Echinococcus presenting as painless jaundice

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A previously well, 24-year-old male resident of Vancouver, British Columbia, presented to the emergency department with a two-month history of gradual onset of painless jaundice, dark urine and pale stool. At the time of presentation, he experienced a dramatic rise in the levels of the following liver enzymes: alkaline aminotransferase 272 U/L; alanine aminotransferase 244 U/L; gamma-glutamyl transferase 487 U/L; and alkaline phosphatase 1085 U/L, with an international normalized ratio of 1.0 and an albumin level of 39 g/L. His serum bilirubin level was highly elevated (367 μmol/L) with a conjugated fraction of 282 μmol/L. Hepatitis was the presumptive diagnosis based on positive serology for hepatitis A. However, a computed tomography scan of the abdomen revealed a complex, cystic mass 10.9 cm × 17.9 cm in size in the epigastric region with rim calcifications (Figure 1). This was concerning for a malignant or infective process and the patient was referred to the hepatobiliary surgery service.

The patient experienced no fever, cough, nausea, vomiting or abdominal pain. He had no known contacts with similar symptoms. He had travelled to Belgium, but had not travelled to the tropics or Northern Canada. There was no known exposure to animals, including canines.

He underwent an endoscopic retrograde cholangiopancreatography with placement of bilateral biliary stents and ultrasound-guided percutaneous drainage of the hepatic abscess. The subsequent culture of the abscess fluid grew Candida albicans, which was treated with fluconazole. The patient became febrile and his jaundice did not improve and, thus, a right hepatectomy and cholecystectomy was performed. The surgical specimen contained a complex hydatid cyst 12 cm in diameter (Figure 2). There were numerous hyalinized profiles in the cyst wall consistent with daughter cysts (Figure 3). The final pathological diagnosis was echinococcus with features suggestive of Echinococcus multilocularis. The patient was treated postoperatively with albendazole, 400mg/day orally. Subsequent serology was positive for serum immunoglobulin G antibodies to echinococcus confirmed by immunoblot. He remains asymptomatic without evidence of recurrence and his liver enzyme levels have normalized.

DISCUSSION

Echinococcus species are small tapeworms (cestodes) of carnivores; the main reservoir are canines. Two forms of Echinococcus cause disease in humans. The more common Echinococcus granulosus causes cystic echinococcosis and the rare Echinococcus multilocularis causes alveolar echinococcosis (AE) (1). Within canines, the definitive host, Echinococcus produce eggs that are excreted in the feces of infected animals. Humans are typically infected either through handling of infected animals or by ingestion of food contaminated with the eggs. The larvae of Echinococcus travel through the portal system to the liver where they form metacestode tissue: in the case of cystic echinococcosis (CE), a single cyst (hydatid); in the case of AE, a mass of small cysts or vesicles. The cysts are often associated with an inflammatory reaction in adjacent tissue. As the cyst(s) expands there are mass effects and, in the case of AE, the multiple cysts invade surrounding structures and metastasize, resulting in a high mortality rate without appropriate treatment (2,3).

Echinococcus infection in humans often remains asymptomatic for five to 15 years and typically presents with gradual onset of subtle symptoms. Similar to the present case, patients may present with cholestatic jaundice related to the mass. The diagnosis of echinococcus is based on the triad of clinical history and exposure, imaging studies and immunodiagnostics, which typically include serum antibodies detected by ELISA and confirmed by immunoblot (2). Computed tomography imaging may reveal large cysts with peripheral calcifications or scattered calcific foci (4). Surgery and adjuvant therapy are the standard treatment. In the case of AE, although there is limited evidence and no prospective randomized controlled trials, it appears that radical surgery including pericystectomy and hepatectomy is the most effective treatment (5). Adjuvant therapy with albendazole improves survival and should be continued for two years postoperatively (2).

The present case represents the clinical and pathological findings of echinococcus presenting as painless jaundice related to a mass that was successfully treated with surgery and medical therapy.
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REFERENCES

Figure 2) A Lining of echinococcal cyst demonstrating the thick laminated layer produced by the parasite with adjacent degenerated and inflamed tissue (hematoxylin and eosin stain, original magnification ×100). B Internal aspect of a cyst with parasite remnants and debris (hematoxylin and eosin stain, original magnification ×200). C A protoscolix with hooklets (hematoxylin and eosin stain, original magnification ×400)

Figure 3) Multiple collapsed small cysts in the hepatic parenchyma. A The wall is composed of carbohydrates as demonstrated by periodic acid-Schiff positivity (original magnification ×100). B The presence of multiple small cysts produces an infiltrating appearance mimicking a neoplastic process (hematoxylin and eosin stain, original magnification ×100)
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