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Gastrointestinal Radiology

Gastrointestinal Mucormycosis

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Abstract. Mucormycosis is an acutely malignant fungal disease causing local thrombosis and infarction. Gastrointestinal infection most commonly occurs in diabetics or in the immunologically compromised. This case demonstrates the radiological appearances of the gastrointestinal lesion and prolonged survival with amphotericin-B therapy.

Key words: Gastrointestinal tract, inflammation – Mucormycosis, diagnosis.

Mucormycosis is a rare fungal infection most often occurring in diabetics or immunologically compromised subjects. Infection of the gastrointestinal tract, however, has been particularly reported from Africa and most commonly in otherwise healthy patients. The radiological appearances of invasive gastrointestinal mucormycosis have not been previously reported, and this is also the first reported case of survival beyond the immediate postoperative period.

Case Report

A 23-year-old farm worker employed in the apple orchards and stables of a fruit farm was admitted in November 1977 complaining of bilious vomiting, epigastric pain, loss of weight, and constipation for 3 weeks. There was no previous surgery or other illnesses except for an episode of Stevens-Johnson syndrome 9 months earlier for which no cause could be found. His father had died of tuberculosis but the rest of his family were well.

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On examination, he was thin, dehydrated, and feverish (38.4° C) with a gastric succussion splash and tenderness in the right upper abdomen. Plain radiographs and barium examination (Fig. 1) showed a distended stomach with a large cavitating lesion in the region of the pylorus and first part of the duodenum. It contained air and fluid with multiple irregular mounds on the inner surface of its wall. Barium trickled into it but would not pass beyond. At laparotomy this was confirmed as a sealed-off cavity containing dark fluid, covering a 7 cm defect in the wall of the distal stomach and proximal duodenum, and a 3 cm defect in the wall of the gallbladder. No other intra-abdominal pathology was found.

The cavity was closed around a duodenostomy tube and a gastrojejunostomy was performed. Multiple biopsies from the wall of the lesion showed dense subserosal fibrosis and extensive ulceration to the inner aspect of the muscularis externa. Inflammatory exudate was present on the serosa as well as on the ulcerated submucosa. Numbers of folded, nonseptate fungal hyphae with the appearances of the Mucoraceae were seen within the inflammatory exudate and also inside several small blood vessels (Fig. 2). The extensive subserosal fibrosis raised the possibility that the infection had been superimposed on a pre-existing chronic peptic ulcer.

Extensive investigation after postoperative recovery failed to reveal any other site of infection or any predisposing illness. This included nasal mucosal biopsy, isotope brain scan, bone marrow biopsy, glucose tolerance test, and individual immunoglobulin levels.

The patient was treated with amphotericin-B. Barium examination after 800 mg had been given showed significant improvement (Fig. 3). The duodenal bulb could now be identified and barium went easily beyond it without obstruction. The previous cavity could now be discerned only as an abnormal size and irregularity of the bulb with a fistula between it and the biliary tract. Due to the drug's side effects, the patient refused further treatment and left the hospital. After 3 months, repeat barium examination showed some deterioration in that the jejunum close to the stoma was narrowed (Fig. 4). Biopsy via gastroscopy confirmed mucormycosis in the region of the stoma with histological appearances similar to those of the original lesion. A further course of 1000 mg amphotericin-B along with IV mannitol, hydrocortisone, and promethazine was given, but he again left the hospital before the aimed for total treatment dose of 3 g could be achieved. At his latest attendance, however, 9 months after the operation, he remains clinically well and has gained weight.

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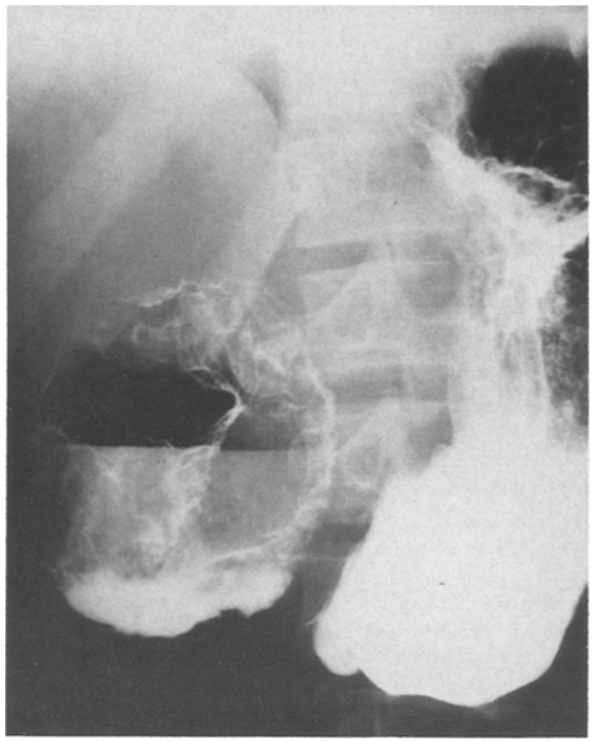

Fig. 1. Large cavitating lesion due to mucormycosis in pyloroduodenal and gallbladder region, causing gastric outlet obstruction. It contains gas and fluid and the inner surface of its wall is irregular. Barium trickled into it but not beyond

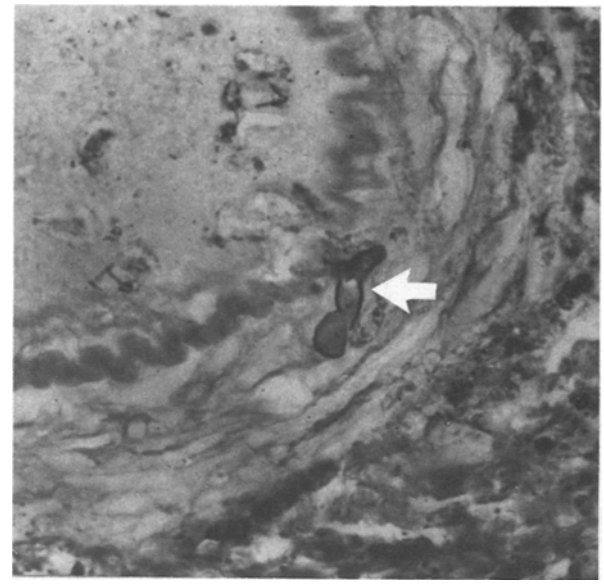

Fig. 2. High-power view of portion of a small vein in the stomach wall shows a nonseptate hypha (*arrow*) within the media close to the internal elastic lamina. Other hyphae are sectioned transversely within the occlusive luminal thrombus (Methenamine Silver stain, ×600)

Discussion

Mucormycosis refers to infection by fungi of the family Mucoraceae (order Mucorales, class Phycomycetes). Genera found responsible for human infection have been, in decreasing order of frequency, Rhizopus, Mucor, Absidia, Mortierella, Cunninghamella, and Saksenaea [1]. They are widely distributed in nature, in food, soil, and manure. They produce in humans a malignant infection by invading and thrombosing blood vessels causing local infarction, ulceration, and perforation [2]. This is responsible for its clinically acute and fulminant course which is unlike the more chronic course of other fungal infections [1]. The broad nonseptate hyphae, branching at almost right angles, are typical of the Mucoraceae, and finding such hyphae within thrombosed blood vessels is diagnostic of the infection [3]. They stain well with Methenamine Silver and with periodic acid–Schiff, and may be detected on routine hematoxylin and eosin stain [1]. The organisms can be difficult to culture (on Sabouraud's glucose) even when seen histologically [4]. Subtle morphological differences between the pathogenic genera can be discerned by the expert mycologist, but they all produce the same histopathological and clinical manifestations [1].

Most commonly affected are the nose and paranasal sinuses, from where infection spreads easily to orbit and brain. Less frequently affected are the lungs and gastrointestinal tract. Burns can be infected [5]. Widespread dissemination may occur. Poorly controlled diabetes is the underlying condition in most cases of rhinocerebral mucormycosis [2], whereas lymphoma, leukemia, or leukopenic states underlie most cases of pulmonary mucormycosis [1].

Gastrointestinal mucormycosis is unusual in that it has occurred most often in otherwise healthy people, although a significant minority have had malnutrition, diabetes, or debilitating diseases [6–9]. Many of these cases have been reported from among the black peoples of South Africa [3, 10–15]. This may be due to cultural dietary factors, particularly the many fermented foods and drinks which encourage fungal growth [16, 17], and this leads one to suspect that it probably occurs in other parts of sub-Saharan Africa too. Sites of infection in decreasing order of frequency have been stomach, colon, small bowel, and lower esophagus. Typically the presenting lesion is a large perforated ulcer with a black discoloration of the surrounding indurated tissues and of the ulcer slough [17]. The typical clinical presentation is therefore of perforation and peritonitis, sometimes with massive bleeding as well [14]. All previously reported

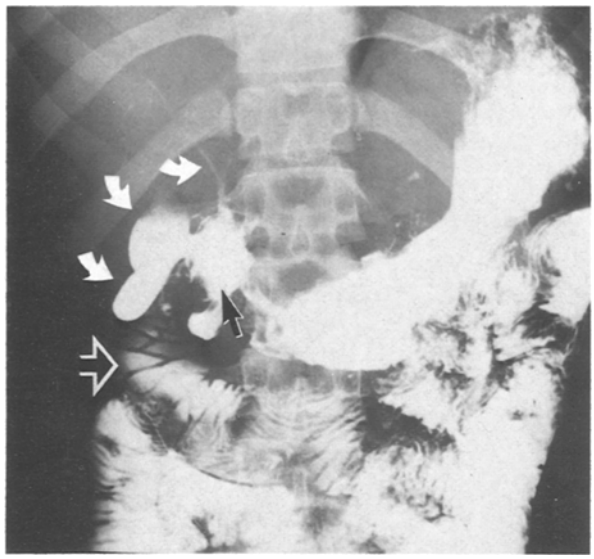

Fig. 3. Improved appearances after 800 mg amphotericin-B. Barium passes through the gastrojejunostomy and also via the duodenum. Large irregular duodenal bulb (*black arrow*) communicates with gallbladder and bile ducts (*curved white arrows*). Barium now passes beyond this region into second part of duodenum (*large open arrow*)

cases of gastrointestinal mucormycosis have been rapidly fatal due to continuing infection even after resective surgery or closure of the perforation.

Mucormycosis responds to amphotericin-B [1], but its toxic effects are frequent and serious [18]. Rhinocerebral infection has been cured by this drug [2, 19] and so, rarely, has pulmonary infection [20], but ours is the first published case of gastrointestinal mucormycosis surviving beyond the immediate postoperative period. It has also provided the first documented radiographic appearance of the lesion itself; a gastric lesser curve lesion reported from Indonesia in 1956 [21] was almost certainly a benign peptic ulcer with noninvasive secondary colonization, as none of the observed hyphae were within blood vessels and the clinical course was slow and benign.

Our initial radiological differential diagnosis for the massively excavating lesion in the pyloroduodenal region lay mainly between submucosal neoplasm (such as lymphoma and leiomyosarcoma) and local perforation of peptic ulcer. Mucormycosis was not considered. It should, however, become a member of diagnostic gamuts for excavating gastrointestinal lesions and for free perforation into the peritoneal cavity if the patient is an indigenous black African; and possibly as a rare consideration in other parts of the world if the patient has poorly controlled diabetes or a compromised immune system.

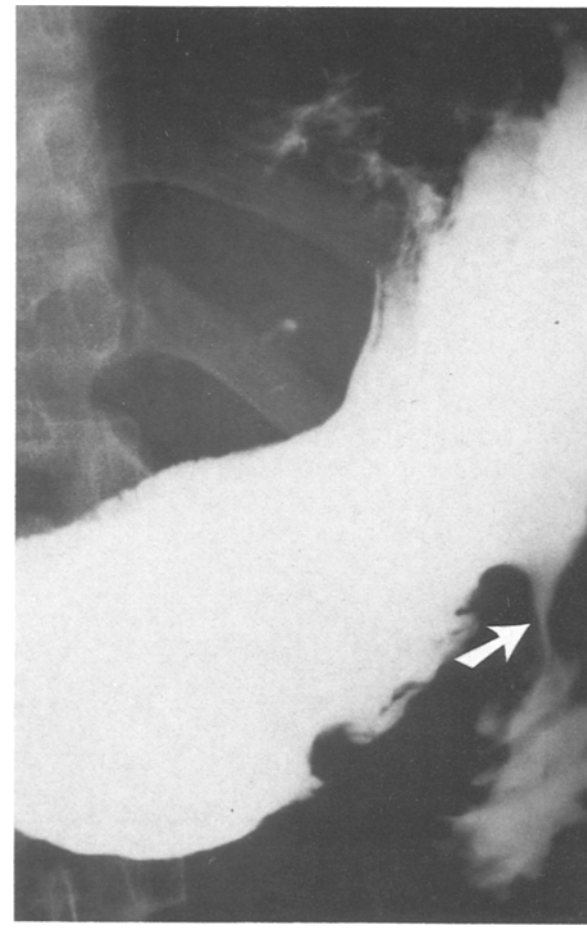

Fig. 4. Mucormycosis infection of the gastrojejunostomy 3 months after interrupting treatment. The afferent limb at the stoma is stiff and narrowed (*arrow*)

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