A Case of Hepatic Hydatids Causing Biliary Obstruction: A Case Report and Review of the Literature

A 51-year-old white male presented to the emergency department of Strong Memorial Hospital in November with an abrupt onset of fever, and right sided chest and shoulder tip pain in the days preceding admission. He also had pale stool and dark urine. He denied nausea or vomiting but had noticed jaundice. His past medical history was significant for known HHD. He had previously had resection of an echinococcal cyst in May, 2000 and had been largely asymptomatic in the intervening 4 years. He had also been treated for 1 year (1999–2000) with lamivudine for chronic hepatitis B. He took no regular medications nor did he have any known drug allergies. Social history was revealing for an individual who had moved from Greece in 1970 and had worked as a property manager in the USA. He had a 20-pack-year smoking history and had on occasion used nasal cocaine. He denied I.V. drug use and drank socially. He had returned to Greece in 2003 on vacation.

Amongst the causes of cystic hepatic disease, hydatids (caused by the parasite *E. granulosus*) is common (endemic) in the Middle East, India, Africa, the south pacific (Australia and New Zealand), South America, China, and parts of Europe, but is less frequently seen in North America. Many of the cases in the USA are associated with immigrants from highly endemic areas. Significant morbidity and mortality can result from hepatic hydatids disease (HHD) and definitive therapy involves surgical resection with pre- and post-operative anti-parasitic medications. Rupture of the cyst into the biliary tree can lead to serious cholangitis.

We describe the case of acute cholangitis in an immigrant from Greece with a known history of HHD.
Family history was non-contributory. At admission he was noted to have a temperature of 39.2°C, blood pressure of 139/69 and respiratory rate of 18/minute. He was noted to be jaundiced and his abdominal exam was significant for a palpable liver edge 4cm below the right costal margin. His abdomen was soft and non-tender at admission with normal bowel sounds. The remainder of his exam was unremarkable. Labs were drawn and revealed an elevated white cell count of 14.5 with 62% segmented neutrophils, 19% lymphocytes and 11.5% eosinophils. His blood electrolytes and renal function were within normal limits. Liver function testing showed elevated transaminases (ALT 318 and AST 133), and evidence of cholestasis (total bilirubin 8.0 with direct bilirubin 6.2, and ALK 493). Amylase and lipase were within normal limits. His albumin at admission was 2.4. Ultrasound scan showed multiple cystic lesions in the liver and complex cystic masses. *(largest = 4.8 × 7.7 cm). There was marked intrahepatic ductal dilatation. CT of the abdomen showed hepatomegaly with multiple cysts in the gallbladder fossa. The common bile duct was dilated to 1.4 cm distal to the pancreatic head.

A diagnosis of acute cholangitis was made and it was suspected that biliary obstruction was directly related to hydatid cyst rupture with debris blocking the common bile duct (CBD). He was thus started on piperacillin/tazobactam and taken for ERCP on hospital day two. ERCP was performed with ampullary sphincterotomy leading to drainage of a large amount of yellow-white membranous debris from the CBD (Figures 1, 2). Two stents were placed in the CBD and the biliary drainage sent for culture. Out of the culture grew alpha hemolytic Streptococcus and coagulase negative Staphylococcus.

He was put on albendazole as well as piperacillin/tazobactam for HHD associated cholangitis. Post ERCP his t-bili came down and the patient was discharged home after 6 days in SMH on Augmentin and albendazole.

He was treated for acute cholangitis secondary to cyst rupture with cyst biliary communication and after completing >1 month of treatment with albendazole and Augmentin, he was electively admitted 1/25/05 for resection. Intraoperatively he was found to have inflammatory adhesions between intra-abdominal viscera and the hydatid cyst.

After operative exposure, aspiration, injection with hypertonic saline and eventual resection of the cyst was performed. A common wall was identified between the gall bladder and the cyst and a modified cholecystectomy was also performed. An intra-operative cholangiogram revealed debris within the common bile duct. The post operative course was complicated by fevers and the patient was treated again with IV antibiotics. CT of the abdomen at this time revealed that he had developed pneumoperitoneum and fluid (continued on page 73)
around the liver which was subsequently tapped and sent for culture. Both Candida albicans and vancomycin resistant enterococcus (VRE) grew out of the cultured peritoneal fluid and his antibiotics were rationalized to linezolid and Diflucan. He was discharged home with a percutaneous drain in situ draining 400–1200 cc/day. He had a repeat ERCP after 4–6 weeks with removal of debris from the CBD. He remains well after 10 months of follow-up.

In summary this is a 51-year-old male from an endemic area with a past medical history of hepatic hydatids disease who presented to the ER with acute cholangitis secondary to rupture of echinococcal cyst with biliary communication leading to debris in the common bile duct and obstruction.

DISCUSSION

Cystic lesions of the liver include simple cysts, hydatid cysts, cystadenocarcinomas/cystadenomas, abscesses and polycystic or multi-cystic liver disease. These can often be distinguished from one another based on symptomatology and radiological appearance.

Individuals with hydatid cysts are often asymptomatic but may develop pain as the cyst(s) enlarge. These cysts can rupture into the peritoneum leading to shock and sepsis or into the biliary tree causing cholangitis. Rupture into the pleural cavity can lead to empyema. In addition, secondary infection of the cysts can occur.

The hydatid cyst formation is directly related to the life cycle of E. granulosus. The mature worm lives in the bowel of several carnivores. Eggs are passed in the stool of dogs/wolves/other carnivores and are inadvertently consumed by grazing animals (e.g. sheep). The larvae migrate through the bowel wall and mesenteric vessels to the liver where they grow and become encysted. The hydatid cyst typically consists of a central germinal membrane which gives rise to daughter cysts surrounded by a thickened outer layer of inflammatory tissue. Carnivores devour the livers of these intermediate hosts and the scolices of the daughter cysts are released into the intestine where they grow further into mature worms.

Although the parasite E. granulosus can theoretically infect any organ the most common sites for human hydatid disease are the liver (55%–70%) followed by the lung (18%–35%). In about 5%–13% of cases these two organs are affected simultaneously. The lesions can be detected by ultrasound or CT. Definitive diagnosis is made on serologic testing (EIA, IHA, CIEP/Western Blot). Treatment is based on surgical resection and treatment with albendazole.

Radiologic tests and serology usually establish the diagnosis of HHD, but in cases where there is a suspicion of biliary obstruction ERCP is the test of choice since it can relieve the obstruction as well. Since the extruded material is thick, a large sphincterotomy is helpful so as to prevent stent clogging with parasitic debris. Also, a large stent must be left behind to prevent obstruction with the cyst remnants. Broad spectrum IV antibiotic coverage should be given to all patients with suspected biliary obstruction and cholangitis. This case clearly outlines the diagnostic and management strategies in patients who present with biliary obstruction in the context of hydatid cyst disease and cholangitis.

References