Mucormycosis-induced ileocecal perforation: A case report and review of literature

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ABSTRACT
Gastrointestinal mucormycosis is a rare form of invasive mucormycosis with high fatality rate due to difficulty in establishing its diagnosis. The classic risk-factors include immunosuppression and metabolic derangement. A case of ileocecal mucormycosis following intracardiac repair of congenital heart disease in a 17-year-old boy is described here who lacked the typical risk-factors for mucormycosis. Ileocecal mucormycosis affecting an individual without the classic risk-factors is uncommon.

KEY WORDS: Ileocecal, mucormycosis, steroid

Introduction

Mucormycosis is the third most common cause of invasive fungal infection¹ and affects individuals with classic risk-factors like immunosuppression, corticosteroid use, or metabolic derangement like diabetes and increased levels of available serum iron due to acidosis or administration of desferoxamine. Clinical presentation varies depending on the portal of entry of the fungus that includes rhino-orbital-cerebral, pulmonary, cutaneous, gastrointestinal, and disseminated forms. Gastrointestinal mucormycosis is rare, accounting for about 5–10% of all the cases of mucormycosis.²⁻⁴ It can affect any part of the alimentary system, but most commonly involves stomach followed by colon and ileum.

Here, we present a case of gastrointestinal mucormycosis manifesting as a large ileocecal perforation following a cardiac surgery, a short course of steroid therapy, and lacking the classic predisposing factors. Literature search for ileocecal or cecal mucormycosis yielded only eight cases [Table 1].⁵⁻¹²

Case Report

A 17-year-old boy diagnosed with a cyanotic congenital heart disease at 8 months of age was admitted for intracardiac repair. He had undergone two palliative surgeries at 8 months and 10 years of age and was also treated for cerebral abscess at 10 years of age. The present intracardiac repair of the congenital anomaly was uneventful. However, the immediate postoperative period was marked by intrapericardial bleed for which re-exploration was performed. Abnormal breathing pattern was noted on postoperative day (POD) 4. Upon evaluation, ischemic myelopathy was diagnosed and steroid therapy was started. On POD 25, he developed peritonitis due to a large ileocecal perforation warranting right hemicolectomy. Peritoneal fluid culture showed heavy growth of Enterobacter aerogenes and Klebsiella pneumoniae. Prior to the surgery, the absolute neutrophil count was 2400/µl.

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Gross examination of the right hemicolectomy specimen showed a large near-circumferential perforation at the ileocecal junction with exudate covered serosal and adventitial surfaces [Figure 1]. The perforated edges were congested and rest of the ileal and colonic mucosa were unremarkable. Microscopy from the ileal and the cecal ends of the perforation exhibited transmural necrosis with acute inflammatory exudate and vegetable matter along the serosa with the presence of numerous fungal hyphae transmurally. The hyphae were broad, irregular, and aseptate with right-angled branching suggestive of Mucorales. There was evidence of angioinvasion with many of the small and large caliber blood vessels completely occluded by fungal ball with thrombosis and vascular wall invasion [Figure 2]. The mycosis was also involving the appendix.

The patient was immediately started on antifungal therapy. On POD 32, he was re-operated for dehiscence of ileocolic anastomosis and the resected samples again showed mucormycosis. He developed hypotension and bradycardia on POD 36 and succumbed to it.

**Discussion**

Invasive mucormycosis is characterized by angioinvasion, thrombosis, and extensive tissue necrosis resulting in a fulminant clinical course. Rhino-orbito-cerebral mucormycosis is the most common form followed by pulmonary, cutaneous, disseminated, and gastrointestinal forms.[3]

There are two groups of individuals predisposed to acquiring gastrointestinal mucormycosis: premature or malnourished infants and children who manifest as necrotizing enterocolitis[2,13,14] and individuals with associated risk-factors like hematolymphoid malignancies, diabetes mellitus, and prolonged corticosteroid use.[13,14] In the alimentary tract, stomach is the most common site of involvement (67%) followed by the colon (21%), small intestine (4%), and esophagus (2%).[15]

About 25 cases of colonic mucormycosis are published in the literature, of which eight cases involved the ileocecal junction or the caecum [Table 1].[6,13] All the eight cases occurred in immunocompromised adults—primarily associated with

![Figure 1: (a) right hemicolectomy specimen with the exudate covered serosal surface and a large perforation at the ileocecal junction (arrow pointing to the cecal end and arrowhead to the ileal end of the perforation); (b) mucosal surface shows gangrenous change at the cecal (arrow) and ileal (arrowhead) ends of perforation.](image)

![Figure 2: (a and b) ileal edge of perforation with transmural necrosis and invasion by mucormycotic fungal hyphae; (c) broad and irregular hyphae with right-angled branch points. Angioinvasion of small (c, arrow) and large (d) caliber blood vessels is present. [(a) Hematoxylin and Eosin, (b–d) Grocott’s methenamine silver; Magnification = scale bar (a and b) 500 µm; (c) 100 µm, (d) 200 µm].](image)

**Table 1: Published literature on ileocecal or cecal mucormycosis**

| Study          | Age | Sex | Predisposing factor | Location                  | Type of lesion          | Procedure            | Treatment  | Outcome |
|----------------|-----|-----|---------------------|----------------------------|-------------------------|----------------------|------------|---------|
| Agha et al., 1985[5] | 21  | F   | Acute promyelocytic leukemia | Cecum                      | Ulcer and mass lesion   | Biopsy followed by resection | Amp B      | Survived |
| Elnakeby et al., 1996[6] | 54  | F   | Acute lymphoblastic leukemia | Cecum                      | Infarction and perforation | Right hemicolecotomy | Amp B      | Expired  |
| Mazza et al., 1999[7] | 35  | M   | Liver Transplant | Cecum + liver              | Cecal perforation        | Primary repair        | None       | Expired  |
| Karanth et al., 2005[8] | 56  | F   | Acute lymphoblastic leukemia | Ileocolic                  | Perforation              | Right hemicolecotomy | Lip Amp    | Expired  |
| Echo et al., 2005[9] | 43  | F   | Renal Transplant | Cecum                      | Mass lesion and perforation | Right hemicolecotomy | Lip Amp    | Survived |
| Lo and Law 2010[10] | 42  | F   | NK cell lymphoma | Cecum and Omentum          | Dilated and inflammed    | Autopsy              | Not available | Not available |
| Kim et al., 2011[11] | 35  | F   | Acute myeloid leukemia | Distal ileum and proximal ascending colon | Ischemia and perforation | Not available | Not available | Not available |
| Chawla N et al., 2012[12] | 65  | M   | Oral cannabis consumption and smoked marijuana | Distal ileum, caecum and ascending colon | Ischemia and mass lesion | Right hemicolecotomy | i.v antifungal | Survived |
| Present case          | 17  | M   | Nil | Ileocecal junction | Perforation              | Right hemicolecotomy | Amp B      | Expired  |

Amp B: Amphotericin B, i.v: Intravenous, Lip Amp: Liposomal amphotericin, NK: Natural killer
hematolymphoid malignancies and solid organ transplantation. Most often, the presentation was in the form of acute abdomen due to perforation. Mass lesion and ulcers were uncommon presentations of cecal mucormycosis. Among the seven cases for which data on the outcome was available, only three survived after intravenous Amphotericin B treatment.

What is unique about the present case is that gastrointestinal mucormycosis occurred in an immunocompetent adolescent resulting in ileocecal perforation. Over the last two decades, gastrointestinal mucormycosis has been increasingly reported in individuals without the classic risk-factors of immunosuppression or uncontrolled diabetes mellitus. These so-called healthcare-associated gastrointestinal mucormycosis occur in immunocompetent adults admitted to the intensive care unit and after prolonged hospitalization and major surgery.

In the present case, a temporary suppression of the immune system due to a combination of prolonged hospitalization and steroid therapy for ischemic myelitis could have predisposed to invasive mycosis.

Diagnosis of gastrointestinal mucormycosis during life is difficult. In a study on 129 cases of mucormycosis, none of the gastrointestinal cases were diagnosed during life as opposed to the rhinocerebral and cutaneous forms, most of which were detected ante-mortem (91% and 100%, respectively). The present case is again an exception as the diagnosis was made during the ante-mortem period even though the patient succumbed to the disease.

In conclusion, this case illustrates that gastrointestinal mucormycosis can also affect individuals without the classic predisposing factors of immunosuppression or diabetes mellitus. Prolonged hospitalization and major surgery seem to be contributing factors in immunocompetent individuals. Hence, under these circumstances, gastrointestinal mucormycosis should be considered in the differential diagnoses of nosocomial infections.

Declaration of patient consent
The authors certify that appropriate patient consent was obtained.

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Conflicts of interest
There are no conflicts of interest.

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