CORRESPONDENCE

Mucormycosis—A Possible Trigger Pathogen for Encapsulating Peritoneal Sclerosis

Editor:

A 54-year-old woman with diabetes mellitus and hypertension was started on emergency hemodialysis in November 2001. After stabilization, she was converted to peritoneal dialysis (PD) in December 2001. She had a severe episode of *Xanthomonas maltophilia* exit-site infection in March 2003, which prompted removal of her PD catheter. After another period on hemodialysis, she was restarted on PD in November 2003.

The patient had no other infections related to PD until March 2010, when she was admitted with an upper gastrointestinal bleed and peritonitis. Endoscopic examination of her upper gastrointestinal tract showed gastric erosions. Although no pathogen was identified from the PD effluent, her peritoneal white cell counts did not settle after 2 weeks on empirical ampicillin-sulbactam, ceftazidime, and fluconazole. Her PD catheter was removed in April 2010, and she recommenced hemodialysis. Her symptoms settled quickly, and no suggestion of bowel obstruction was noted at that time.

This patient was re-admitted to her local hospital in June 2010 with vomiting and abdominal pain suggestive of small-bowel obstruction. Computed tomography imaging of her abdomen confirmed the diagnosis of small-bowel obstruction. In addition, evidence of thickened peritoneum and loculated fluid collections was noted. Based on those findings, urgent laparotomy was performed on 22 June 2010, which revealed extensive adhesions, with pockets of fluid and pus. The bowel was noted to be covered with a cocoon-like structure suggestive of encapsulating peritoneal sclerosis (EPS).

The obstructed bowel was relieved with adhesiolysis, and a total of 1.5 L pus was drained during the course of the surgery. There was evidence of wide, ribbon-like aseptate hyphae branching at right angles, suggestive of mucormycosis infection. The pathology report

later confirmed mucormycosis in the fluid and pus specimen and the peritoneal tissues from the laparotomy (Figures 1–3).

Figure 1 — Hyphae are broad (3 – 20 μ m), rarely septate, and haphazardly branching (Grocott silver stain).

Figure 2 — A single filament, showing occasional septation (periodic acid Schiff stain).

Figure 3 — Granuloma, with palisading histiocytes and polymorphonuclear leukocytic infiltration (hematoxylin and eosin stain).

The patient was started on fluconazole after the confirmation of fungal growth. Her condition did not improve, and so she was transferred to the tertiary hospital on 2 July 2010 for further treatment. An alternative antifungal treatment—for example, amphotericin—was not considered because of liver dysfunction.

In the tertiary hospital, the patient was treated with intravenous fluconazole. After 5 days, the fluconazole was stopped because of improvement in septic markers and liver dysfunction. Unfortunately, the patient's clinical condition deteriorated, and she died 25 July 2010 from septicemia, presumably secondary to bacterial and fungal intra-abdominal collections.

DISCUSSION

Encapsulating peritoneal sclerosis is a rare cause of intestinal obstruction that is characterized by a thick grayish-white fibrotic membrane encasing the small bowel. Histologic examination of peritoneal tissue typically shows proliferation of fibroconnective tissue with nonspecific chronic inflammatory reaction. The diagnosis is usually based on intraoperative findings, but clinical suspicion may be raised by recurrent episodes of small-bowel obstruction, together with relevant imaging findings. In most reported cases, by the time the diagnosis of EPS is made, the condition has reached an advanced stage visible on imaging. In our case, this condition was obvious only upon surgery, indicating early-stage EPS.

Encapsulating peritoneal sclerosis has been associated with PD, liver cirrhosis, beta-blocker exposure, and

recurrent peritonitis (1). Paradoxically, discontinuation of PD has also been implicated in the development of EPS (2). Infective and inflammatory processes within the peritoneal cavity are likely to be important factors that trigger EPS. Chronic infection with tuberculosis has also been implicated in its development (3,4). As far as we know, no other organism has been directly implicated in the pathophysiology of EPS.

Mucormycosis is a term used for fungal infections caused by fungi from the order Mucorales (class Zygomycetes) (5). The infection frequently involves the sinuses, brain, or lungs as the primary area of infection, although cases involving the gastrointestinal tract, skin, and other organs are reported (6). Patients with hematologic diseases or diabetes mellitus and those receiving deferoxamine are especially susceptible (6). Not surprisingly, infections in renal patients, who are often immunocompromised and receiving deferoxamine, are not uncommonly reported (7). Sedlacek *et al.* (8) summarized the reports of PD-related mucormycosis peritonitis and reported a total of 15 published cases from 1980 to 2007. Only 1 of the 15 reported cases showed exposure to deferoxamine; 6 of 15 patients experienced an episode of bacterial peritonitis before contracting mucormycosis peritonitis. Interestingly, none of the reported cases showed evidence of EPS.

Mucormycosis peritonitis is usually diagnosed by PD effluent culture. In the published series of 15 cases of mucormycosis peritonitis summarized by Sedlacek *et al.*, the infection was identified by culture of the peritoneal effluent in nearly all cases. Only 1 case was diagnosed using peritoneal tissue (which was acquired during laparotomy after perforation of the colon).

The presence of mucormycosis in our patient's peritoneal tissue specimen suggests that the infection may have been present for a significant amount of time, possibly since March 2010, when the patient experienced a culture-negative peritonitis. It is possible that mucormycosis may have remained in a dormant or chronic form in the peritoneum as has often occurred in reported cases involving intracranial and maxillofacial sources (9). Chronic exposure to mucormycosis pyogens in the subsequent few months, together with underlying predisposing factors such as long duration on PD, prior bacterial infections, and discontinuation of PD, may have led to the rapid development of EPS. An alternative explanation might be that chronic mucormycosis caused adhesions and pus collections within the abdominal cavity, leading to a picture of sepsis and bowel obstruction. However, the incidental surgical discovery of abdominal cocoon, which is usually pathognomonic for EPS, may weaken that hypothesis.

CONCLUSIONS

Mucormycosis peritonitis may cause symptoms of EPS either by the true development of fibrotic encasement of the bowel, or by adhesions from chronic infective and inflammatory insults.

DISCLOSURES

The authors have no financial conflicts of interest to declare.

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