Case Snippets

Hepatobiliary dysfunction as initial manifestation of disseminated cryptococcosis

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Disseminated cryptococcosis presenting as biliary obstruction is rare. We report a 35-year-old HIV-negative man who presented with clinical features suggestive of obstructive jaundice, and radiological features suggestive of Ktalskin's tumor, but who ultimately was found to have cryptococcal involvement of the liver and biliary tract as part of disseminated cryptococcosis. The patient responded to antifungal therapy. [Indian J Gastroenterol 2004;23:145-146]

Keywords: Obstructive jaundice

C. neoformans usually causes an asymptomatic pulmonary infection followed by development of meningitis, which often is the indication of the disease. Disseminated cryptococcosis presenting with primary hepatocellular dysfunction is rare, with only nine cases reported in literature. Immune defects are common in patients with meningitis or disseminated cryptococcosis; however, only two reported cases were associated with immunocompromised status.

A 35-year-old man presented with jaundice and right upper quadrant dull achying abdominal pain of 1-month duration. He also had itching and nausea, with occasional vomiting. There was history of passing clay-colored stools. There was no history of fever, weight loss, anorexia or previous episode of jaundice. Examination revealed 2 cm tender hepatomegaly below the right costal margin. No other palpable lump was present.

Investigations: total bilirubin 13 mg/dL (direct 10), serum alkaline phosphatase 5000 KA units. Serology was negative for HBV, HCV and HIV. Ultrasonography was suggestive of Ktalskin's tumor. The patient underwent spiral CT scan that revealed moderately dilated intrahepatic biliary radicles with obstructing lesion at upper end of common bile duct. MRCP was also suggestive of Ktalskin's tumor (Fig).

After about 10 days, the patient developed severe persistent headache, followed by excessive drowsiness and altered sensorium. CT scan of the brain was normal. CSF analysis was positive for cryptococcal antigen on India ink staining; CSF culture grew Cryptococcus neoformans. A 1 cm x 1 cm sized left-sided enlarged cervical node was biopsied. It was suggestive of cryptococcosis.

The patient was treated with amphotericin B 1.5 g over 2 weeks, followed by fluconazole for 12 weeks; percutaneous biliary drainage was done for relief of jaundice. Bile culture grew Cryptococcus neoformans. Over 6 weeks, the biliary drainage from the catheter decreased from 400 mL/day to 3-5 mL/day. Follow-up cholangiogram did not show any obstructive lesion. Bile cultures following antifungal treatment were negative.

C. neoformans var. gattii is an encapsulated yeast and has four serotypes: A and D (Cryptococcus neoformans var. neoformans) and B and D (Cryptococcus neoformans var. gattii). Serotype A causes most cryptococcal infections in patients with defective cell-mediated immunity. Patients infected with C. neoformans var. gattii usually are immunocompetent, respond slowly to treatment, and are at risk of developing intracerebral mass lesions, e.g., cryptococcomas.

C. neoformans var. gattii infection results in a spectrum from harmless colonization of the airways to meningitis or disseminated disease. Lung and neural infection are the two most common sites. Skin, prostate, and medullary cavity of bones are the next most common involved organs in disseminated cryptococcosis. Other less common organs involved are myocardium, retina, liver, peritoneum, kidney, adrenals and muscles.

Only nine cases of cryptococcal infection with hepatitis, cholangitis and cholecystitis as initial manifestation are reported in literature. Four of these had been subjected to laparotomy for clinical suspicion of acute abdomen. Cryptococcosis is known to occur in immunocompromised patients, yet only two reported cases presenting as hepatitis were associated with immunocompromised status.

The diagnosis can be established by biopsy or fluid examination by fungal stains and cultures. In our case, India ink, which outlines the organisms by negative contrast, was used to identify the yeast cells. In addition, CSF culture, bile culture and histology of lymph node also revealed the infection.

In immunocompetent patients, the aim of treatment is to achieve permanent cure of the fungal infection. Amphotericin B can be administered alone for 6-10 weeks or in conjunction with fluconazole for 2 weeks, followed by fluconazole for a minimum of 10 weeks. None of the alternative forms of amphotericin B are superior to standard non-lipid amphotericin B. Fluconazole is unreliable if used alone and resistance develops very rapidly.

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Gastric zygomycosis – an unusual cause of massive upper gastrointestinal bleed

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We report a 62-year-old man with cardiac failure and acute renal failure, who had massive hematemesis. Upper GI endoscopy showed a large gastric lesser curvature ulcer. Bilroth II gastrectomy specimen showed fungal invasion. He received amphotericin B postoperatively, and recovered uneventfully. [Indian J Gastroenterol 2004; 23:146-147]

Key words: Mucormycosis, stomach

Zygomycosis refers to a group of uncommon but frequently fatal mycosis caused by fungi of class Zygomycetes. The spectrum of disease ranges from localized cutaneous to disseminated disease. Rhinocerebral and pulmonary disease are the most common forms, followed by gastrointestinal infections. Stomach is the most frequent site of involvement (67%), followed by colon (21%), small intestine (4%) and esophagus (2%). Although invasive gastrointestinal mucormycosis has been reported with fatal outcome, our patient, who presented with massive upper gastrointestinal bleed, had successful outcome after resectional surgery followed by antifungal therapy.

A 62-year-old man, a known hypertensive, was admitted in the cardiac unit with cardiac failure and acute renal failure. He was diabetic for the last seven years and had good glycemic control on oral hypoglycemic agents. He was recovering well when after one week of hospitalization, he had sudden onset of massive upper gastrointestinal bleed with hemodynamic instability. Upper gastrointestinal endoscopy revealed a large ulcer in the body of the stomach on the lesser curvature. With a suspicion of malignancy, the patient was operated upon. After gastrectomy, multiple ulcers were seen in the body of the stomach, varying in size from 1 cm x 1 cm to 4 cm x 4 cm, with black slough covering them. Bilroth II gastrectomy was performed.

Microscopic examination of the resected stomach showed broad, aseptic hyphae with irregular walls and frequent perpendicular branching, foreign body giant cells and eosinophils; the hyphae were invading the gastric wall as well as blood vessels. Postoperatively the patient received amphotericin B (1 mg/Kg/day) for three weeks and had uneventful recovery.

Gastrointestinal variants of zygomycosis are uncommon; most previous reports have come from South Africa with few cases from America, Europe and the Indian subcontinent. Gastric zygomycosis can be categorized into three forms: colonization, infiltration, vascular invasion. Colonization usually occurs in pre-existing gastric ulcers and is not fatal. Invasive form of gastric mycosis has variable presentation and is usually fatal. This form can either invade pre-existing peptic ulcer or can invade the stomach de novo.

The most frequent presentation of gastric zygomycosis is perforation, bleeding or epigastric discomfort. Lawson and Schuman reported seven cases of invasive gastric mycosis, all presenting with perforated viscus, diagnosed either intra-operatively or at autopsy. Thomson et al. reported that all their seven patients with peptic ulcer disease and gastrointestinal mucormycosis, presented with complications. At upper gastrointestinal endoscopy, the ulcers usually appear malignant with rolled edges, but are usually covered with black necrotic tissue unlike the white slough of gastric ulcer. Biopsy usually reveals fungal hyphae.

In the absence of complications medical management with antifungal agents is usually successful. In those with complications, resectional surgery followed by amphotericin B therapy is preferred. Non-resectional surgery carries high mortality. In one series all patients died after simple suture of perforation. Ninety percent of patients in another series underwent surgery for gastric mucormycosis and had successful outcome. Park et al. reported success with resectional surgery followed by antifungal therapy in bleeding peptic ulcer complicated by gastric mucormycosis.

Increased awareness and early resective surgery in complicated gastric zygomycosis, followed by effective antifungal therapy, provides improved survival in patients with this otherwise fatal disease.

References

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