SMALL INTESTINAL MUCORMYCOSIS: A CASE REPORT

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INTRODUCTION

Mucormycosis is a rare but serious fungal infection that rapidly attacks and usually kills its untreated victims. It is caused by the large, nonseptate, branching and saprophytic fungus of the genera mucorales.

Gastrointestinal Mucormycosis is rare with the gastroduodenal form the most commonly reported followed by that of the colon. Involvement of the small intestine is very rare with only two reports of the ileal disease found in the literature[1,2].

Antemortem diagnosis has been stressed as important for effective treatment and given the rarity of the small intestinal disease a high index of suspicion, based on sound knowledge of the macroscopic features of the diseased bowel, is mandatory at surgery to enable early diagnosis and effective treatment.

A case of small intestinal mucormycosis diagnosed postmortem in a thirteen-year-old boy is presented and the literature reviewed.

CASE REPORT

A thirteen-year-old boy was admitted to the general surgery department of the Korle-Bu Teaching Hospital on 26th July 2003 complaining of mild abdominal pain and vomiting for four days. The abdominal pain was constant and located in the lower abdomen. The pain became generalized and severe with associated distension, constipation and fever three days prior to presentation. These symptoms were preceded by a week's history of headache, palpitations, fever, chills and dry cough for which reason he was treated for malaria elsewhere. He appeared to have improved on this management but his condition deteriorated three days prior to presentation. He was neither a known diabetic nor a sickle cell anaemia patient and was not on any regular medication before the onset of the symptoms.

He was febrile on examination, with severe dehydration. He was neither anaemic nor jaundiced. Besides a tachypnoea of 30/min. the respiratory system was unremarkable. A regular pulse of good volume at 100/min and a blood pressure of 110/60mmHg were recorded. The abdomen was distended with generalized tenderness, rebound-tenderness and guarding. Bowel sounds were absent. The rectum was empty with smooth mucosa.

A diagnosis of peritonitis secondary to typhoid perforation was made and the patient prepared for surgery. His haemogram was; Haemoglobin-12.6g/dl, WBC-7.1 $\times$ 10^9/L (Neutrophils-63%, Lymphocytes-37%). The serum sodium was 143mmol/L, the potassium - 4.1mmol/L, urea - 18.1mmol/L, creatinine - 151umol/L, total protein-58g/L and albumin-29g/L. The HIV screening test was negative.

A Foley's urethral catheter was passed and the urine output monitored. He was resuscitated with fluids given intravenously (Normal saline and Ringers Lactate), antibiotics (Metronidazole and Ciprofloxacin), and analgesics (Pethedine). Laparotomy was done ten hours after adequate resuscitation.

A 1.5cm wide ileal perforation at 9cm from the ileocaecal junction and between the mesenteric and antimesenteric borders of the ileum was found. A few ileocaecal mesenteric lymph nodes were enlarged. The Peyer's patches were not inflamed. A segment of the bowel with the perforation was resected with 3cm margin and an end-to-end, double layer, inverting anastomosis done with chromic 2/0 suture.

He recovered from the surgery and was started on supplementary oral fluids on the fifth post operative day. There was a discharge of faecal matter from the wound on the 8th day and he developed peritonitis on the 10th day for which reason a relaparotomy was done the same day.

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The findings at operation were;

1. Two areas of segmental necrosis of the ileum. The distal segment was 10cm long and extended to the ileocaecal junction. The second was 10cm proximal to the distal necrosed segment and was 6cm long. The necrosis extended into the adjoining mesentery.

2. There were six other isolated perforations; five involving the ileum and one the jejunum. The largest was 6cm and the smallest 3cm wide.

A right hemicolectomy was done with ileotransverse anastomosis. The perforated segment of jejunum was resected and an end-to-end anastomosis performed.

A diagnosis of enteritis necroticans (Pig-bel) was suspected at this second laparotomy based on the necrotizing nature of the lesions. The patient died twenty-four hours after surgery from severe sepsis causing multiple organ dysfunction.

Trichuris trichura were found in the contents of the resected bowel. Cultures of ileocaecal exudates were not done. A microscopic examination of histological sections of the resected specimen stained with haematoxylin and eosin and accentuated with a Grocott stain revealed extensive areas of infarcted bowel and the adjoining mesentery as well as the large, thin-walled, nonseptate, branching fungal hyphae invading the bowel wall and showing a predeliction for vessels as shown in Figures 1,2 and 3. The diagnosis of mucormycosis was thus made postmortem.

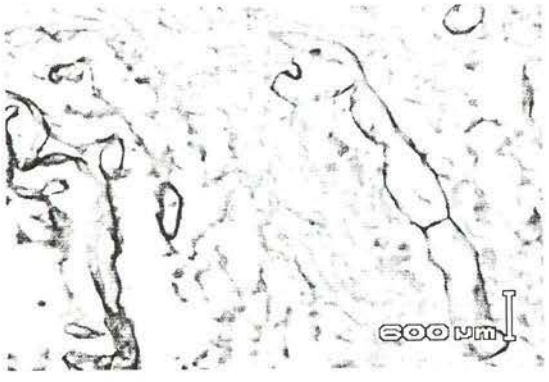

Figure 1 Haematoxylin and Eosin staining enhanced by Grocott stain of Histological specimen of the intestine showing the fungal hyphae in necrotic tissue.

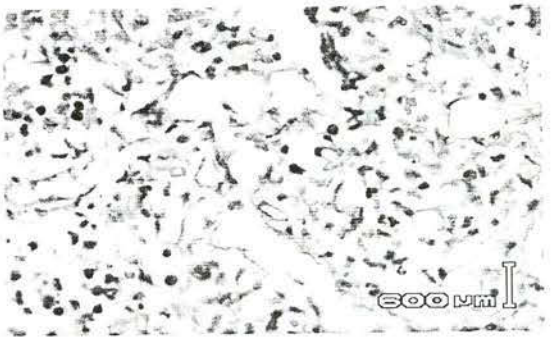

Figure 2 Haematoxylin and Eosin staining enhanced with Grocott stain of Histological specimen of the intestine showing the fungal hyphae in inflamed tissue

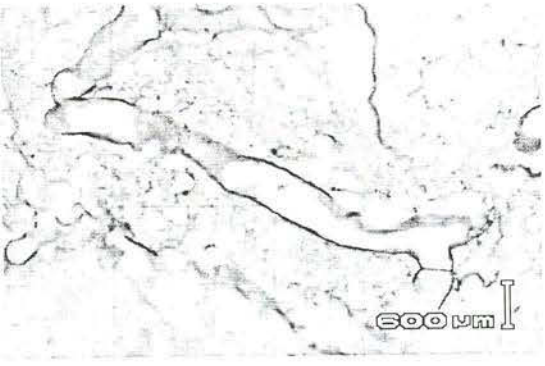

Figure 3 Haematoxylin and Eosin staining enhanced by Grocott stain of Histological specimen of the intestine showing the fungal hyphae in necrotic tissue.

DISCUSSION

Mucormycosis is a relatively uncommon, frequently fatal, opportunistic fungal infection of the genera *mucorales*. Species pathogenic to man in this genera include; *Rhizopus, Absidia, Mortierrella* and *Mucor*.

They are saprophytic and ubiquitous, thriving on dead and decaying organic matter including bread.

These fungi have little intrinsic pathogenecity in normal host but can cause serious infections in those immunocompromised by age, drug therapy, malnutrition or underlying disease. Not infrequently an underlying predisposition is not demonstrable in affected individuals, and fatal cases of invasive infection not associated with obvious systemic disease have been reported[3, 4].

Classical pathological forms of mucormycosis include rhinocerebral, pulmonary, skin and soft tissue, gastrointestinal and disseminated disease.[5] The gastrointestinal form is uncommon and most reports have come from Southern Africa[4]. It has

been reported sporadically in America, Europe and Asia[1,6,7].

The gastroduodenal disease, which frequently complicates chronic peptic ulcer, is the commonest followed by that of the colon[2]. Small intestinal disease is extremely rare with two reports of the ileal disease made in the literature[1,2].

Local abnormalities of the gastrointestinal tract have been emphasized as predisposing to the disease in the gut. Peptic ulcer disease, amoebic colitis, post-traumatic peritonitis, gastroenteritis, typhoid enteritis, pellagra and Kwashiorkor are the local gut abnormalities described[5,7]. The case being reported had *Trichuris trichura* in his gut and was malnourished (with a serum total protein of 59g/L and albumin of 29g/L) which could be the predisposing factors to the fulminant disease noted.

The pathogenesis of gastrointestinal mucormycosis is unclear, however, ingested spores can germinate and invade the mucosa through a chronic peptic ulcer or ulceration in the gastrointestinal mucosa in the course of debilitating illness. Gastrointestinal haemorrhage, obstruction or perforation (as was noted in the case presented) with dissemination of the infection may then occur.

While mucormycosis should be regarded as a serious and potentially life-threatening condition, it presents with a spectrum of severity and occasionally assumes an indolent and less aggressive course[8]. When gastrointestinal mucormycosis presents as invasive fungal infection the prognosis is extremely poor.

With the invasive disease the gross appearance of the lesion on the gut is characteristic and at laparotomy should aid diagnosis. The characteristics include:

1. The size of the lesion from the serosal aspect is much larger than would be expected in the usual ulcer from peptic, typhoid or amoebic ulceration

2. There is extreme hardness of the surrounding tissue.

3. There is a characteristic black appearance of the serosa.

4. There is a zone of hyperaemia between the black area and the normal bowel wall.

5. Seen from the inside, the obvious feature of the ulcer is the amount of black necrotic tissue as opposed to the white slough of peptic or typhoid ulcers.

If perforation occurs, as is frequently so in intestinal disease, these feature may be missed.

Diagnosis of gastrointestinal mucormycosis is difficult, but this is more so with the intestinal type. With improvement in diagnostic techniques antemortem diagnosis (an essential for effective treatment) is becoming possible[9]. Cultures of exudates and aspirates from affected bowel have persistently yielded no growth; growths have however; been achieved from cultures with tissue biopsy[8].

Histology of resection specimen occasioned by intestinal obstruction or perforation gives the diagnosis in most cases. Tissue invasion by the hyphae of mucormycosis must be seen microscopically to establish the diagnosis, but tissue culture is required to identify the fungal species involved.

A serological assay based on immunodiffusion for fungal antibodies holds promise for early diagnosis. Preliminary studies with this test indicate that it is specific for mucormycosis but cross-reaction occurred among the species in the genera. It demonstrated a sensitivity of 73% with a high negative predictive value[10].

Intravenous Amphotericin B is the mainstay of treatment in a daily dose of at least 0.8-1.0mg/kg. Surgical debridement (resection) to remove all infected material is a necessary adjunct as the presence of necrotic tissue prevents distribution of the drug to infected tissue.

Given the high mortality of the disease, attention should turn towards prevention since a source of infection could be removed if clustering of infection is recognized to come from a common source, e.g. contaminated ventilation system in hospital, or surgical dressing material[3,7,11].

The development of other preventive measures depends on detailed knowledge of the role of the factors predisposing to mucormycosis. In the gastrointestinal disease attention should focus on the unifying factor in the myriads of factors that predispose to fungal infiltration and invasion.

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