

# Mucormycosis of Intestine in an Immunocompetent Individual

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**Abstract-** Mucormycosis is a relatively uncommon opportunistic infection caused by fungus *Mucor*. This usually occur in association with diabetes, leukemia, lymphoma, following long term use of corticosteroids and cytotoxic drugs. We present a case of gastrointestinal mucormycosis in a 40 year old immunocompetent patient with a mass in the jejunum mimicking Gastrointestinal stromal tumor (GIST).

**Index Terms-** Intestine, Mucormycosis

## I. INTRODUCTION

Mucormycosis is a rare invasive fungal infection caused by *Mucorales* of class *Zygomycetes* that occurs in immunosuppressed individuals.<sup>1</sup> Rhinocerebral and pulmonary involvements of this infection are the most common followed by gastrointestinal (GI) infections. All parts of the intestine are vulnerable to infection, with stomach, ileum and colon most commonly involved. Primary GI mucormycosis is an uncommon disease associated with a high mortality rate, as they are usually diagnosed late.<sup>2</sup>

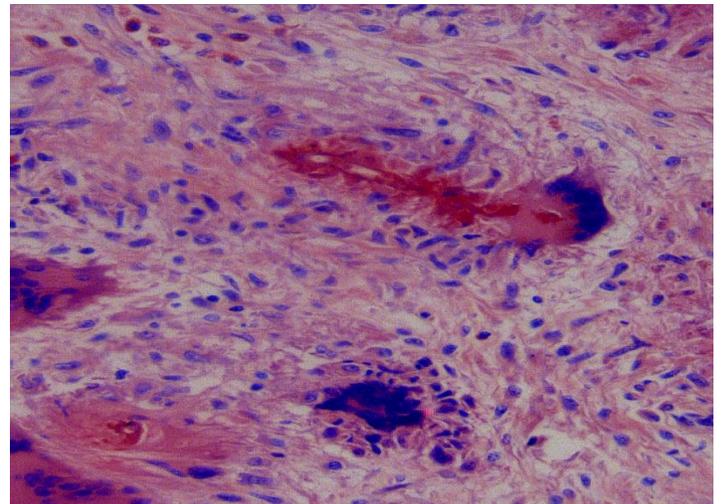
## II. CASE REPORT

A 40 year old male presented with abdominal pain and abdominal mass of 4 months duration. His routine investigations including blood sugar was normal. There was no history of any significant illness or drug intake in the past. On examination an ill defined mass measuring 3x 4 cm was present in the para umbilical region. USG abdomen showed a thickening in the wall of the jejunum forming a mass. Clinical diagnosis of gastrointestinal stromal tumor was made and the jejunum with the lesion was resected. On gross examination the segment of jejunum with mesentery measured 55 cm in length and weighed 591 grams. Jejunum appeared dilated and on cut section showed circumferential thickening of the bowel wall with ulcerated mucosa and black necrotic areas.



**Fig 1. Segment of intestine showing black necrotic areas in the wall**

On histopathological examination the entire thickness of the bowel wall showed granulomatous reaction with eosinophils and multinucleated foreign body giant cells. The granulomas and giant cells showed empty looking wide and non-septate fungal hyphae. The fungal stains PAS and Methenamine silver highlighted the thin walled broad aseptate fungal hyphae.



**Fig 2. Empty looking fungal hyphae surrounded by granulomas and giant cells H&E x200**



**Fig 3 Thin walled broad aseptate fungal hyphae  
.Methenamine silver x 400**

Thrombosed blood vessels with hyphal invasion was seen. The mesenteric lymph nodes also showed granulomatous reaction with eosinophils and fungal hyphae. A diagnosis of Gastrointestinal mucormycosis was made based on the above findings.

### III. DISCUSSION

Mucormycosis, also known as phycomycosis or zygomycosis was first reported by Paultauf in 1885. It is a rare invasive fungal infection, most often seen in immunosuppressed patients. Mucormycosis of the gastrointestinal tract is rare. It mainly occurs in infants or children, who are extremely malnourished. It is thought to arise from ingestion of the fungi.<sup>1</sup> Some patients with mucormycosis have no identifiable risk factors as in the present case.<sup>2</sup>

Mucorales and Entomophthorales, the two orders of the class zygomycetes, are closely related fungi with different manifestations. Mucorales are opportunistic fungi, that cause rapidly disseminating, often fatal infection in immunocompromised and diabetic patients. Fungi tend to invade blood vessels causing mycotic emboli. Entomophthorales are ubiquitous fungi usually causing chronic subcutaneous or nasofacial infections in immunocompetent individuals. The fungi has no tendency to invade blood vessels.<sup>3</sup>

All parts of the alimentary tract are vulnerable to gastrointestinal mucormycosis infection, with stomach (the most common site), ileum, and colon being commonly affected.<sup>4</sup> The route of infection is believed to be secondary to ingestion of the fungi, which then may colonize the gastrointestinal tract. Gastric mycoses have developed after nasogastric intubation or other conditions leading to the development of gastric or colonic ulcers, suggesting that an initial ulceration may facilitate fungal entry. The fungus invades the bowel wall and blood vessels, leading to bowel ischemia, necrosis, perforation, peritonitis or massive hemorrhage.<sup>5</sup>

The symptoms of gastrointestinal mucormycosis are varied and depend on the site affected. Nonspecific abdominal pain and distention associated with nausea and vomiting are the most common symptoms. Fever and hematochezia may also occur. The patient is often thought to have an intra-abdominal abscess. In premature neonates, mucormycosis may present as necrotizing enterocolitis. Gastric mucormycosis can be categorized into three forms: colonization, infiltration, and vascular invasion.<sup>2</sup> Sharma and associates reported isolated gastrointestinal mucormycosis in 8 patients, 2 of whom were middle-aged individuals without predisposing factors. The diagnosis may be made by biopsy of the suspected area during surgery or endoscopy.<sup>6</sup> The most frequent presentation is perforation, bleeding, or epigastric distention.

Mucoraceae are moulds in the environment that become hyphal forms in tissues. Once the spores (sporangiospores) begin to grow, fungal hyphae (angioinvasive forms) invade blood vessels, producing tissue infarction, necrosis, and thrombosis.<sup>2</sup> Neutrophils are the key host defense against these fungi, thus individuals with neutropenia or neutrophil dysfunction (diabetes, steroid use) are at highest risk. Two major hallmarks of histopathology are direct penetration and growth through the blood vessel wall, which explains the propensity for thrombosis and tissue necrosis with black eschar and discharge that are pathognomonic of this infection.<sup>4</sup>

Diagnosis of this condition can be elusive and is most often revealed by histopathology. There are no reliable, serologic, polymerase chain reaction (PCR) based or skin tests for zygomycosis. Therefore, the diagnosis is usually made by biopsy of infected tissues.<sup>1</sup> It is not possible to differentiate the various mucormycoses in tissue sections and isolation of Mucorales is difficult; material has to be cultured immediately.<sup>3</sup>

The biopsy demonstrates the characteristic wide, ribbon-like, aseptate hyphal elements that branch at right angles, as seen in the present case. A hallmark of zygomycosis infections is the virtually uniform presence of extensive angioinvasion with resultant vessel thrombosis and tissue necrosis. Tissue diagnosis of mucormycosis is essential to lower mortality, which can be higher than 50% in patients with hematological malignancies. Unfortunately, diagnosis is difficult and requires a biopsy of the organ(s) involved.<sup>5</sup> Aspergillus can sometimes produce a similar tissue reaction with multinucleated giant cells. It is readily distinguished by the presence of narrow hyphae with dichotomous branching at acute angles.

Antifungal therapy alone is typically inadequate to control mucormycosis, and surgery to debulk the fungal infection and/or resect all infected tissues is often required for effective cure.<sup>6</sup> Management of the underlying medical disease if any is also required.<sup>8</sup>

### IV. CONCLUSION

Increased awareness of this condition as a possible cause of intestinal ischemia and high index of suspicion is required to make an early diagnosis and management in these patients. It is important to initiate vigorous supportive medical management and antifungal agents after adequate surgical excision with clear margins.

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