Case Report

Colonic basidiobolomycosis in a child: report of a culture-proven case

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SUMMARY

Basidiobolomycosis is a rare fungal disease caused by Basidiobolus ranarum, which is endemic in tropical and subtropical regions of Africa, Asia, and Latin America. Gastrointestinal basidiobolomycosis poses diagnostic difficulties due to the non-specific clinical presentation and absence of predisposing factors. Eight pediatric cases of the disease have been reported recently from Iran, but none of these, or most of the other cases reported in the literature, have been proven by culture, which is the gold standard method for diagnosis. We present a case of culture-proven colonic basidiobolomycosis that occurred in a 3-year-old boy. The outcome was successful following surgical excision and antifungal therapy with posaconazole.

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1. Introduction

Basidiobolomycosis is a rare fungal disease caused by Basidiobolus ranarum, a fungus belonging to the Entomophthoraceae family of the class Zygomycetes. B. ranarum is an environmental saprophyte with worldwide distribution. The disease mainly presents as a chronic non-angioinvasive subcutaneous infection in immunocompetent individuals. Involvement of the gastrointestinal tract is a rare presentation, which in all reported cases has posed diagnostic difficulties. We report a case of culture-proven colonic basidiobolomycosis masquerading as malignancy in a 3-year-old boy.

2. Case report

A 3-year-old boy was referred with a 1-month history of colicky abdominal pain, nausea, and vomiting. He came from Zahedan, located in Sistan and Baluchestan Province, Iran. Physical examination revealed a large tender palpable mass in the right upper quadrant and epigastric areas. Laboratory investigations showed anemia (hemoglobin 9.2 g/dl), leukocytosis (12.5 × 10^9/l), eosinophilia (6%), and elevated serum IgE (2034 IU/ml, reference range <81). A computed tomography (CT) scan showed a mass in the descending colon accompanied by adhesion of the bowel loops to the anterior abdominal wall (Figure 1a). As it was suspected to be an infiltrative process in the colonic wall, the patient underwent a laparotomy. With a presumptive diagnosis of intestinal tumor, laparotomy and resection of part of the involved colon was done.

Histopathological examination showed extensive acute and chronic inflammation with foreign body granulomas and numerous eosinophils and neutrophils that involved the entire colonic wall (Figure 1b). There were also broad, thin-walled septated fungal hyphae that were surrounded by strongly eosinophilic material (Splendore–Hoeplli phenomenon) and stained positively with periodic acid-Schiff (PAS) and Gomori methenamine silver stains (Figure 1c). These features were suggestive of colonic basidiobolomycosis. Fungal culture was also consistent with B. ranarum (Figure 1d).

The patient was treated for 3 months with posaconazole 200 mg every 12 h. The patient's condition improved and the disease showed a dramatic response to antifungal treatment. The antifungal treatment was continued with oral itraconazole for 1 year, with follow-up by abdominal sonography without signs of relapse.

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3. Discussion

Gastrointestinal basidiobolomycosis (GIB) is a rare infection that, unlike other fungal infections, is mainly seen in immunocompetent hosts. As this fungus is an environmental saprophyte and is present in soil and decaying vegetables and fruits, the routes of infection appear to be ingestion of soil or feces or food contaminated by either of these. According to the literature, this infection is rare; however Geramizadeh et al. recently reported 14 cases from Shiraz, Iran. It appears that the small number of reported cases of GIB is probably related to the limited knowledge of this disease. Basidiobolomycosis has been reported mainly from tropical and subtropical areas of the world, as in our case. Most of the previously reported cases of pediatric GIB have been male and without any predisposing factors, as was our case. The main symptoms include fever, abdominal pain, diarrhea, weight loss, constipation, and an abdominal mass. Peripheral blood leukocytosis with marked eosinophilia were found in our case, as in other reported cases.

On physical examination, intra-abdominal masses were always detected and confirmed by imaging studies, including ultrasonography and CT scan, as well as during surgical exploration. Nearly all reported cases of GIB have shown involvement of the large intestine. In all cases, a provisional diagnosis of either an inflammatory bowel disease or a malignant process was suggested.

There are many factors that cause difficulties in reaching the diagnosis of GIB, such as the non-specific clinical presentation, absence of predisposing or risk factors in patients, non-representative colonoscopic biopsies due to involvement of non-mucosal layers of the gastrointestinal tract, and non-specific inflammation or granulomatous reaction, with possible overlooking of the fungal hyphae.

The unequivocal diagnosis requires culture of the organism, which can be isolated from surgically resected tissues. Sabouraud dextrose agar is a satisfactory medium, and visible growth is usually present 2–3 days after incubation at 25–30 °C. Colonies are white to gray with radial folds, and beaked zygosporae can be identified on microscopic examination of the culture material. Although culture is the gold standard for diagnosis, in all reported pediatric cases from Iran and other countries, fungal cultures were not done or were negative and the diagnosis was made on the basis of histological findings. Histological examination shows a granulomatous reaction, admixture of inflammatory cells rich in eosinophils, and the presence of radiating eosinophilic granular material (Splendore–Hoeppli phenomenon) surrounding broad branching, pleomorphic, and sparsely septated hyphae. Hyphal structures are silver- and PAS-positive. The absence of vascular invasion and a dense eosinophilic infiltrate in the lesional tissue are important findings in distinguishing this disease from mucormycosis, which usually shows a dense neutrophilic infiltrate and vascular invasion.
According to the available data, the prognosis of GIB in pediatric patients is usually favourable.\(^2\)\(^-\)\(^4\) A few cases of GIB with a fatal outcome due to disseminated infection have been reported, mostly in adults.\(^1\)

Surgical resection of all affected bowel segments and debridement of the involved tissue in combination with prolonged antifungal therapy appear to be effective.\(^1\)\(^,\)\(^3\)\(^,\)\(^4\) In a large study including 91 patients with mucormycosis, posaconazole was given for \(\geq 30\) days in 80% of the subjects. Amongst the enrolled patients, 13 had a complete response at 12 weeks and 42 had a partial response.\(^5\) Our patient responded well to 3 months of posaconazole treatment, and a CT scan performed after 4 months demonstrated complete disappearance of the mass. Treatment was continued with oral itraconazole for 1 year, and subsequent follow-up after 1 year revealed a totally asymptomatic image in our patient.

In conclusion, GIB is a rare and little-known fungal disease that has non-specific presenting features in healthy individuals with no identifiable risk factors. It can be misdiagnosed clinically as malignancy or inflammatory disease. This entity should be considered in the differential diagnosis of patients with abdominal mass and eosinophilia. Familiarity of both the clinician and pathologist with this condition, as well as using tissue culture, can lead to a definite diagnosis and prevent a fatal outcome.

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**References**