

GIANT PRIMARY PANCREATIC ECHINOCOCCOSIS

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Received: 29-XI-2024

Accepted: 13-I-2025

Abstract

Hydatid disease, a zoonosis caused by *Echinococcus granulosus* (EG), primarily affects the liver and lungs. Pancreatic involvement is exceedingly rare, representing only 0.2% of cases. In the reported case of a giant pancreatic hydatid cyst in a 22-year-old female patient.

This patient from La Rioja, Argentina, presented with abdominal pain, distension, and early satiety. A CT scan revealed a 16 x 12 cm cyst in the pancreatic head. Pre-operative albendazole was administered, followed by a cephalic duodenopancreatectomy due to cyst contact with the superior mesenteric vein. The postoperative period was uneventful, and the patient was discharged on postoperative day five. Pathological examination confirmed a hydatid cyst, with no recurrence at eight months' follow-up.

Pancreatic hydatid cysts are rare, with most cases being asymptomatic due to the slow growth of the cysts. Imaging modalities like CT and MRI are crucial for diagnosis, while serological tests can help, although their sensitivity is limited. The treatment approach depends on the cysts location and size. In this case, surgical intervention was necessary due to the size and symptomatic nature of the cyst. The patient's preoperative and postoperative management included albendazole to minimize recurrence risk.

Primary pancreatic echinococcosis is a rare manifestation of hydatid disease. This case represents the larg-

est pancreatic cyst reported to date in our knowledge, successfully managed with surgical intervention and albendazole therapy, with no disease recurrence during follow-up. Surgical treatment remains the gold standard for giant or symptomatic cysts.

Key words: echinococcosis, pancreas, abdominal pain

Resumen

Equinocosis pancreática primaria gigante

La enfermedad hidatídica, una zoonosis causada por *Echinococcus granulosus* (EG), afecta principalmente al hígado y los pulmones.

La afectación pancreática es extremadamente rara, representando solo el 0.2% de los casos. Presentamos un caso de un quiste hidatídico pancreático gigante en una paciente de 22 años.

Esta paciente de 22 años, procedente de La Rioja, Argentina, presentó dolor abdominal, distensión y saciedad temprana. Una tomografía computarizada reveló un quiste de 16 x 12 cm en la cabeza del páncreas. Se administró albendazol preoperatorio, seguido de una duodenopancreatectomía cefálica debido al contacto del quiste con la vena mesentérica superior. El período postoperatorio transcurrió sin complicaciones y la paciente fue dada de alta al quinto día postoperatorio. El

examen patológico confirmó un quiste hidatídico, sin recurrencia a los ocho meses de seguimiento.

Los quistes hidatídicos pancreáticos son poco frecuentes y la mayoría de los casos son asintomáticos, debido al lento crecimiento de los quistes. Las modalidades de imagen como la TC y la RMN son cruciales para el diagnóstico, mientras que las pruebas serológicas pueden ayudar, aunque su sensibilidad es limitada. El enfoque del tratamiento depende de la ubicación y el tamaño del quiste. En este caso, la intervención quirúrgica fue necesaria debido al tamaño y la naturaleza sintomática del quiste. El manejo preoperatorio y posoperatorio de la paciente incluyó albendazol para minimizar el riesgo de recurrencia.

La equinococosis pancreática primaria es una manifestación rara de la enfermedad hidatídica. Este caso representa, en nuestro conocimiento, el quiste pancreático más grande informado hasta la fecha, tratado exitosamente con intervención quirúrgica y terapia con albendazol, sin recurrencia de la enfermedad durante el seguimiento. El tratamiento quirúrgico sigue siendo el estándar de oro para los quistes gigantes o sintomáticos.

Palabras clave: equinococosis, páncreas, dolor abdominal

Hydatid disease is a zoonosis caused by the *Echinococcus* parasite larvae, which occurs globally but is endemic in South America¹. Four species of *Echinococcus* can infect humans, with *Echinococcus granulosus* (EG) being the most prevalent, responsible for over 95% of cystic echinococcosis cases². The EG life cycle involves dogs and other canids as definitive hosts, where the adult tapeworm resides in their bowels. Humans, acting as accidental intermediate hosts, become infected by consuming food contaminated with parasite eggs from the stools of these hosts³. The parasite can affect several organs, most commonly liver and lungs, which together account for approximately 90% of cases. Less frequently affected sites include muscles, bones, kidneys, brain, spleen, and extremely rarely pancreas, representing just 0.2% of cases⁴⁻⁶.

In Argentina, between 2019 and 2023, 3792 cases of hydatid disease were reported, of which 62.5% (n=2368) were confirmed. During the period under review, a decrease in cases was observed in 2020 (COVID-19 pandemic) compared to 2019. However, since 2021 the trend has been upward, reaching a total of 643 confirmed cases

in 2023, achieving the highest incidence rate for the period under analysis⁷.

In the reported case of a giant pancreatic hydatid cyst in a 22-year-old female patient, which was successfully treated with surgical intervention. To our knowledge this case represents the largest cyst reported to date in the bibliography.

Clinical case

A 22-year-old female patient from La Rioja province, Argentina, presented with general discomfort, nonspecific abdominal pain, abdominal distension, and early satiety. On physical examination, she was afebrile and hemodynamically stable. Her abdomen was flat and soft, with a palpable, slightly immobile mass in the right hypochondrium.

During the consultation, her Arc 5 test result was negative.

Epidemiology was positive for echinococcosis, considering her origin from an endemic region in our country and the fact that a first-degree family member from the same area had recently undergone surgery for a hepatic hydatid cyst.

Laboratory tests were unremarkable, with inflammatory markers such as erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) within normal limits. Liver and pancreatic enzyme levels were also within the normal range.

An abdominal scan CT with intravenous contrast revealed a giant cyst, measuring 16 x 12 cm, located in the head of the pancreas and in close contact with liver segment III. Inside the cyst, daughter cysts and parasitic membranes were clearly visible (Fig. 1).

An interdisciplinary committee was convened, and a surgical approach was decided, with albendazole administered for two weeks preoperatively and for four weeks postoperatively.

Exploratory laparoscopy was initiated, revealing the previously described giant solid-cystic tumor. A segmental resection of the affected liver segment III was performed using CUSA® ultrasonic aspiration. Following the Kocher maneuver, close contact between the tumor and the superior mesenteric vein was observed, leading to the decision to convert to an open approach.

A cephalic duodenopancreatectomy was performed using the Whipple technique. The cyst was successfully dissected from the superior mesenteric vein. The postoperative course was uneventful, and the patient was discharged on the fifth postoperative day.

Pathology report: Macroscopy: A 16 x 12 x 10 cm unilocular cystic lesion containing serous fluid and membranes, with a wall up to 5 mm thick.

Microscopy: The wall consists of an acellular laminated membrane and a cellular layer (germinal membrane) containing protoscolices (Fig. 2).

At eight months of follow-up, the patient remains asymptomatic, with no recurrence of the disease.

Informed consent was obtained for the publication of the case.

Discussion

The treatment of hydatid cysts in the pancreas is extremely rare, with few cases reported in the literature. The case described here involved

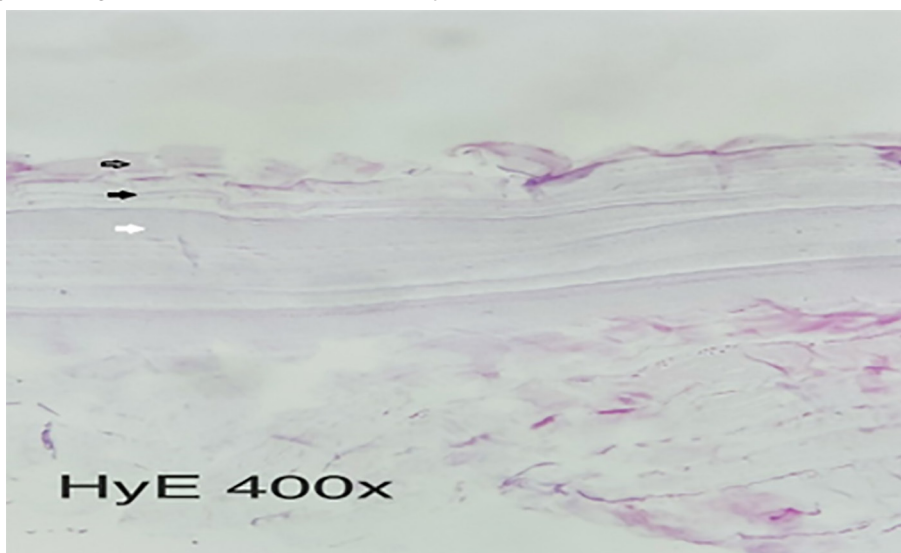
a large unilocular cyst in the head of the pancreas, in contact with liver segment III, requiring a complex surgical approach. This is the largest cyst reported to our knowledge.

Echinococcosis is an endemic zoonosis caused by the *Echinococcus granulosus* parasite, responsible for 95% of cases^{2,3}. Is a common disease in many parts of the world, with highest incidence in Uruguay (32 cases/100 000/year), Argentina (21 cases/100 000/year) and Morocco (7.2 cases/100 000/year)⁸. hydatid disease is exceptionally rare (0.2-1%) and can be classified as primary when it exclusively involves the pancreas (72%) or secondary when multiple organs are affected (28%). Cysts are usually solitary

Figure 1 | Abdominal CT scan with intravenous contrast revealed a giant cyst, measuring 16 x 12 cm, located in the head of the pancreas and in close contact with liver segment III



Figure 2 | Microscopic cut with hematoxylin-eosin staining where acellular laminate membrane is observed (arrow without filling), a cell layer called germ membrane (black arrow) and protoscolices (white arrow)



(90%), with 50% located in the pancreatic head, as in the present case, 30% in the body, and 20% in the tail of the pancreas⁹.

The majority of cases remain asymptomatic due to the slow growth rate of the cysts (0.3 - 2 cm per year), often taking several years before symptoms appear or being detected incidentally during routine examinations⁹.

The clinical presentation depends on the size, location, and relationship with neighboring organs. The most common manifestations include jaundice, cholangitis, epigastric pain, nausea, vomiting, or a palpable abdominal mass. Complications such as infection, acute pancreatitis, spontaneous or traumatic rupture into the peritoneal cavity, and mesenteric thrombosis have been reported. Cyst size can vary significantly; in our case, the hydatid cyst was considered giant because it exceeded 10 cm in diameter⁹.

The diagnosis of hydatidosis cyst is based on epidemiological history, physical examination, imaging studies, and serological tests. Hydatid disease should be suspected in the presence of a cystic mass, particularly when located in the abdomen or thorax and associated with epidemiological factors (such as place of origin, contact with dogs, or a family member previously diagnosed)⁹.

For diagnosis, detection, and monitoring of recurrence, the following serological tests are used: indirect hemagglutination (IHA), enzyme-linked immunosorbent assay (ELISA), immunofluorescence assay (IFA), and the Arc 5 test for echinococcosis. The seropositivity rate is higher in hepatic hydatid cysts compared to cysts in other organs⁶. However, seronegativity does not exclude hydatid disease, as serological tests report an average sensitivity of 62%¹⁰. In our case, the patient had a negative antibody measurement against the parasite's antigen 5, but she had an endemic epidemiological history and a first-degree family member from the same region who had undergone surgery for a hepatic hydatid cyst.

Markers of systemic inflammation, such as ESR and CRP, can be elevated in the presence of infection; however, they remained within normal limits in the patient described.

The diagnosis of pancreatic echinococcosis typically relies on imaging techniques such as

ultrasound (US), computed tomography (CT), and magnetic resonance imaging (MRI)^{9,11}. In more complex cases, a thorough evaluation may require the use of advanced invasive diagnostics, including endoscopic ultrasound (EUS) and endoscopic retrograde cholangiopancreatography (ERCP)⁹.

Ultrasound (US) is a non-invasive, cost-effective, and highly sensitive diagnostic tool⁸. Gharbi outlined the typical ultrasound appearance of hepatic hydatid cysts; however, while its application for pancreatic cysts is equally valid, it is less frequently used due to the retroperitoneal position of the pancreas and interference from bowel gases^{9,11}.

CT is highly effective in defining the cyst's size, location, and its relationship with the pancreatobiliary system, as well as in identifying cysts in other organs. It is also widely used for monitoring treatment progress and for the early detection of recurrences⁹.

In our case, the patient presented with a giant cyst that could be classified as Gharbi type I, displaying clear signs of parasitic activity.

The treatment decision depends on the cyst's location, specifically whether it is to the right or left of the portomesenteric axis. For small cysts (<5 cm) in asymptomatic patients without complications, regular monitoring with US or CT can be used to observe any changes in size or symptomatology¹⁰. Albendazole should be administered whenever parasitic activity is suspected¹⁰.

Preoperative treatment with albendazole was essential to reduce the risk of spillage and dissemination of parasitic material during surgery, as well as to minimize the risk of recurrence.

If the cyst is in the cephalic region of the pancreas, the trend is toward a more conservative surgical approach whenever possible (evidence level 5; recommendation D)⁹. Duodenopancreatectomy should be reserved exclusively for select patients. If the cyst is located in the tail or body of the pancreas, the resolution typically involves more radical surgery, with resection of the affected portion. In our case, a cephalic duodenopancreatectomy was performed due to the presence of a giant symptomatic cyst with liver involvement and contact with the superior mesenteric vein, which caused gastrointestinal symptoms¹⁰.

In symptomatic patients who are not candidates for surgery due to pre-existing comorbidities, or who have surgically unresectable cysts, one option is the PAIR technique (puncture, aspiration, injection of a scolical agent, and reaspiration of the cyst)⁸. This is a minimally invasive procedure performed under CT or ultrasound guidance, primarily useful for hepatic cysts. Potential complications include parasitic dissemination, risk of anaphylaxis, and cyst recurrence¹⁰.

Primary pancreatic echinococcosis is a rare manifestation of hydatid disease. We present the largest cyst reported to date. Surgical treatment remains the gold standard for giant or symptomatic cysts that impair the patient's quality of life.

Postoperative recovery was uneventful, and follow-up confirmed the absence of recurrence, consistent with expected outcomes in such cases when managed appropriately.

Conflict of interest: None to declare

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