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Case Report

Abdominal mucormycosis in a child: a case report

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Abstract

A 2-year-old Brazilian female child from the countryside in Bahia State presented with pain in the right flank of the abdomen, accompanied by a daily fever for about 2 weeks before admission. A large mass in the abdomen was resected by the surgical team. The biopsies revealed the mass was an intra-abdominal mucormycosis. However, the diagnosis was late, and despite treatment (amphotericin B) initiation, the patient eventually died.

Keywords: Mucormycosis. Pediatric abdominal pain. Amphotericin B.

INTRODUCTION

Mucormycosis is a rare fungal infection caused by fungi of the Zygomycetes class Mucorales and Entomophthorales order. It usually occurs in tropical area and presents as a subcutaneous infection. Mucormycosisis caused by opportunistic pathogens and rarely causes disease in immunocompetent patients, yielding mainly processes that cause neutropenia or neutrophil dysfunction. After aspergillosis and candidosis, it is the third most common invasive fungal infection, representing 8.3-13.0% of all fungal infections found in autopsies of hematologic patients[1,2].

Clinical manifestations are variable, including rhinocerebral commitment, often 44-49% of the reported cases; localized or generalized primary cutaneous involvement (10-19%); lung (10-11%); disseminated (6-11%); and gastrointestinal (2-11%)[3,4].

Gastrointestinal presentation of mucormycosis is uncommon and rarely diagnosed in living patients. In such cases, the diagnosis is late, and the mortality rate is high, at approximately 85%[3]. Only 25% of cases of gastrointestinal mucormycosis are diagnosed antemortem, and the disease is reported mainly in premature infants, newborns, patients, and malnourished children with oncological diseases, diabetes mellitus, or a history of corticosteroid use[5,6]. It can be acquired by ingesting pathogens in foods such as fermented porridge, alcoholic beverages and drinks derived from corn, herbs contaminated with spores, and homeopathic remedies[7].

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Mucormycosis rarely appears in children. In an analysis of all reports of pediatric mucormycosis cases published until 2004, 157 patients (64% male) with an average age of 5years were identified[8]. Twenty-eight (18%) patients had hematological diseases, and 9 (6%) had undergone transplantation. A series of an additional 30 pediatric cases from 2004 to 2008 was also reported in 2009[9].

We report an unusual gastrointestinal mucormycosis presentation in a previously healthy child, whose diagnosis was challenging to the entire team.

CASE REPORT

A 2-year-old Brazilian female child, weighing 12kg (z-score: 0), from the countryside (Ribeira do Pombal, Bahia State) presented with pain in the right flank of the abdomen, accompanied by daily fever for about 2 weeks before admission. She was attended by different doctors in her hometown; however, only symptomatic drugs for pain were prescribed, and the symptoms increased. The patient's condition worsened, and dysuria appeared, associated with inappetence, in the last week before admission.

On admission, she presented as malnourished, with pain and a palpable abdominal mass in the right flank region of the abdomen. An abdominal ultrasound showed decreased right kidney echogenicity, suggesting pyelonephritis. Hemoglobin was 8.9mg/dL, and white blood cell count was 17,600 cells/mm^3,with 4% immature neutrophils. Intravenous ceftriaxone (100mg/kg/day) was administered; however, the abdominal pain worsened. The large abdominal mass led to acute obstructive abdomen. The child underwent a laparotomy, during which a large abdominal mass was observed (**Figure 1**). The mass had adhered to the caecum, right kidney, and small bowel. It was resected, and biopsies were conducted. The mass

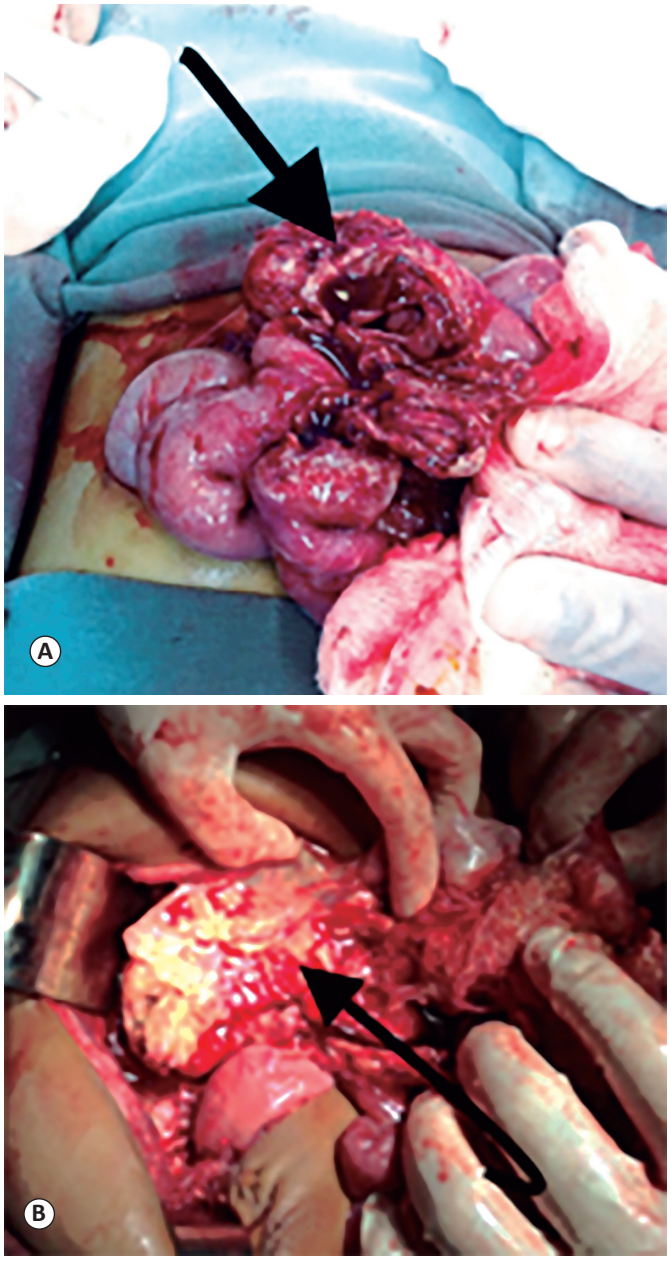

FIGURE 1. (A and B): Surgical finding of the abdominal mass (arrows).

had an irregular shape, measured 9cm at its greatest diameter, and weighed 185g.

We initiated empirical treatment for tuberculosis due to the severity of clinical symptomsand the suspicion of visceral disease (insidious daily fever, loss of weight and appetite) as well as broad-spectrum antibiotic therapy for sepsis. In the meantime, the anatomopathological analysis results suggested mucormycosis (**Figure 2**): frequent hyphae with thick walls, irregular branching, chambers, shoots forming on refractile walls, and heavy staining by silver salts. In addition to vascular wall invasion in the abdominal region, the bone marrow immunophenotyping was suggestive of myelodysplasia with no other remarkable findings. No other focus of mucormycosis was identified in other organs.

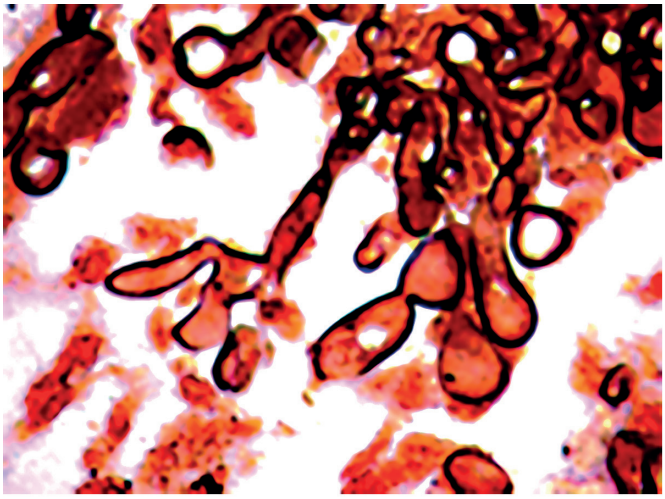

FIGURE 2. Pathological anatomy of the abdominal tumor revealing hyphae with thick walls, chambers, and bud formation.

Intravenous amphotericin B (1mg/kg/day) was started on hospital day 41. However, the patient already had multiple organ dysfunction at that time. Despite clinical support in the pediatric intensive care unit,her clinical condition deteriorated, and the patient died on day 54 after admission.

DISCUSSION

Mucormycosis is a rare and often life-threatening disease that is commonly seen in patients who have immunodeficiency syndromes such as malignancies,undergone solid organs transplant, received long-term steroid therapy, and poorly controlled diabetes. However, no underlying condition is discovered in 19% of cases. Of these cases, 9% have gastrointestinal manifestations of the disease[10]. In the reported case, the patient had no reported underlying comorbidities, which contributed to the delayed diagnosis. However, the bone marrow immunophenotyping revealed myelodysplasia, which could have been a sign of a previous underlying condition, such as a myelodysplastic syndrome, that would better explain the presence of mucormycosis in this child.

The most common presentation of abdominal mucormycosisis reported in adults, althoug husually in the abdominal wall instead of intraabdominally, which made the diagnosis in our patient more difficult.

Mucormycosis infections are difficult to diagnosis and treat due to the variable presentations, delayed diagnosis, and aggressiveness of the disease course. Clinical suspicion is an indicator for treatment, and physicians should not wait for the results of biopsy or culture. There is no reference for the sensitivity of the mycological examination, which is a new diagnostic tool, but the depth of tissue infection should be limited as a sensitive method given the difficulty of obtaining suitable material for the exam. The cultivation of the agent is achieved from the biopsy fragment seeding on Sabouraud dextrose agar at room temperature. The identification of the causative species is not always possible, estimated at 30% of positive cultures from fragments obtained from surgical specimens[11].

The therapeutic approach relies on an attempt to reverse or reduce the predisposing framework, facilitating surgical debridement and the immediate start of antifungal therapy.The most commonly used drug is liposomal or classical amphotericin B in high daily doses, which is 94% effective against *Mucor* spp, with a minimum inhibitory concentration<1ug/mL.The suggested dose is 1.0mg/kg/day for conventional amphotericin B (sodium deoxycholate) and 5-7.5mg/kg/day for liposomal amphotericin B. The antifungal efficacy of the combination of amphotericin B plus a triazole or caspofungin, for example, has yet to be demonstrated. The use of posaconazole, a second-generation triazole derivative, is considered a rescue option for patients refractory or intolerant to amphotericin B[12].

In conclusion, mucormycosis with anintra-abdominal presentation in pediatrics remains a disease that is poorly understood and with a high mortality, requiring a high degree of clinical suspicion for the diagnosis. Currently, the triad of clinical awareness, prompt initiation of treatment, and timely surgical intervention effectively controls the disease, avoids the main complications, and increases the chance of a better outcome.

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Conflict of Interest

The authors declare that there are no conflicts of interest.

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