CASE OF THE MONTH

A patient with neutropenic fever and abdominal pain showing absent bowel wall on CT

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A 35-year-old Asian female with acute myeloblastic leukaemia developed fever and right lower abdominal pain 6 days after second induction chemotherapy. The white blood cell count was $10 \mu\text{l}^{-1}$ with 0% neutrophils. Peripheral blood culture was negative for aerobic and anaerobic organisms. Haematologists changed the antibiotics and started amphotericin B on the presumptive diagnosis of typhlitis. However, 13 days later, the patient again complained of severe abdominal pain. An urgent contrast enhanced abdominal CT was performed.

CT showed a long segmental wall thickening of the terminal ileum, caecum and ascending colon with target-like appearance. A relatively long segment of postero-medial wall of the ascending colon was not visualised, in association with a small amount of concentrated air bubbles close to the bowel wall, regional mesenteric haziness and a large amount of loculated fluid collected along the right psoas muscle (Figure 1-4).

What is the cause of these imaging findings?

Figure 1. Coronal reformatted contrast enhanced CT image shows abrupt discontinuity of the medial wall of proximal ascending colon (open arrow) and a long segmental bowel wall thickening of ascending colon (arrows).

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Diagnosis

Emergency laparotomy revealed diffuse necrosis of the distal ileum and proximal ascending colon and an 8 cm sized perforation in the posteromedial wall of the ascending colon, consistent with extensive ischaemic necrosis caused by mucormycosis (Figure 5).

Discussion

Mucormycosis, a fungal infection caused by the fungal order *Mucorales*, class *Zygomycetes*, is a relatively uncommon opportunistic infection, primarily seen in immunocompromised patients, such as those with diabetes mellitus, leukaemia or lymphoma. Most of these conditions are associated with impairment of normal neutrophil and macrophage function, resulting in increased risk for invasive fungal infections. Although aspergillosis and candidiasis are the most common invasive fungal infections in such patients, the incidence of infectious diseases caused by *Zygomycetes* has risen significantly over the past decade [1]. According to the literature, five clinical manifestations caused by mucormycosis have been described, including rhinocerebral, pulmonary, cutaneous, gastrointestinal and disseminated diseases. If the gastrointestinal tract, the rarest spectrum of mucormycosis, is involved, the stomach is the most common site, followed by colon and small bowel [2].

The most characteristic feature of mucormycosis is invasion of blood vessels, resulting in ischaemia, haemorrhagic infarction and tissue necrosis, irrespective of the involved organs [3]. In the gastrointestinal tract, the fungi invade through the bowel wall and blood vessels, leading to bowel ischaemia, perforation, peritonitis or massive gastrointestinal haemorrhage. Imaging findings of mucormycosis involving the gastrointestinal tract have been rarely reported in the radiological literature, but in the early stage of gastrointestinal mucormycosis, CT may show non-specific bowel wall thickening with or without target sign and decreased bowel wall enhancement, representing bowel ischaemia. When the involved bowel

Figure 2. Contrast enhanced CT image from the upper to lower level show a long segmental wall thickening of the terminal ileum, caecum and ascending colon with target-like appearance (arrow).

Figure 3. Contrast enhanced CT image from the upper to lower level show a long segmental wall thickening of the terminal ileum, caecum and ascending colon with target-like appearance (arrow). A relatively long segment of posteromedial wall of the ascending colon (open arrows) is not visualised, in association with a small amount of concentrated air bubbles close to the bowel wall, regional mesenteric haziness and a large amount of loculated fluid collected (F) along the right psoas muscle.

segment becomes necrotic, the bowel wall is thinned or even worse, disappears, as in our case. Direct visualisation of the discontinuity of the bowel wall is one of the most specific imaging findings of bowel perforation, but this finding is infrequently seen in most patients with gastrointestinal tract perforation, partly owing to the small size of the lesion [4]. However, our case showed an unusually long segmental absence of the bowel wall, which was caused by extensive haemorrhagic infarction and perforation caused by mucormycosis. In 2006, Song et al [5] reported a case of mucormycosis resulting in gastric

Figure 4. Contrast enhanced CT image from the upper to lower level show a long segmental wall thickening of the terminal ileum, caecum and ascending colon with target-like appearance (arrow). A relatively long segment of posteromedial wall of the ascending colon (open arrows) is not visualised, in association with a small amount of concentrated air bubbles close to the bowel wall, regional mesenteric haziness and a large amount of loculated fluid collected (F) along the right psoas muscle.

Figure 5. Microscopic examination revealed colonic vessel occlusion and transmural invasion by many fungal hyphae with right angle branching, consistent with mucormycosis (arrow) (haematoxylin and eosin stain, $\times 100$ magnification).

perforation in a patient with acute myelogenous leukaemia. In that case, CT showed a long segmental absence of posterior wall of the gastric fundus with a massive collection of haematoma, which is very similar to our case. We think that CT findings of a long segmental discontinuity of bowel wall along with a large amount of haematoma and localised peritonitis may be characteristic findings suggestive of extensive infiltration of angioinvasive fungus in the proper clinical setting, although definite diagnosis can be made only by histological examination of tissue.

Successful treatment consists of a combination of surgical removal of devitalised necrotic tissue along with long-term administration of intravenous antifungal therapy. Early diagnosis of angioinvasive fungal infection at the time when they are still localised is very important for prompt patient management and improved prognosis [3]. In disseminated cases, the prognosis remains

unfavourable, in spite of aggressive treatment. The mortality rate in patients with haematological malignancy who have mucormycosis is greater than 50% [6]. Therefore, timely suggestion of presumptive radiological diagnosis of angioinvasive fungal infection including mucormycosis can have important bearing on the prognosis.

Conclusion

Imaging findings of early gastrointestinal mucormycosis may be non-specific, but CT findings of a long segment of absent bowel wall along with a large amount of haematoma can be suggestive of angioinvasive fungus in immunocompromised patients with neutropenic fever and abdominal pain unresponsive to the antimicrobial treatment.

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