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Disseminated angioinvasive basidiobolomycosis with a favourable outcome

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ABSTRACT

Basidiobolomycosis, a rare fungal infection, is of worldwide distribution but areas commonly involved include the tropical areas of Africa, USA and South East Asia. 88% of the cases are reported among patients younger than 20 years. Many of the case reports in Saudi Arabia are from Tohama area where our patient lives. The diagnosis tends to be overlooked as the presentation may mimic colonic carcinoma in adults or inflammatory bowel diseases and tuberculosis in both children and adults. Angioinvasion seen in our patient is extremely rare suggesting the diagnosis of mucormycosis and resulting in a delay in choosing the most appropriate treatment. We report this case to remind physicians and surgeons to consider this diagnosis in patients from endemic area presenting with such conditions.

1. Introduction

Basidiobolomycosis is a rare fungal disease caused by *Basidiobolus ranarum*. It is increasingly recognised as one of the chronic non-angioinvasive subcutaneous fungal infections in immunocompetent patients. Gastrointestinal involvement is rare. The majority of reported cases of gastrointestinal basidiobolomycosis (GIB) were from the southwestern USA as well as subtopics regions of Asia [1]. There are currently less than 80 reported cases of GIB; of these, 23 occurred in the USA (Arizona), 23 in Saudi Arabia, and 17 in Iran. It is believed that a warm and humid climate enhances the growth of *B. ranarum* in these environments. Many case reports in Saudi Arabia are from the Tohama area, Asir province, in the southern region of the Kingdom [2]. Most cases (88%) occur among male patients younger than 20 years old [2]. Of 71 cases, only 6 were in women [3].

The diagnosis of GIB tends to be overlooked in most patients since the presentation of the disease can mimic colon carcinoma in adults or inflammatory bowel diseases and tuberculosis in both children and adults [4].

A conclusive diagnosis of GIB demands isolation of *B. ranarum* in culture. As most patients are identified postoperatively, many patients never underwent a microbiological examination. Consequently, the diagnosis is frequently established based on distinctive findings in a histopathological examination. The Splendore–Hoeppli phenomenon, consisting of fungal hyphae surrounded by star-like, deeply eosinophilic amorphous substance, is a characteristic feature of GIB [5]. Though not pathognomonic, these features aid in establishing the diagnosis of GIB, particularly in the presence of the classical symptoms and epidemiologic setting [6]. In addition to tissue eosinophilia, leukocytosis with peripheral eosinophilia that regresses dramatically following surgery supports a diagnosis of GIB. A recent review by Vikram et al. observed peripheral eosinophilia in 76% of cases.[1] The persistence of peripheral eosinophilia may indicate an on-going source of infection [1].

Different antifungal drugs are used to treat basidiobolomycosis. Itraconazole is the most commonly used azole and has shown promising results [7]; both voriconazole and posaconazole have also been used successfully [8]. Conversely, results with amphotericin B and related compounds were unsatisfactory, with 50% of *B. ranarum* isolates being resistant to amphotericin [9]. Whereas some patients with GIB have been managed medically only, most have undergone surgery combined with prolonged medical treatment.

2. Case

A 58-year-old man was admitted to the hospital in May 2014 with a 1-month history of left iliac fossa (LIF) pain, worsening constipation, bleeding of the rectum, and weight loss. The abdominal pain had recently worsened, and he had developed a fever. The patient had

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bronchial asthma and chronic hepatitis B. He denied the use of herbals or antacids but admitted to using a homemade enema with a hose and tap water.

Examination showed a pulse rate of 93/min, blood pressure of 122/61 mmHg, respiratory rate of 30/min, temperature of 38.3 °C, and oxygen saturation of 98%. The abdomen was diffusely tender, with a tender mass in the LIF and external haemorrhoids. The results of the examination were otherwise normal apart from quadriplegia and a scar from spinal surgery following a traffic accident 20 years previously.

The white blood cell, eosinophil, and platelet counts were 5.7×10^9/L, 0.8×10^9/L, and 703×10^9/L, respectively; the haemoglobin concentration was 7.3 g/dL. The C-reactive protein, creatinine, and albumin concentrations were 141 mg/L, 43 μmol/L, and 25 g/L, respectively. HbA1c was 5.8%, liver enzymes were normal, and he tested negative for HIV.

An initial computed tomography (CT) scan of the abdomen at another hospital had displayed a mass and perforated descending colon with pericolic fluid collection, suggesting colon carcinoma. Emergency exploration on **day 0** identified a mass in the sigmoid and descending colon extending to the splenic flexure. The mass was dissected, followed by the Hartmann procedure to create a diversion colostomy. On gross pathology, a hard mesenteric mass measuring 11.0 × 5.0 cm with a yellowish necrotic cut surface encasing the bowel segment was identified. Histopathology showed severe chronic granulomatous inflammation with necrosis **(**Fig. 1**A)** and extensive fungal elements with marked necrosis, consistent with mucormycosis (Fig. 1**B**). In addition, multiple foci of angioinvasion (Fig. 1**C)** were noted.

The patient was started on liposomal amphotericin 5 mg/kg/day. A CT scan for abdominal pain on **day 16** showed segmental perfusion defects of the liver and segmental wall thickening of the colon (Fig. 2**A**). A subsequent colonoscopy revealed a circumferential mass at the hepatic flexure with a tiny polyp in the caecum (Fig. 2**B**). A biopsy of the mass showed necrosis and ulcerations but was negative for malignancy and mucormycosis.

After one month of amphotericin treatment, the patient was discharged home on posaconazole suspension 400 mg twice daily.

A follow up abdominal CT scan on **day 90** showed 2 hypodense hepatic lesions, measuring 2.1 × 1.9 cm and 4.0 × 4.0 mm, and a suspected filling defect in the portal vein. The patient was not compliant to treatment. He was advised to continue on posaconazole and attend the outpatient clinic.

On **day 180**, the liver lesion worsened, measuring 5.7 × 4.4 × 8 cm with central hypodensity, suggesting a fungal abscess (Fig. 3). Gross segmental biliary tract dilatation was noted. Furthermore, increases in the eosinophil count (3.3×10^9/L) and alkaline phosphatase levels (1360 IU/L) were seen.

Despite drainage of the abscess to decompress the biliary tract, the liver lesions continued to worsen. Thus, left lobe hepatectomy was performed on day 270. Histopathology reviewed at our centre and the Mayo Clinic confirmed necrotizing granulomatous inflammation with eosinophilia and sparsely septate, thin-walled, irregularly branching fungal hyphae surrounded by eosinophilic deposition (Splendore–Hoeppli phenomenon) (Fig. 4**A, B**).

A review of the initial colon biopsy showed similar changes. The

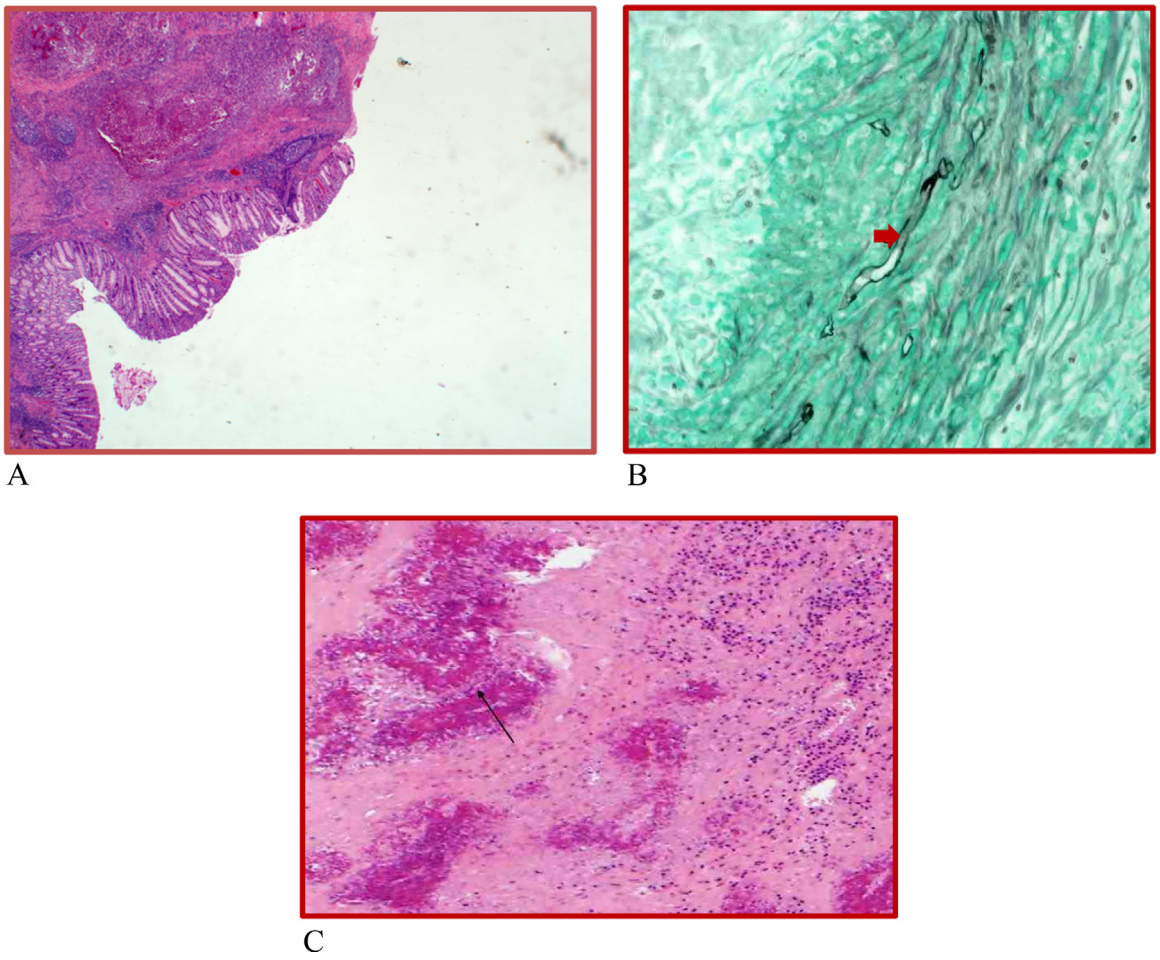

Fig. 1. A. Colonic resection showed severe chronic granulomatous inflammation with necrosis. H&E x 200. **B**. Colon tissue; Fungal hyphae (red arrow) with morphological features most consistent with basidiobolomycosis (thin -walled, branching, sparsely septated and surrounded by eosinophilic deposition/splendore-Hoeppli phenomenon); GMS x 600. **C**. Colon tissue. Angioinvasion (arrow); H&E stain; Magnification 100x.