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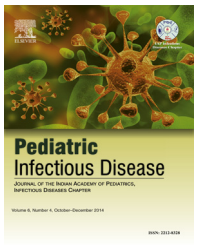

Case Report

Pediatric gastrointestinal basidiobolomycosis

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ABSTRACT

Basidiobolomycosis is a rare fungal disease caused by *Basidiobolus ranarum*, which is an environmental saprophyte. It is a chronic inflammatory disease that is generally restricted to the subcutaneous tissue and rarely involves the gastrointestinal tract. With the intent to spread awareness of this potentially life threatening and rare infection, we report a 4-year-old boy presenting with abdominal pain and fever with eventual diagnosis of gastrointestinal basidiobolomycosis. We discuss the nonspecific and confusing symptoms of this rare infection and available treatment options in detail.

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1. Introduction

Basidiobolomycosis ranarum is a rare known fungal organism, a member of the subphylum of Entomophthoramycotina.[1] Basidiobolus is an endemic infection in tropical and subtropical regions of the world, especially Africa, Latin America, and Asia.[2] The first human case of infection caused by *Basidiobolus ranarum* was one of subcutaneous mycosis, reported in 1956 in Indonesia, and other cases subsequently occurred in India, Africa, and South America.[3] Although the incidence of this infection is very low, recent studies have shown an emerging number of cases of visceral involvement due to *B. ranarum*.[4] In this article, we report a four-year-old boy with the eventual diagnosis of gastrointestinal basidiobolomycosis (GIB) from a southern province of Iran.

2. Case presentation

A 4-year-old boy from a southern province in Iran was referred to Bahrami Children hospital, a tertiary children's hospital, Tehran, Iran on December 2013 with chief complaint of abdominal pain and intermittent fever that first began approximately 45 days prior to his admission.

The very first symptoms of the patient began on September 2013, when he was admitted to a regional hospital because of abdominal pain. According to the documents of the mentioned hospital, in his physical examination, he was mildly dehydrated and ill. In his first abdominal examination, a generalized tenderness was detected. His vital signs were stable except for a temperature of 38 °C axillary. His past medical history was significant for a history of a unilateral undescend-

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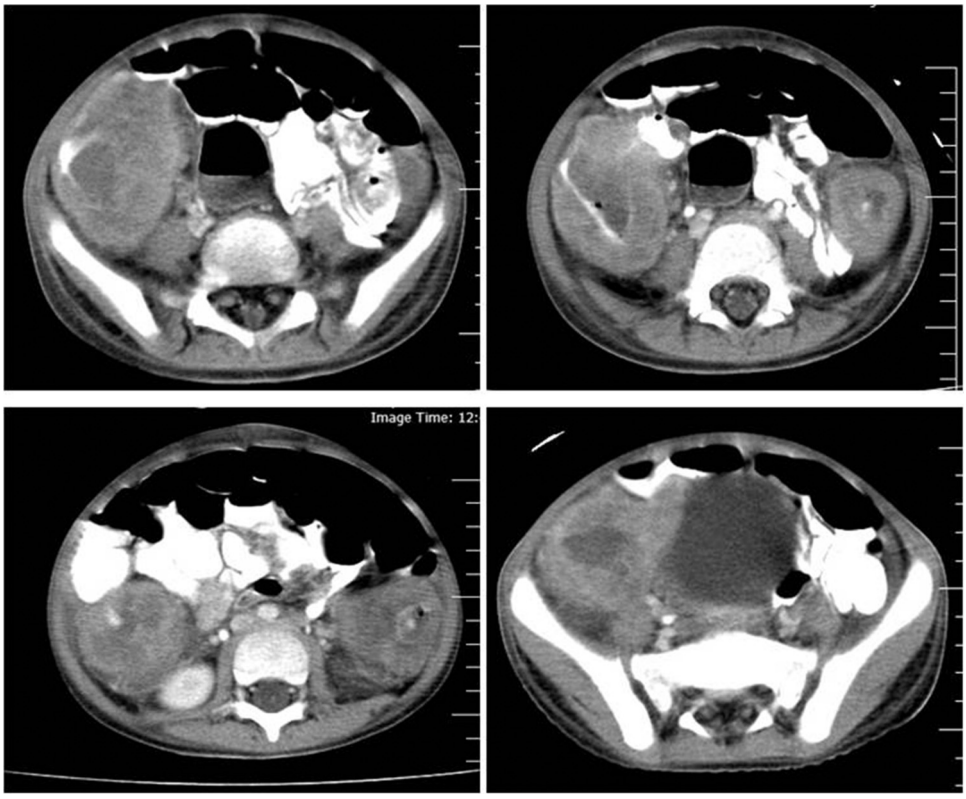

Fig. 1 – Abdominopelvic contrast-enhanced CT scan of our patient. Two extensive heterogeneous masses in the terminal ileum, which were extended to the cecum.

ed testis and cleft palate that was repaired surgically. He was otherwise a healthy boy.

He was diagnosed to suffer from cholelithiasis by ultrasonography. Unfortunately, he went through a laparoscopic surgery for cholecystectomy.

After the surgery, his abdominal pain and fever did not resolve. Intermittent bilious vomit was also added to his symptoms. The patient was referred to another hospital for further evaluation and management.

In the second hospital, during the physical exam, the patient appeared to be icteric and febrile. In his abdominal examination, a tender palpable mass in the right lower quadrant was discovered. Rectal examination was unremarkable. A repeated abdominal ultrasonography and abdominal computerized tomography (CT) scan were performed. Two extensive masses in the terminal ileum, which were extended to the cecum, were reported in the imaging process (Fig. 1).

The patient underwent a right hemicolectomy. During the surgery, three more masses were detected in the peritoneum. Histopathologic findings from the resected intestinal masses revealed extensive inflammation, granulomas with eosinophils and numerous fungal hyphae, and zygospores surrounded by eosinophilic material called the Splendore-Hoeppli phenomenon suggestive of basidioblomycosis infection. We unfortunately could not isolate the fungus from the intestinal specimen. Therefore, he was first treated with IV amphotericin B with 0.5 mg/kg stat and then 1 mg/kg/day, and when there was no significant response to treatment after about a week, it was changed to itraconazole capsules 10 mg/kg/day. After about ten days of unsuccessful treatment, the patient was referred to our hospital for further management.

On his admission in our hospital, the patient appeared ill and emaciated. In his first physical exam, his pulse rate, blood pressure, and temperature were normal. His abdominal examination revealed marked distention and a tender palpable mass in the left lower quadrant. An ileostomy, colostomy, and a bile drainage tube were noted in abdominal examination.

Results of laboratory investigations showed white cell count: 13,400/mm^3 (neutrophils 77% and lymphocyte 21%), hemoglobin: 11.5 mg/dl, platelet: 188,000/mm^3, ESR: 7, CRP: 57, AST: 67, ALT: 21, total bilirubin: 1.8 mg/dl, and direct bilirubin: 1.1 mg/dl. Electrolyte levels, blood urea nitrogen values, creatinine levels, and urinary analysis were normal.

Since his clinical symptoms persist and hematemesis and hematochezia were added to his symptoms, the antifungal treatment was changed to intravenous voriconazole 6 mg/kg/day BID for the first 24 h and then continued with 4 mg/kg BID. Because of his poor general condition, he was inoperable and no further surgical procedure could be performed. Unfortunately, though aggressive antifungal therapy was continued for the patient, his general condition deteriorated and he experienced multiorgan failure and expired on the 10th day of his admission in our hospital.

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3. Discussion

Basidiobolomycosis is caused by *B. ranarum*, a rare known fungal organism, and it is mainly seen in immunocompetent hosts. While the first presumed case of gastrointestinal basidiobolomycosis was reported in a 6-year-old Nigerian boy in 1964, the first culture-proven invasive case of basidiobolomycosis of the maxillary sinus was reported in 1978 in the United States.[4] The fungus is mainly found in soil, decaying organic matter, and gastrointestinal tract of amphibians, fish, and reptiles.[2,9] The exact mode of transmission of the infection is not well understood, but it seems that infected insects and animals like frogs and lizards can disseminate the fungus in the environment where humans can be infected.[4] There are some clues that GI basidiobolomycosis is acquired through ingestion of contaminated soil or food.[15]

Generally, basidiobolomycosis is distributed worldwide, while most of the cases were reported from tropical and subtropical regions.[4] Additionally, there have been some reports from countries in middle Asia like Saudi Arabia, and Iran.[6] Consequently, there is a probable suspicion that warm and arid climate, common in these countries, may be a contributing factor although the exact mechanism is not clear.[7] These facts were true for our patient, who was the resident of a southern province in our country with known arid weather. Since he was living in a rural area, he was with close contact with animals and their waste materials.

Basidiobolomycosis usually causes subcutaneous infection, which mainly involves the trunk and extremities; however, there have been emerging cases of visceral involvement in recent years that made this fungal infection a potent life-threatening one. The mode of transmission in the subcutaneous form of the disease is usually a trauma to the skin by a simple scratch or insect bite, which forms a nodule that usually becomes widespread to the adjacent tissue afterwards.[10] Involvement of maxillary sinuses, palate and turbinate, GI tract, and also lungs has been reported in recent clinical reports.

GI basidiobolomycosis is extremely rare in the world and only about 44 cases of immunocompetent children and adults with this disorder have been reported until now.[6] In India, there has not been any GI basidiobolomycosis case reports. Singh et al. reported a rare case of basidiobolomycosis, which involved maxillary sinuses in 2008.[1]

According to a recent review that was performed by Virkam and his colleagues in 2012, the most common sites of involvements in gastrointestinal basidiobolomycosis were the colons and rectum (82%), small intestines (36%), and liver and gallbladder (30%).[4] As colon was the main GI involvement in our mentioned case, it seems that the above fact is reliable.

The most common presenting symptoms of gastrointestinal basidiobolomycosis are abdominal pain, along with constipation, abdominal distention, and palpable mass, fever, and diarrhea.[5,10] The nonspecific features of the symptoms could procrastinate the diagnosis and so increase the risk of morbidity of the patients. The lab findings may reveal an elevated white blood cell count along with peripheral eosinophilia.[4] Histopathological characteristic finding is presence of granulomatous inflammation, tissue eosinophilia, and thin walled, broad, septate hyphae, surrounded by eosinophilic material (Splendore-Hoeppli phenomenon). Use of imaging techniques, especially computed tomography, may reveal the extension of the infection throughout the body.[4–8,11]

Our findings in history, physical examination, lab data, and biopsy were consistent with previously reported cases.

Since GI basidiobolomycosis is an aggressive disorder that can be potentially life threatening, management options are in great value of attention. Unfortunately, there are limited data about a definite treatment option for GIB, especially in children. But according to available experiences, it seems that surgical resection of masses and debridement of the involved tissue in combination with prolonged antifungal therapy can be effective.[12,13]

The choice and the duration of antifungal treatment is not well understood.[6] Among the azoles, fluconazole and ketoconazole have been shown to be effective on some cases of subcutaneous forms,[14] while itraconazole remains the best treatment for GIB, along with a surgical resection of the mass.[4] The preferred dose of itraconazole is 100 mg twice daily continuing for 4 months to 1 year.[6] Additionally, there are few reports of successful treatment of GI basidiobolomycosis with posaconazole.[7,16] Unfortunately, in our case, though intensive surgical and medical treatment with antifungal drugs were performed, the patient did not respond well. It seems that the aggressive course of GI basidiobolomycosis and delay in the course of diagnosis of the patient has lead to this failure.

4. Conclusion

Basidiobolomycosis is a very rare fungal infection in tropical and subtropical regions of the world. GI involvement of this infection usually has nonspecific symptoms and lab findings that can prolong the course of diagnosis and subsequently delay the initiation of the appropriate treatment. Since this fungal infection can be potentially lethal and delay in diagnosis can increase the risk of mortality and morbidity, physicians, especially pediatricians, working in regions where the disease is epidemiologically common should be aware of this fungal infection. Even in immunocompetent patients presenting with chronic GI symptoms and infiltrative lesions or eosinophilic granuloma in their GI tract, gastrointestinal basidiobolomycosis should be considered as an important diagnosis along with excluding of other differential diagnoses.

Conflicts of interest

The authors have none to declare.

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