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## Biliary cryptococcosis in a child: MR imaging findings

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**Abstract** Cryptococcosis is a systemic mycosis with a worldwide distribution. It frequently occurs in patients who are immunologically compromised or chronically ill. Clinical manifestations are usually confined to the central nervous system, lungs and skin. Involvement of the hepatobiliary system is very rare. We describe the MR imaging appearance of a rare case of disseminated cryptococcosis in an immunocompetent child in whom the clinical presentation was dominated by biliary and lymph nodal involvement.

**Keywords** Biliary cryptococcosis · Obstructive jaundice · MRI · Child

### Introduction

Cryptococcosis is systemic fungal infection caused by *Cryptococcus neoformans*. It is frequently seen in immunocompromised patients and the most common predisposing factor worldwide is AIDS [1, 2]. It commonly involves the lungs, skin and central nervous system. Involvement of

the biliary system is very uncommon. We present here the MR imaging appearance of biliary involvement in a child with disseminated cryptococcosis presenting as obstructive jaundice.

### Case report

A 5-year-old boy was admitted to the paediatric ward with high-grade intermittent fever of 2 months' duration. One week after the onset of fever, he developed painless progressive jaundice, dark urine and clay-coloured stool. Two days prior to admission, he had developed an erythematous rash over his face that progressed to involve the abdomen and extremities. On physical examination the child appeared sick and irritable. He was febrile and had tachycardia, pallor, deep icterus and clubbing. There was a maculopapular rash over the face, upper abdomen and lower limbs. Multiple cervical and bilateral axillary lymph nodes were palpable. The nodes were firm, non-tender, discrete and mobile. Abdominal examination revealed firm hepatomegaly and splenomegaly. Chest, cardiovascular and neurological examinations were normal. Laboratory investigations showed a haemoglobin level of 7.2 g/dl and total leucocyte count of 29,000/mm<sup>3</sup> with 76% neutrophils. Liver function tests were abnormal, with a total bilirubin of 6.7 mg/dl (conjugated 4.6 mg/dl), alanine aminotransferase 169 IU/l, aspartate aminotransferase 197 IU/l and serum alkaline phosphatase 903 IU/l. Serum tested negative for all viral hepatitis markers. Blood and urine cultures were sterile.

Abdominal ultrasonography revealed dilatation of intrahepatic biliary radicals and diffuse irregular wall thickening of the common bile duct (Fig. 1). MR imaging coupled with MR cholangiopancreatography (MRCP) was performed to further characterize the bile-duct wall thickening and to determine the severity of biliary obstruction. Axial T2-W images (TR/TE 4,500/80 ms) showed moderate dilatation of intrahepatic biliary radicals in both lobes of the liver with periductal soft-tissue thickening involving the right hepatic duct, left hepatic duct and confluence

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(Fig. 2). There was gross circumferential mural thickening of the common bile duct causing luminal narrowing as far as its lower end (Fig. 2). The periductal soft tissues appeared hypointense on T2-W images (Fig. 2). MRCP also revealed moderate dilatation of intrahepatic ducts in both lobes of the liver with a blocked confluence. Dilatation of the main pancreatic duct was also seen (Fig. 3).

Liver biopsy revealed canalicular cholestasis with inflammation of portal tracts. Axillary lymph node aspiration biopsy and fine-needle aspiration cytology from the common bile duct wall revealed granulomatous inflammation and numerous encapsulated fungal elements positive for mucicarmine stain consistent with *Cryptococcus* (Fig. 4).

The patient was investigated for immunocompromised status. He was HIV-negative with a normal CD4 and CD8 counts. Phagocytic function of the neutrophils was assessed with the nitroblue-tetrazolium test, and this was also normal. He was treated with intravenous amphotericin B (0.7 mg/kg per day) for 10 weeks with close monitoring of cell count and renal function. He became afebrile following treatment. There was significant improvement of his liver function tests. Follow-up MRCP after 6 weeks showed resolution of the periductal soft tissue around the common bile duct. The patient was discharged in a stable condition on oral fluconazole and at the time of this report 6 months later remained under regular follow-up.

## Discussion

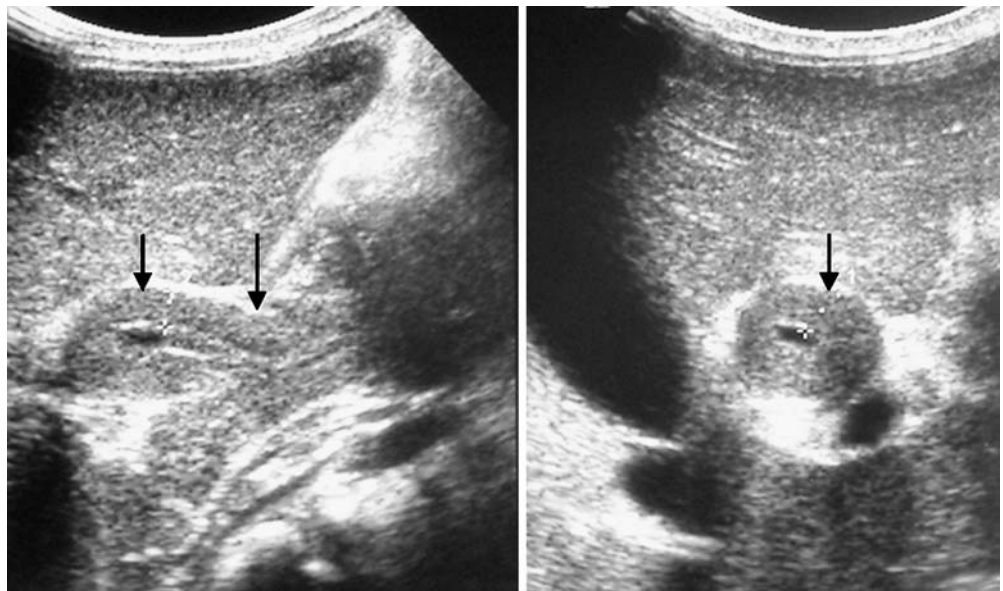
Cryptococcosis is caused by a yeast-like fungus *Cryptococcus neoformans*, which reproduces by budding and forms round yeast-like cells. It is an uncommon disease and is classified into two groups according to the immune status of the patient. The localized form is seen in immunocompetent patients, while the disseminated form is common in

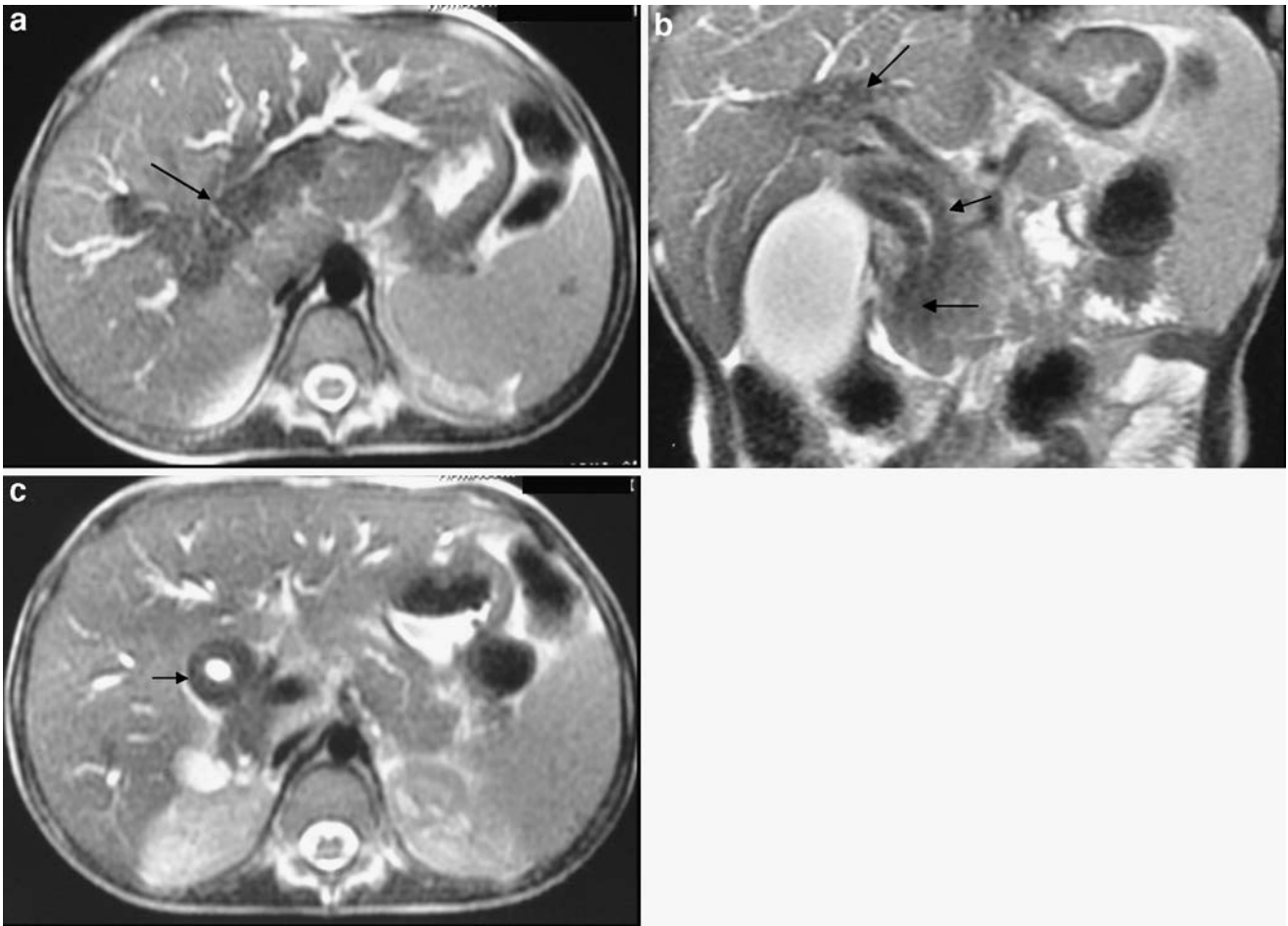
immunocompromised patients [1]. The most common predisposing factor for cryptococcosis worldwide is AIDS and more than half of patients have a history of receiving glucocorticoids or other immunosuppressive drugs prior to the fungal infection. Other predisposing conditions include solid organ transplantation, lymphoma, sarcoidosis and idiopathic CD4 lymphopenia [2]. Cryptococcosis is acquired by inhalation of the fungus into the lungs, although rare cases of cutaneous cryptococcosis appear to arise after minimal trauma. Pulmonary infection has a tendency for spontaneous resolution, while most patients present with meningoencephalitis [2].

Cryptococcosis presenting as diffuse biliary wall thickening or hepatobiliary dysfunction as the initial presentation is very rare. On review of the literature we found six such case reports to date, one of which was diagnosed on post-mortem examination and only one case was reported in an 8-year-old immunocompetent boy [3–8]. Apart from hepatobiliary involvement, two patients had lymphadenopathy and three patients developed meningoencephalitis in the later part of their illnesses [3–8]. Five patients received antifungal therapy, of whom three improved clinically following treatment. One patient had cirrhosis, even after treatment, and another young girl died before completion of antifungal treatment due to upper gastrointestinal bleeding and hepatorenal syndrome [8]. There have been a few additional case reports of hepatic cryptococcosis without biliary system involvement presenting as viral hepatitis, acute abdomen and hepatic failure [9–12]. Although one report was of a child with disseminated cryptococcosis who presented with sclerosing cholangitis and hepatobiliary dysfunction [5], our patient uniquely illustrates the MR and MRCP appearance of biliary cryptococcosis.

The radiological imaging of suspected surgical obstructive jaundice in a child comprises ultrasonography, contrast-enhanced CT and MR imaging coupled with MRCP. All these techniques can show the degree, as well

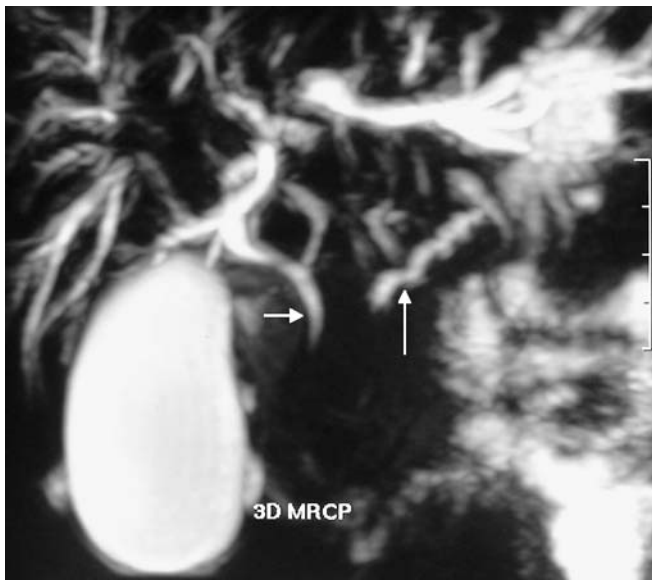
**Fig. 1** Abdominal US scans showing diffuse irregular wall thickening of the common bile duct (arrows)



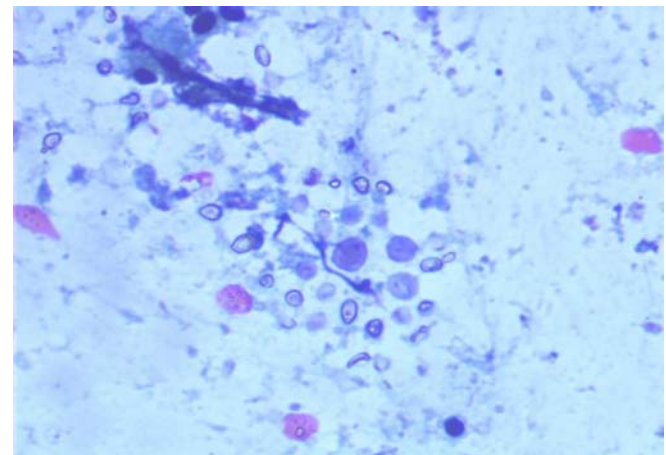


**Fig. 2** MR images. **a** Axial T2-W image showing moderate dilatation of intrahepatic biliary radicals in both lobes of the liver with periductal soft-tissue thickening (*arrow*) extending to the confluence. **b** Coronal T2-W image showing hypointense soft tissue

around the confluence (*arrow*) extending up to the lower end of the common bile duct (*arrow*). **c** Axial T2-W image showing hypointense gross circumferential mural thickening of the common bile duct (*arrow*)



**Fig. 3** MRCP showing dilated intrahepatic ducts and common bile duct (*short arrow*) with abrupt cut-off of its lumen in the lower part. The proximal main pancreatic duct is also dilated (*long arrow*) whereas the distal duct is not seen



**Fig. 4** Photomicrograph showing budding hyphal forms of cryptococcus in a necrotic background (Papanicolaou smear, x200)

as the level of obstruction. Sonography and MR are the preferred modalities because of the lack of ionizing radiation. The differential diagnosis in children includes choledochal cyst, chronic pancreatitis, primary sclerosing cholangitis or extrinsic compression by lymph node, portal

cavernoma or pancreatic mass [13–17]. The findings on sonography, CT and MR imaging include intra- and extrahepatic biliary dilatation, wall thickening, pruning, beading and skip dilatation [13–16]. MRCP has now virtually replaced endoscopic retrograde cholangiopancreatography (ERCP) for the diagnosis of biliary pathology. Breath-hold techniques that are very robust in adults cannot be used in uncooperative or sedated children. The signal acquisitions can be increased when respiratory and cardiac gating are used to increase the signal-to-noise ratio.

In conclusion, the MR imaging appearances of a rare case of disseminated cryptococcosis in an immunocompetent child with obstructive jaundice are presented. Unique to our case was the extensive periductal soft tissue as seen on MR imaging and MRCP, which resolved following antifungal treatment.

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