Hydatid Disease Presenting as Pancreatitis

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Abstract

A 20 year old Turkish man presented with a one year history of weight loss and epigastric pain. Pancreatitis was confirmed with hyperamylasaemia. CT scanning of the pancreas revealed a complex pseudo-cyst and ERCP demonstrated a pancreatic duct stricture. Hydatid disease was suggested on Ultrasound scan but serological tests were negative. After ERCP the patient deteriorated with sepsis and this prompted surgery as gas was seen within the pseudocyst on repeat CT.

Laparotomy was performed and a large infected cyst in the head of the pancreas was opened and the contents evacuated. Cystostomy was anastomosed to a Roux loop of jejunum to allow drainage. Medical treatment with albendazole was given and the patient made a good recovery.

Hydatid cysts of the pancreas are rare and an even rarer cause of pancreatitis. Awareness of hydatid disease should be maintained even in non endemic areas as early diagnosis and treatment can avoid serious and potentially life threatening complications.

Key Words: Hydatid, Pancreatitis, Echinococcus granulosus, Cystostomy, Albendazole

Case history

A 20 year old Turkish man who had been living in the UK for 5 months presented with a one year history of epigastric pain and weight loss. Worsening abdominal pain and vomiting over the preceding week had necessitated hospital admission. He was noted to have worked with sheep and cattle in his youth. Clinical examination revealed epigastric fullness with localised tenderness, and pancreatitis was confirmed by a serum amylase of 1282 U/ml, with a White Cell count of 8.6 x10^9/L and an eosinophilia of 2%. Abdominal ultrasound demonstrated a large cyst in the left side of the abdomen with some features suggestive of hydatid disease (Fig. 1). Computerised Tomography showed features of chronic pancreatitis with multiple pseudocysts at the head of the pancreas. There was also dilatation of the pancreatic duct. Stool samples contained no ova, cysts or parasites and hydatid serology was negative. The patient was treated conservatively for chronic pancreatitis and discharged ten days after admission. He remained well, and an ultrasound performed 8 months later showed reduction in the size of the pseudocyst.

The patient however was readmitted 2 years later with another attack of pancreatitis. Again an obvious epigastric mass was palpable with localised tenderness. Serum amylase was elevated at 2089 U/ml, White Cell count of 19.3 x10^9/L and an eosinophilia of 68%. CT scan showed that the complex pseudocyst had enlarged from previously, and subsequent ERCP demonstrated a pancreatic duct stricture with a proximally dilated segment. The pseudocyst was in communication with the junction of the stricture and the dilated duct (Figs. 2,3). Following ERCP, the patients’ condition deteriorated with abdominal pain and sepsis. A further CT scan confirmed gas within the pancreatic collection and free fluid in the right para-renal space (Fig. 4). A laparotomy was therefore performed and the infected pseudocyst was drained via a cyst enterostomy using a Roux-en-Y anastomosis. The fluid cultured was heavily bloodstained pus with no organisms identified. The cyst was reported as being hydatid in nature, although no scolaces were seen. The post-operative course was uneventful, and he was discharged ten days later on albendazole.

Clinical Evidence

Cystic hydatid disease is caused by larvae of the canine tapeworm *Echinococcus granulosus*. Dogs are the chief mediators of the disease in humans. The tapeworm lives in the small intestine of the dog, and the terminal end containing eggs is shed in the dogs’ faeces. The intermediate hosts – usually sheep or humans – ingest these ova, they hatch in the upper jejunum and enter the portal circulation. The site of the final development of the larva is determined by the portal blood flow, the liver therefore being affected in approximately 70% of cases [1]. Less likely sites include the lungs, spleen, bone or brain. Rarely the breast,
muscles or pancreas are involved. Primary pancreatic involvement is found in less than 0.2% of cases of hydatidosis [2].

Hydatid disease therefore thrives in parts of the world where humans, sheep and dogs co-exist. In South America, Eastern Europe, Mediterranean countries and Australasia it is a major health problem. In the UK, Wales is recognised as a hydatid high risk area with a large proportion of farms having one or more resident infected dog.

Awareness of the possibility of hydatid disease however should be maintained even in non-endemic areas. Infestation usually occurs in childhood and the cysts can remain asymptomatic for years. Patients may complain of upper abdominal pain or a palpable mass, although they usually present as either an incidental finding on Ultrasound scanning or CT, or with one of the complications.

The natural history is for the majority of cysts to enlarge and the walls may calcify. Complications include jaundice, due to pressure of cysts on, or the presence of cysts within the bile ducts. Rupture of the cysts into the gut, biliary channels, pleura or abdominal cavity can also occur causing anaphylaxis with profound circulatory collapse.

Plain X-Rays may show calcification of the cyst although this does not necessarily infer death of the larvae. Typical Ultrasound and CT findings include multivesicular cysts and these can be pathognomonic.
Serological tests include Complement fixation, Indirect fluorescent antibody testing, ELISA and Specific immunoelectrophoretogram although they can be unreliable and false negatives are seen in up to 45% of patients with histologically proven hydatidosis [3]. If USS and CT do not show any diagnostic features, then
negative serology cannot exclude hydatid disease in a patient with an anechoic cyst. Blood count usually shows an eosinophilia of 6% or more.

Treatment of hydatid disease consists of surgery, chemotherapy and percutaneous aspiration. Surgery is the traditional treatment with conservative resection being appropriate in most patients. Care to excise the whole of the cyst wall, at the same time avoiding contamination of the abdominal cavity and wound with the cystic contents are the main principles.

In the presence of a stricture and a pseudocyst, cyst decompression using a Roux-en-Y anastomosis was a logical surgical treatment to prevent rupture of the cyst into the abdominal cavity. Distal pancreatectomy has been used in similar cases [4], and for others where the cause was considered to be pancreatic carcinoma [5]. We submit that Roux-en-Y is easier and safer.

Anti-helminthic therapy with albendazole is an important aspect of treatment. Toxicity includes liver function abnormalities, anaphylactic shock and rupture of the cyst following treatment. It is therefore usually confined for recurrent, disseminated or inaccessible cysts as well as for adjuvant therapy. It is unclear, however, whether the uncertain prognosis as occurs with the drug treatment of liver hydatid disease also applies in the pancreatic infection

Unusual features

The patient described here presented certain diagnostic difficulties and was initially managed as a case of chronic pancreatitis. Ultrasound scanning suggested the possibility of hydatid disease, although negative serology and CT appearances suggested that formation of a pancreatitis associated pseudocyst was more likely. Possibly there was too much reliance on the negative serology. The signs of sepsis, and gas within the pseudocyst as well as the clinical deterioration of the patient suggested that pancreatic necrosis was the cause and prompted the surgery.

The sequence of events leading to the pseudocyst formation is likely to have started with the hydatid infection of the pancreas. Rupture of hydatid cysts into the duct, or periductal pancreatic inflammation could both have produced the effect of ductal inflammation and fibrosis. After a stricture had been formed, further episodes of pancreatitis occurred, which predisposed to the formation of pancreatic cysts. These then have the potential to be colonised by the local hydatid infection.

Lesson

Awareness of the possibility of hydatid disease should be maintained even in non-endemic areas. Infestation usually occurs in childhood and the cysts can remain asymptomatic for years. Serological tests can be unreliable and false negatives are seen in up to 45% of patients with histologically proven hydatidosis.

References