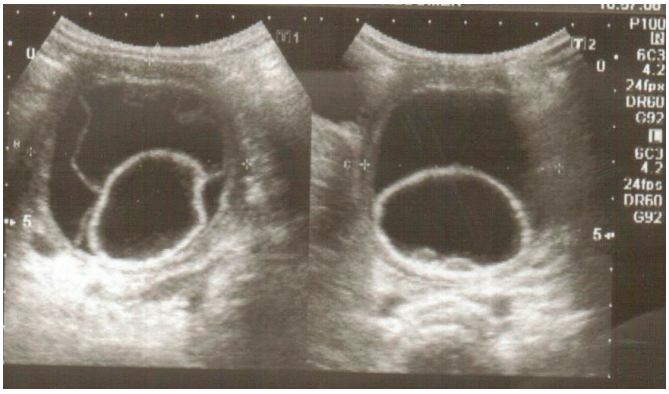


Figure 2. Computed axial tomography image showing a well-defined cystic structure that measures approximately 6.4 cm in diameter, without enhancement (black arrow), which is in continuity-contiguity with the head of the pancreas, with internal hyperdense foci. It shows involvement of the porta hepatis with marked dilatation of the intrahepatic and extrahepatic bile duct and the gallbladder (white arrow)

CASE REPORT

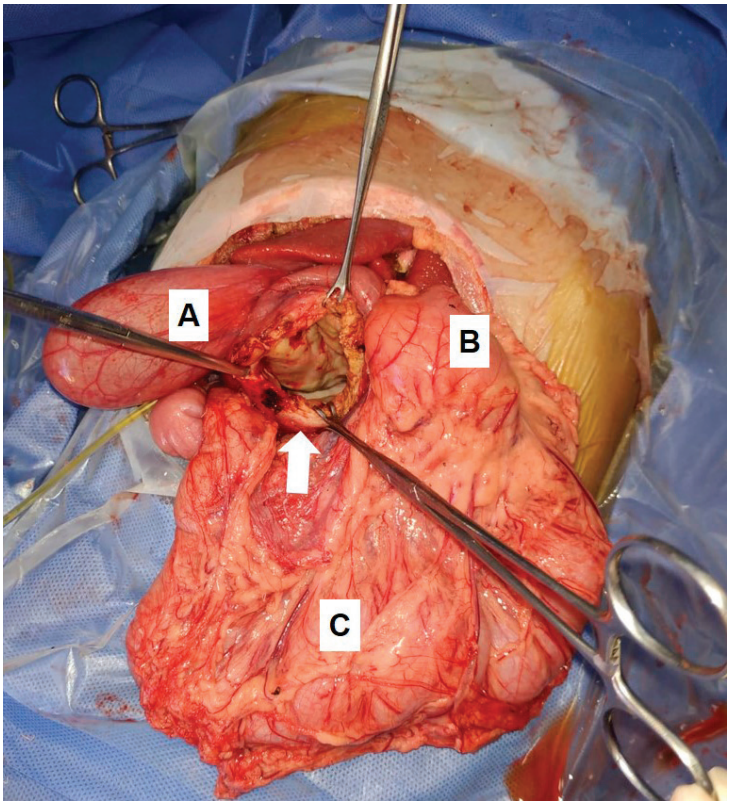
This was a previously healthy 5-year-old girl born in a rural area of the province of Chaco, in the Northeast region of Argentina. She lives with her mother and father in a brick house with well water, surrounded by sheep and goats.

The patient started with asthenia, nausea, loss of appetite, and progressive abdominal pain. The abdominal ultrasound (US) showed homogeneous liver, dilated intrahepatic and extrahepatic bile duct, tortuous, 12-mm common bile duct. The pancreatic cyst image showed 2 components: one had a collapsed internal membrane (type CE3) and the other contained daughter vesicles (type CE2),[6] and movement of internal echoes when the position was changed (falling snow sign), with a diameter of 60 mm (*Figure 1*). Other results were within normal ranges. Albendazole was administered orally at 15 mg/kg/day twice daily with a poor clinical course; the medication was discontinued due to vomiting and abdominal pain. The patient was referred to Hospital de Niños Dr. Ricardo Gutiérrez in the City of Buenos Aires. At that time, she had a regular general condition; low-grade fever (37.5 °C); a weight of 15.2 kilograms (3rd–10th percentile); a heart rate of 130 beats per minute; a respiratory rate of 20 breaths per minute; a blood pressure of 90/55 mmHg; soft, depressible, non-tender abdomen with hepatomegaly; and generalized jaundice. The rest of the physical exam was unremarkable. Intravenous hydration and expectant antibiotic management were indicated.

The abdominal computed tomography (CT) scan showed a cyst contiguous to the head of the pancreas that compressed the bile duct (*Figure 2*). The chest CT was normal. Indirect hemagglutination (IHA) was negative for echinococcosis. No alterations were observed in the blood count and kidney function. The liver function tests showed elevated total bilirubin, predominance of direct bilirubin, and a 3-fold increase in liver enzymes.

The condition was diagnosed as pancreatic echinococcosis and surgery was indicated. A laparotomy was performed with intra-operative cholangiography that showed that the pancreatic

Figure 3. Intra-operative images show a cystic image in the head of the pancreas (white arrow). A: gallbladder; B: stomach; C: greater omentum



cyst was compressing the extrahepatic bile duct. A cholecystectomy, cyst puncture and microscopic examination of cyst fluid, cyst unroofing (*Figure 3*), and germinative membrane removal (*Figure 4*) were performed; the specimen was sent for biopsy. Following the surgery, the patient had a favorable course, was discharged 9 days later, and returned to her usual activities.

Two months later the liver function tests returned to normal values. She continued taking albendazole for 3 months after the surgery and showed a good clinical course. The pathological study confirmed the diagnosis of echinococcosis. Six-monthly serological controls using IHA remained negative after 2 years. The control US at 3 months did not show cyst recurrence. The patient's weight and height remained in the 10th percentile during follow-up.

Her family was screened using IHA, US, and chest X-ray. Her father's tests were normal; her mother had a negative IHA and a normal chest X-ray, but her US showed a CE1 cystic lesion in the left hepatic lobe that measured 32 × 30 mm. She was started on albendazole 800 mg/day with an adequate response at 3 months of follow-up.

DISCUSSION

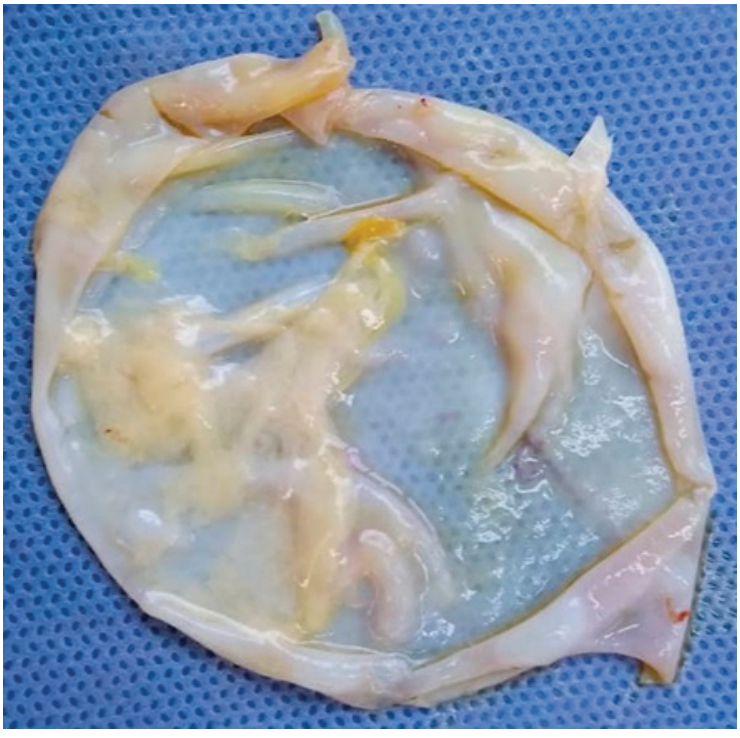
In endemic regions, the incidence rates of echinococcosis can reach more than 50 per 100 000 person/years, and its prevalence is as high as 5–10% in some areas of Argentina, Peru, East Africa, Central Asia, and China.[7] In Argentina, during 2018 and 2019, 658 and 555 cases in humans were reported to the National Health Surveillance System, respectively.[8]

Echinococcosis develops mainly in rural areas, where the presence of parasitized dogs, livestock (sheep, goats, pigs, sheep), and humans form a trilogy that sustains the parasitic cycle. Since dogs are the definitive hosts, the pediatric population is the most vulnerable due to close contact with them.[1]

Location in the pancreas accounts for 0.2–0.6% of cases;[2] it is found in the pancreatic head in 50–58%; and in the body and tail of the pancreas in 24–34% and 19%, respectively.[9] The main differential diagnoses include pseudocyst, common bile duct cyst, and tumors such as Frantz's tumor, serous or mucinous cystadenoma, and cystadenocarcinoma.[2]

Patients may be asymptomatic for many years, until the size of hydatid cysts causes clinical signs

Figure 4. Germinative membrane of hydatid cyst: whitish laminar fragment measuring 9.5 × 2.5 cm



due to pressure on adjacent structures, infection, rupture, and dissemination of cystic contents into neighboring body cavities.[10] In most published series, echinococcosis is usually asymptomatic or nonspecific, identified incidentally during imaging tests due to other problems or complications.[11] In our experience, most cases are diagnosed by findings in imaging studies or by palpation of a tumor during the usual physical exam or due to some other condition.[12]

Diagnosis is based on imaging tests and epidemiology. Our patient lived in a rural area, in contact with dogs without veterinary care and fed with raw offal of infected animals. She lived close to meat processing plants and slaughterhouses where animals are slaughtered for consumption.

The US is the technique of choice to diagnose abdominal echinococcosis and is usually complemented by CT and/or magnetic resonance imaging (MRI). The US allows to identify cyst course, size, location, and relationship with other organs. The US is the method of choice to assess the response to the parasiticide treatment by observing the collapse of the germinative membranes that fold into the cyst.[13] The CT and MRI are useful for detecting smaller cysts in unusual locations and for pre-operative studies in complicated cysts. Rounded or oval masses containing fluid, with well-defined edges and without enhancement after intravenous contrast are observed.[7,9] In our patient, the CT allowed us to better define the pancreatic origin.

Serology is performed using IHA and ELISA. Sensitivity ranges from 35% to 90%, depending on cyst stage. False negative results are usual in recent, type CE1 cysts—the most frequent type in children. Therefore, a negative serological test does not rule out infection.[13] In our patient, serology was negative at diagnosis and persisted unchanged after surgery. Serology is useful for post-treatment follow-up; cyst recurrence may be evidenced by seroconversion in previously negative patients or an increase in titers may be observed in patients with a previous positive serology. Although the optimal follow-up time for echinococcosis has not been established, it should be long term, at least 3–5 years, with no established maximum duration.[14]

An established diagnosis requires direct visualization of the scolex or a pathological study of the membranes.[3,14]

Currently, abdominal cysts require medical management with albendazole for at least 3 months[13] and, in the case of large cysts or those in a location at risk for complications, the resolution is via a surgical approach. If the cyst is in the pancreas, the treatment of choice is surgery in the form of a distal pancreatectomy for lesions of the distal pancreas and pericystectomy for lesions of the pancreatic head.[15] If surgery is not urgent, the suggestion is to indicate albendazole (10–15 mg/kg/day) since 15 days before and up to 1–3 months after the procedure due to the risk for rupture and dissemination of protoscolices.[1,10,12,15]

The early detection of echinococcosis, especially in low-resource settings, remains necessary to assist in deciding on the most appropriate treatment and to avoid further morbidity and mortality. The most important preventive measures are deworming dogs, not feeding them raw offal, preventing children from being licked by them, and hand-washing before eating.■

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