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Unusual presentation of Gastrointestinal Basidiobolomycosis in a 7-year-old Child – Case Report

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Abstract Gastrointestinal basidiobolomycosis (GIB) is a rare fungal infection caused by basidiobolus ranarum. It has been recognized increasingly in the southern region of Saudi Arabia. Here, we report an unusual case of pediatric GIB with anorectal involvement in a 7-year-old Yemeni boy living in the south of Saudi Arabia. He had abdominal pain, abdominal distension, rectal bleeding, weight loss, perianal swelling and redness, and peripheral eosinophilia. The abdominal computed tomographic (CT) scan revealed significant wall thickness of the rectum. Histopathological findings of full thickness biopsy were consistent with basidiobolomycosis. The patient was treated with voriconazole and showed marked improvement. This case highlights the importance of considering GIB in children presenting with bleeding per rectum and eosinophilia associated with anorectal mass.

Keywords: *basidiobolomycosis, gastrointestinal, basidiobolus ranarum, saudi arabia, voriconazole, splender-hoeppli phenomenon*

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1. Introduction

Basidiobolomycosis is a rare but an emerging fungal infection caused by Basidiobolus ranarum [1,2]. Basidiobolus ranarum is a member of the order Entomophthorales in the class of Zygomycetes [3]. It is usually a subcutaneous infection, but rarely gastrointestinal involvement was described [4]. Most of the cases reported were from tropical and subtropical countries [1]. Pediatric gastrointestinal basidiobolomycosis (GIB) has been recognized increasingly in the southern region of Saudi Arabia [5,6,7]. Right-sided colon and liver were the most commonly affected sites with a rare description of anorectal involvement in pediatric GIB [8]. Here, we report a case of pediatric GIB affecting mainly the anorectum.

2. Case Report

A 7-year-old Yemeni boy, living in Jazan, s-western province, Saudi Arabia, was admitted to King Fahd Central Hospital (KFCH) with a history of colicky generalized abdominal pain associated with distension of two months duration. Three weeks before admission, the child developed bright red bleeding per rectum, mucoid stool with tenesmus and painful defecation. No fever, diarrhea, constipation, vomiting or jaundice. He had a history of poor appetite and weight loss of about 2 Kg over the last two months. No oral ulcers, joint pain or swelling. There was a history of contact with animals and ingestion of raw vegetables.

The evaluation revealed a sick looking child, his weight on the 25th percentile, his Height on the 50th percentile, his temperature was 37.8°C, with no significant lymphadenopathy. The abdomen was distended and tender with no palpable mass or hepatosplenomegaly. Anal opening distorted with tender perianal swelling and redness. Rectal digital examination showed multiple small tender masses involving the posterior wall of the rectum.

Laboratory studies revealed high erythrocyte sedimentation rate (ESR) 100 mm/h (normal <11 mm/h) and C-reactive protein (CRP) 18.2 mg/dL (normal <0.5 mg/dL). Complete Blood Count (CBC) was remarkable for an increase in eosinophils (15.9%) with a total white cell count (WBC) of 10.03 × 10^9/L, hemoglobin (Hb) 14 g/dL and platelet (Plt) 480 × 10^9/L. Coagulation, liver and renal profiles were normal. Further investigations showed negative anti-*Saccharomyces cerevisiae* antibodies (ASCA) and peri-nucleolar anti-neutrophil cytoplasmic antibody (pANCA). Quantitative serum immunoglobulins were normal. HIV screening, cytomegalovirus (CMV) IgM and VDRL blood test for syphilis all were negative. Stool culture, stool for ova, cyst and parasites and rectal

swab for *N. gonorrhea* were negative. Contrast-enhanced abdominal computed tomography (CT) study demonstrated diffuse circumferential massive wall thickening of the rectum with enhanced mucosa (Figure 1). Surrounded soft tissue edema and fat stranding with multiple enhanced and enlarged retroperitoneal lymph nodes were noted. The urinary bladder showed thickening and irregularity of the posterior wall with loss of fat plane between the bladder wall and rectal mass suggestive of posterior bladder wall infiltration.

The child underwent colonoscopy which showed inflammation of the distal part of the rectum suggestive of proctitis while the rest of colon was normal. Findings of Microscopic examination of rectal biopsy were suggestive of chronic active proctitis. Periodic acid-Schiff (PAS) and Grimelius methenamine silver (GMS) special stains were negative for fungus. We considered the possibility of Inflammatory bowel disease (IBD) initially and started on systemic steroid, topical mesalazine and oral metronidazole with no improvement over one month course of treatment. Subsequently, the child developed severe abdominal distension and tenderness with radiological features of intestinal obstruction of the lower gastrointestinal tract (GIT). The patient underwent laparotomy on an emergency basis which revealed an obstructive granulomatous lesion of the rectum. The surgeon has done Tru-cut full thickness biopsies and proximal colostomy. Histopathological examination of the biopsies showed transmural Granulomatous lesions with prominent eosinophilic infiltrates (Figure 2A). The hyphae structure of fungus seen in longitudinal and transverse cuts as empty tubes surrounded by an eosinophilic sheath ("Splendore-Hoeppli" phenomenon) were consistent with basidiobolus ranarum (Figure 2B&2C). GMS stain showed fungal hyphae (Figure 2D). Tissue culture using sabouraud agar was negative for basidiobolus ranarum. A fungal molecular diagnosis was not available in our hospital. The biopsy was negative both for acid-fast bacilli using Ziehl-Neelsen stain and polymerase chain reaction for Mycobacterium tuberculosis.

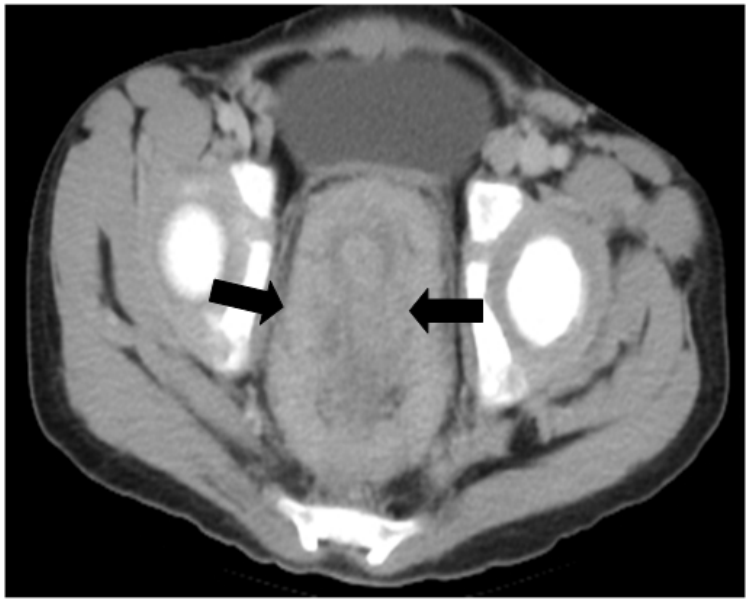

Figure 1. Axial enhanced CT of abdomen Shows diffuse circumferential massive wall thickening of the rectum with enhanced mucosa (Solid arrow)

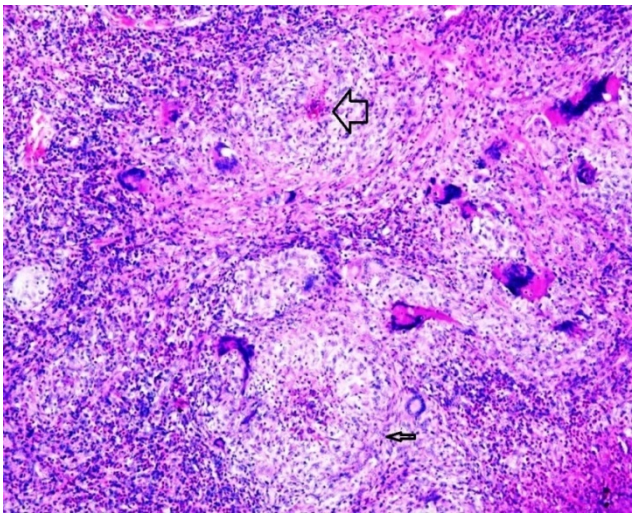

Figure 2A. Granulomatous reaction, narrow arrow showing granuloma and broad arrow showing fungus (H&E stain, × 100 original magnification). JPG

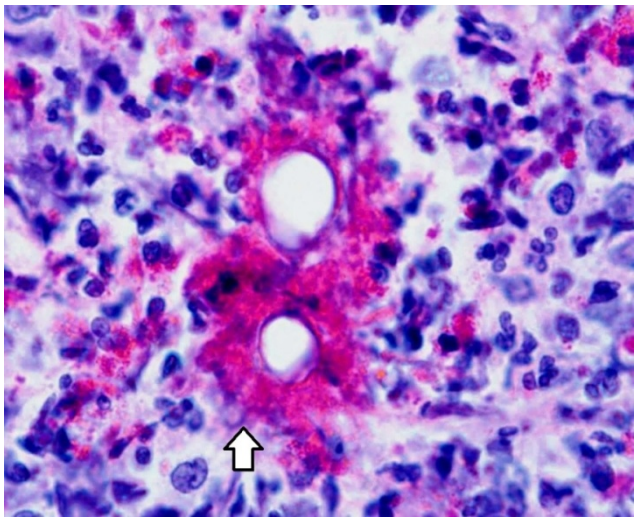

Figure 2B. Transverse section of fungal hyphae showing sun-burst pattern of splendore-hoeppli phenomenon (H&E stain, × 400 original magnification). JPG

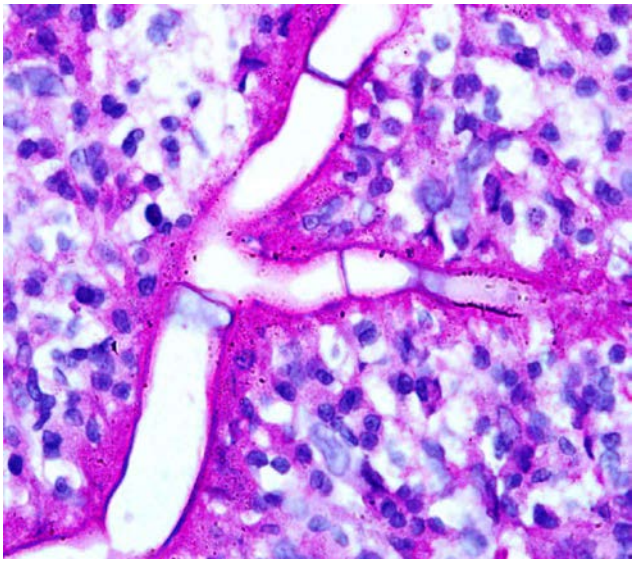

Figure 2C. Broad fungal hyphae longitudinal section showing splendore-hoeppli phenomenon (PAS stain, × 400 original magnification). JPG

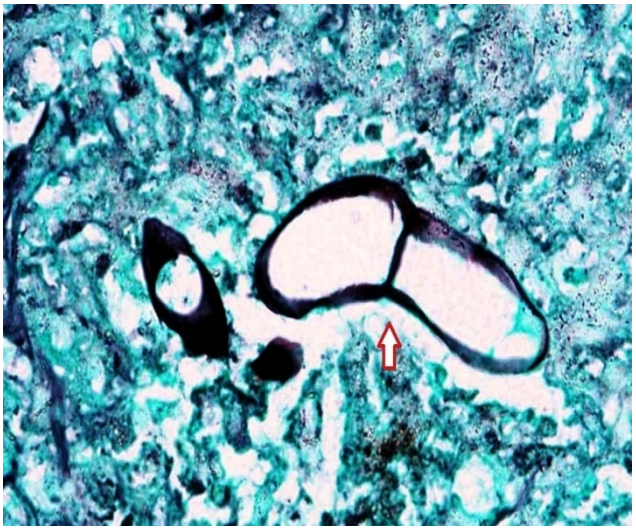

Figure 2D. Septated fungal hyphae (GMS stain, × 400 original magnification). JPG

The child was treated with intravenous voriconazole for one month and then discharged on oral voriconazole 10 mg per kg twice daily. One month after initiation of therapy, the patient showed marked improvement in his general condition. His weight increased by 3 kg; the gastrointestinal symptoms disappeared and the perianal signs resolved completely. We continued antifungal treatment for one year. He remained free of symptoms during follow-up with no drug side-effects. Repeated Contrast-enhanced CT abdomen revealed significant improvement with regression of the mural wall thickening of the rectum, the inflammatory process, and the associated lymphadenopathy.

3. Discussion

Basidiobolus ranarum is an environmental saprophyte commonly found in decaying plant materials, leaves of deciduous trees and soil [1]. It is occasionally present in the gastrointestinal tracts of amphibians, reptiles, fish, and mammals such as horses, dogs, bats, and humans [1,9]. Ingestion of food contaminated with soil or animal feces is the most likely route of GIB infection [2]. GIB is a rare fungal infection with the first case reported in 1964 from Nigeria [10]. Since the first report, there have been 73 cases worldwide [11]. Pediatric GIB is an emerging disease in Saudi Arabia with the first case reported in 2003 [12]. Since the first report till May 2017, there have been 29 pediatric GIB cases reported [5,6,7,8,13-18]. Adult GIB has been described as well in Saudi Arabia [11,19].

The patient lives in Jazan in the S-western part of Saudi Arabia. The warm and humid climate characterizes this area, possibly enhancing the growth of the fungus in this environment [5]. We believe that frequent contact with the animal environment and ingestion of raw vegetables may play a role in the route of infection. The involvement of the anal area may also raise the possibility of trauma to this region and direct inoculation of the fungus [1]. The most frequent presentations in reported cases were abdominal pain and fever. The other variable symptoms include vomiting, diarrhea, constipation, abdominal distension, hematochezia or jaundice [8]. A mass affecting part of the gastrointestinal tract was observed almost in all patients with GIB with the most commonly affected site is the right-sided colon followed by the hepatobiliary system [8,20]. Marked peripheral eosinophilia was a frequently reported laboratory finding [20]. In our report, the child had some of these features. Interestingly, there were perianal lesions in the form of distorted anal opening with perianal swelling and redness. The infection was localized mainly to the anorectum. To the best of our knowledge, there are only two reports of pediatric GIB described in the literature with a rectal and bladder involvement as well as perianal lesions similar to the case presented [21,22]. Recently there is one case report of pediatric GIB from Saudi Arabia with rectal mass but no perianal involvement [16].

Clinical presentation of GIB is nonspecific and could mimic malignancies, intestinal tuberculosis, and IBD. Misdiagnosis is highly possible in these cases [15]. The initial histopathological findings of colonoscopic biopsies were non-representative. It seems that basidiobolus ranarum involves the non-mucosal layers of the gastrointestinal tract (GIT). Therefore small superficial endoscopic biopsies usually show non-specific inflammation and might fail to detect the fungus [8]. In most of the previous reports including our report, the pathological study of the resected specimen or full thickness tissue biopsy confirmed the diagnosis of GIB.

Transmural granulomatous inflammation, prominent eosinophilic infiltration and fungal hyphae surrounded by an eosinophilic sheath (Splendore-Hoeppli phenomenon) are the characteristic histopathological features of GIB and the main clue for diagnosis [1,8]. In the case presented the histopathological finding was typical of GIB. Fungal culture was negative, and the diagnosis was entirely made based on the characteristic microscopic findings. In some reports, the molecular study by ribosomal DNA sequencing on formalin fixed paraffin embedded (FFPE) tissue established the diagnosis of basidiobolomycosis [13]. We do not have this test available in our hospital.

Combined surgical resection and prolonged use of antifungals are the best options for treatment of GIB [23]. Itraconazole successfully controlled the infection in many reports [5,12,16]. Earlier reports showed treatment failure with amphotericin B [20]. Recently voriconazole has been used effectively without surgical intervention for treatment of GIB [8,14]. Surgical intervention in this patient was limited to proximal colostomy for intestinal decompression because of rectal obstruction. We treated our patient successfully with voriconazole. His symptoms disappeared completely, and repeated CT abdomen showed significant improvement during follow-up.

In conclusion, Physician's awareness to the diagnosis of GIB is critical, and we should consider in the differential diagnosis of patients presented with gastrointestinal mass and eosinophilia. The spectrum of pediatric GIB is expanding to involve any part of the intestine including the anorectum. Further studies are needed to identify which patients are at risk for this rare disease.

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