CASE REPORT

Isolated splenic mucormycosis in a case of aplastic anaemia

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SUMMARY

Mucormycosis, a rare opportunistic infection seen in immunocompromised hosts, is caused by fungi of Mucorales family. It may be confined to the organs, such as rhinocerebral and pulmonary mucormycosis, or may cause disseminated infection. A 14-year-old boy presented to our clinic with fever and left upper quadrant abdominal pain, and on evaluation was found to have pancytopenia, and imaging revealed ill-defined splenic collection with thrombus in the splenic vein. He was started on empirical intravenous antibiotics, followed by antifungals empirically as he did not show any improvement clinically. Eventually, splenectomy was done, which on histopathological examination revealed mucormycosis. The patient finally succumbed to his illness as he developed peritonitis and refractory shock. To date, only two cases of isolated splenic mucormycosis have been reported. Aggressive treatment is needed, which includes the use of antifungals (amphotericin B) and surgical debridement or resection of the involved tissues or organs.

BACKGROUND

Mucormycosis, a rare opportunistic infection seen in immunocompromised hosts, is caused by fungi of Mucorales family. Depending on the portal of entry, it may be confined to the organs, with rhinocerebral and pulmonary mucormycosis being the most common, or may cause disseminated infection.¹ However, isolated splenic mucormycosis has been very rarely reported so far.

CASE PRESENTATION

A 14-year-old boy was referred to the clinic with a history of fever and left upper abdominal pain, along with nausea and vomiting for 10 days. He had received 3 units of packed red cells and 6 units of random donor platelet transfusions during his hospital stay for 4 days, in view of severe anaemia and thrombocytopenia, before admission to our hospital. On examination, he had a subconjunctival haemorrhage in the right eye. Abdominal examination revealed left upper abdominal tenderness and guarding, whereas chest examination revealed bronchial breath sounds in the left infra-axillary and infrascapular areas, suggestive of consolidation.

INVESTIGATIONS

Investigations revealed haemoglobin of 5.8 gm/dL, total leucocyte count of $1.2 \times 10^9/L$ ³ with the absolute neutrophil count being $448/mm^3$, and platelet

count of $12\,000/mm^3$. Ultrasound of the abdomen revealed an ill-defined, space-occupying lesion in the spleen. The CT of the abdomen revealed completely infarcted spleen with marked perisplenic inflammatory stranding, which extended to the left perinephric and colonic fat with thickened perinephric fascia. The splenic vein at the hilum showed intraluminal hypoattenuating thrombus (figure 1). Blood culture grew *Enterobacter* and *Acinetobacter*. Bone marrow biopsy was done in view of pancytopenia, which revealed hypocellular marrow with reduced erythropoiesis and thrombopoiesis, suggestive of aplastic anaemia. Flow cytometry was done to rule out paroxysmal nocturnal haemoglobinuria and was negative for CD55/59.

TREATMENT

He was managed initially with intravenous cefepime and amikacin, along with supportive blood transfusions. However, the fever persisted and he was started on empirical intravenous amphotericin (1 mg/kg) on day 6 of admission. As splenic abscess was strongly suspected, he was taken up for surgery and splenectomy was done. Histopathology revealed marked necrosis, and staining with Grocott-Gomori's (or Gomori's) methenamine silver showed broad, aseptate hyphae branching at right angles

Figure 1 Axial contrast-enhanced CT image in portovenous phase showing non-enhancing spleen with marked perisplenic and perinephric inflammatory stranding (black arrow). The splenic vein (white arrow) is distended with hypoattenuating thrombus.

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Figure 2 The fungal hyphae are broad and aseptate with right-angle branching consistent with the morphology of mucormycosis. Gomori's methenamine silver stain. Original magnification $\times 40$.

with invasion into the vessel walls, suggesting angioinvasive mucormycosis (figure 2). He was continued on amphotericin B along with antibiotics.

OUTCOME AND FOLLOW-UP

Three days postoperatively, he developed signs of peritonitis, refractory hypotension and finally succumbed to his illness on day 21 of admission.

DISCUSSION

Mucormycosis is documented 5–10 times less common than other 'mold' infections like aspergillosis. These fungi are ubiquitous in nature and usually do not cause symptomatic disease in an immune-competent individual, although there are case reports.^{2–4} Severe neutropaenia, chronic high-dose corticosteroids, severe graft-versus-host disease, haematological malignancies, stem cell or organ transplantation, uncontrolled hyperglycaemia and/or diabetic ketoacidosis, and iron overload (associated with multiple transfusions of blood products for thalassaemia or leukaemia) are the common predisposing risk factors for the infection.⁵

Depending on the portal of entry, it causes rhinocerebral mucormycosis, the most common variety, followed by sinopulmonary, primary cutaneous, gastrointestinal and rarely disseminated infection. In the disseminated form, the lung is the most common organ involved, followed by brain, kidney, heart and

Learning points

- ▶ A high index of suspicion must be kept for mucormycosis in patients with predisposing risk factors.
- ▶ Although common in immunocompromised individuals, it can also rarely affect immunocompetent individuals.
- ▶ Aggressive and early surgical debridement of the involved tissue and antifungal therapy are the keystones for the successful management of the disease.

the spleen. Single-organ involvement is less commonly seen in patients with underlying haematological diseases as compared with other risk groups.⁵ To date, only two cases of isolated splenic mucormycosis have been reported.^{4,6}

The hallmark in the pathogenesis of mucormycosis infection is the vascular invasion and tissue infarction. The diagnosis is made through the demonstration of the organism, characterised by broad aseptate hyphae that branch at right angles, in the smear of aspirated fluid or biopsied sample of the involved organ. The mortality is very high and the survival rate is reported only up to 36%. Aggressive treatment is needed, which includes the use of antifungals (amphotericin B) and surgical debridement or resection of the involved tissues or organs.⁴

Contributors SKS treated the case and came up with the idea of writing the case report. PB prepared the manuscript. BR did the histopathological examination. MS reported the radiology.

Competing interests None declared.

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