An unusual cause of obstructive jaundice

CASE REPORT

A 65-year-old man presented with a three-month history of progressively worsening jaundice associated with pruritus, clay-coloured stools, dark-coloured urine and weight loss of 15 kg over this period. His medical history was unremarkable for recurrent infection and exposure to pigeons (see Discussion, below). Clinical examination showed deep jaundice and an enlarged liver. Baseline investigations showed obstructive jaundice with a serum bilirubin level of 10 mg/dl (upper limit of normal 1 mg/dl) and serum alkaline phosphatase of 630 U/l (upper limit of normal 150 U/l). The patient's full blood count, globulin and albumin levels were normal. Pulmonary nodules were seen on his chest X-ray. A computed tomography (CT) scan of the chest, abdomen and pelvis showed calcified lung nodules, nodes in the porta hepatis causing biliary obstruction and consequent intrahepatic cholestasis and nodes in the root of the mesentery (Figure 1).

After a review of these investigations, the patient underwent endoscopic retrograde cholangiopancreatography (ERCP), where common bile duct and common hepatic duct stricture extending into the right system were noted (Figure 2). Brushings were obtained at the time of this procedure, and a plastic stent was inserted. A presumptive diagnosis of Klatskin tumour was made, but the brushings showed no evidence of malignancy. Hence a CT-guided fine needle aspiration (FNA) of a pulmonary nodule was performed. The aspirate showed granulomatous inflammation, clusters of yeast cells and no malignant cells (Figure 3). This specimen was smear- and culture-negative for Mycobacterium tuberculosis. The serum cryptococcal antigen and human immunodeficiency virus (HIV) status were checked: the patient tested negative for both. Nevertheless a diagnosis of disseminated fungal (cryptococcal) infection was made.

KEYWORDS Biliary obstruction, Cryptococcus neoformans, disseminated fungal infection, immunocompetent, obstructive jaundice

DECLARATION OF INTERESTS No conflict of interests declared.

ABSTRACT Cryptococcus neoformans is an opportunistic fungal infection that mainly affects immunocompromised patients with or without HIV and can only be seen infrequently in the immunocompetent host. Presentation can be with pneumonia, meningitis or widely disseminated disease. We report the case of a 65-year-old man who presented with obstructive jaundice and weight loss. He had been evaluated at another institution where surgery was offered to him for a presumed diagnosis of cholangiocarcinoma. Our evaluation revealed disseminated cryptococcal infection for which the patient received fluconazole for six months, which resulted in the resolution of his obstructive jaundice.

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FIGURE 1 CT scans of the chest (A) and abdomen (B) showing calcified lung nodules (indicated by arrow), nodes in the porta hepatitis causing biliary obstruction and consequent intrahepatic cholestasis and nodes in the root of the mesentery of a 65-year-old male with a three-month history of progressively worsening jaundice associated with pruritus, clay-coloured stools, dark-coloured urine and weight loss of 15 kg.
and the patient was started on fluconazole 400 mg once daily. On treatment, his appetite improved, he regained his weight and his liver function tests and serum biochemistry improved. A three-month follow-up ERCP revealed the resolution of the biliary stricture, and the stent was removed (Figure 4). A three-month follow-up CT scan revealed a decrease in size of the pulmonary nodules and the disappearance of the hilar lymphadenopathy, but a lumbar mass was still present.

DISCUSSION

A presentation of painless, progressive obstructive jaundice associated with weight loss in a 65-year-old man prompts a diagnosis of either cholangiocarcinoma or carcinoma of the pancreatic head unless proven otherwise. The mean age of patients presenting with pancreatic cancer is 55–65 years. Cryptococcus infection presenting as obstructive jaundice is a rare clinical entity.

C. neoformans is a ubiquitous organism found in mammal and bird faeces, particularly in pigeon droppings. It is present in two forms, either as Cryptococcal neoformans var. neoformans or Cryptococcal neoformans var. gattii. C. neoformans var. neoformans consists of serotypes A and D, which cause disease in patients with immune suppression, while C. neoformans var. gattii consists of serotypes B and C, which may cause disease in normal
hosts. *C. neoformans var. gattii* infection can exist as a harmless colonisation of the airways but may also cause meningitis or be disseminated, most often affecting the lung, brain, skin and prostate. The medullary cavity of bones is somewhat less commonly involved. Other, even less common sites are the myocardium, retina, liver, peritoneum, kidney, adrenal glands and muscles. Those individuals most susceptible to infection are patients with T cell deficiencies.

We could find only nine cases in the reported literature of cryptococcal infection with hepatitis or cholangitis with cholecystitis as the initial manifestation. Laparotomy was reported as being performed in four cases as there was a clinical suspicion of an acute abdomen. To date, the only two reported cases we could find presenting as ‘hepatitis’ were associated with an immunocompromised status. A preliminary diagnosis of cryptococcal infection is made by an identification of the yeast in a compatible clinical setting. Definitive diagnosis is confirmed by culture, most often from the cerebrospinal fluid or blood. Serum cryptococcal antigen has a sensitivity of 90% in cryptococcosis. However, there are case reports of negative serum cryptococcal antigen in disseminated cryptococcosis. A culture of bile can also be helpful in the diagnosis of cryptococcal cholangitis, and one recent case report particularly emphasises the usefulness of bile culture for the diagnosis of cryptococcosis.

The most sensitive diagnostic modality in suspected biliopancreatic diseases is ERCP. Endoscopic retrograde cholangiopancreatography is not only diagnostic but also offers therapeutic advantage with stent placement and release of obstruction. In cryptococcal cholangitis the cholangiogram mimics primary sclerosing cholangitis. Radiological investigation can be of help in diagnosing cryptococcal cholangitis, particularly magnetic resonance (MR) imaging. The intra- and extrahepatic biliary dilatations and contours of the bile duct are more accurately defined by T1- and T2-weighted biliary MR imaging. There are no definitive guidelines for the management of cryptococcal infection in an immunocompetent host. We followed guidelines published by the Infectious Diseases Society of America for the management of cryptococcal disease.

Our case of disseminated cryptococcosis in an HIV-negative (presumed immunocompetent) host presented a unique diagnostic challenge with the involvement of the biliary system, abdominal lymph nodes and lungs.

REFERENCES