Acute Fatty Liver of Pregnancy with Mucormycosis of Bowel: A Rare Case Report

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Abstract

Background: Mucormycosis is an invasive pathogenic fungal infection occurring mostly in immunocompromised patients, though rare cases have been described in immunocompetent patient. Pregnancy has been proposed to partially immunocompromised state and cases of mucormycosis occurring during pregnancy have been reported. Most cases involved rhinocerebral region and rarely involvement of gastrointestinal tract was reported.

Case Report: We herein report a case of a 32-year-old female who presented to outside hospital in third trimester of pregnancy with fever and jaundice for 2 weeks, lower segment caesarean section (LSCS) was done at 36 weeks in view of worsening liver functions and ongoing sepsis. Post LSCS patient became drowsy, had acute kidney injury, worsening coagulopathy and liver functions and was shifted to our hospital. USG abdomen suggestive of fatty liver with hemo-peritoneum. Clinical and lab investigations were suggestive of DIC. Empirical antibiotic and antifungal cover was given. In view of her worsening condition and hemo-peritoneum exploration laparotomy was done, blood and clots were removed. However, patient continued to bleed from surgical site, multiple blood transfusions were done. USG now showed intra-abdominal fluid collection, on exploration gangrenous bowel was found which was resected, jejunostomy with mucous fistula was done. Histopathology of the bowel segment revealed tissue and angio-invasive mucormycosis. Despite all efforts patient continued to deteriorate with increasing vasopressors and succumb to her illness.

Conclusion: Occurrence of mucormycosis with acute fatty liver of pregnancy is a very rare finding. Increased awareness of this condition as a possible cause of abdominal sepsis in critically ill patient may aid in early diagnosis and management.

Keywords: Fatty Liver; Pregnancy; Mucormycosis; Bowel

Introduction

Mucormycosis is an opportunistic invasive fungal infection occurring mostly in immunocompromised states like in transplant recipients, diabetes, HIV infection etc. Cases have also been described in immunocompetent patient and in pregnancy. We herein report a case of invasive mucormycosis in a 32-year-old pregnant female leading to bowel gangrene along with fatty liver of pregnancy.

Case Report

We report a case of a 32-year-old female G2P1 who presented to an outside hospital at 35 weeks of gestation with complaints jaundice for last 2 weeks, along with loss of appetite, nausea vomiting, recurrent hypoglycemia and increased drowsiness. She was initially managed conservatively and later underwent lower segment caesarean section at 36 weeks for same in view of worsening liver function and
ongoing sepsis. She was given 6 units of fresh frozen plasma and 2 units of packed red blood cells during surgery. However, post-surgery her drowsiness increased, urine output decreased along with worsening coagulopathy and liver functions and was shifted to our hospital. Her hemoglobin dropped from 11.5 mg/dl on day of surgery to 7.3 within 2 days. Her LFT post-surgery were bilirubin 14.5 mg/dl, OT 77, PT 85 U/L. On examination patient was drowsy, not obeying commands (E3M5V2), pupils normal reacting to light with bilateral plantar flexors. Edema and icterus was present. Clinical and lab investigations (DIC SCORE + TEG) was suggestive of disseminated intravascular coagulation (DIC). Serology for hepatotropic (Hep A, B, C, D, E) viruses were negative. Her cultures of blood and urine were both negative. D dimer was < 0.5, FDP was < 5, Fibrinogen 85, arterial ammonia 61. Her tests for dengue, malaria, chikungunya were all negative. Thyroid profile was normal. ANA was positive in titer of 1:40. Ultrasound examination revealed raised echo texture of liver with fatty liver. Kidney showed raised cortical echogenicity maintained cortico-medullary differentiation, right kidney measures 10.7 and left kidney 11.5 cms along with moderate ascites, empirical antibiotic and antifungal cover was given. Exploration laparotomy was done, blood and clots were removed. However, patient continued to bleed from surgical site, multiple blood transfusions were done. Repeat USG now showed intra-abdominal fluid collection, on exploration gangrenous bowel was found which was resected, jejunostomy with mucous fistula was done. Intraoperative findings were stiff liver, 800 cc blood clot, ooze from parietal wall and gall bladder bed, multiple ischemic patches involving jejunum and ileum. Post surgery patient become more drowsy. Microscopic examination of the gangrenous bowel segment revealed multiple aseptate fungal hyphae within the mucosa and muscularis. The fungal hyphae were also seen invading the vessel wall at several foci and occluding the lumen. Microscopy from the liver tissue showed ballooning degeneration of hepatocytes with microvesicular steatosis (Figure 1). So final diagnosis of Tissue and angioinvasive Mucormycosis with Acute fatty liver of pregnancy was rendered.

Figure 1: 1a-Showing changes of fatty liver of pregnancy, 1b-Gangrenous bowel gross, 1c-Microscopy, 1d-Aseptate fungal hyphae, 1e-PAS stain and 1f-GMS Stain.
Despite all efforts, patient continued to deteriorate with increasing vasopressors and succumbed to her illness within a week due to sudden cardiac arrest.

Discussion

Mucormycosis is an invasive fungal infection caused by mucorales [1]. These are opportunistic ubiquitous saprophytic organisms that are usually found in soil and decaying matter. Most cases are seen in immunocompromised patients and rarely in immunocompetent persons as seen in our case. Immunocompromised states like diabetic ketoacidosis, transplant recipients, steroid therapy, neutropenia, and malnutrition increase susceptibility to mucormycosis [2].

Clinical categories of mucormycosis include i) rhinocerebral, (ii) pulmonary, (iii) cutaneous, (iv) gastrointestinal, (v) disseminated, and (iv) miscellaneous. Mode of entry into the host will determine the predominant site of involvement. Inhalation leads to rhinocerebral and pulmonary mucormycosis, direct contact leads to cutaneous whereas ingestion of fungal spores leads to gastrointestinal mucormycosis [3].

Most common form of mucormycosis is rhino-orbito-cerebral (44 - 49%), followed by cutaneous (10 - 16%), pulmonary (10 - 11%), disseminated (6 - 11.6%) and gastrointestinal (2 - 11%) presentations [4,5].

Within gastrointestinal mucormycosis, stomach is the commonest site of involvement followed by colon and ileum [6]. Bowel involvement can occur in three forms: colonization, infiltration, and vascular invasion. The most common presentation is perforation, bleeding, or epigastric distention [7]. Gastrointestinal involvement is commonly seen in malnourished adults and in individuals with underlying gastrointestinal disease like Crohn’s disease or abnormalities such as kwashiorkor, colitis, typhoid, or pellagra [8].

The fungal hyphae have the ability to invade the blood vessels locally leading to decreased blood supply to tissue thereby causing ischemic necrosis of the area which further promotes fungal growth. Impairment of phagocytosis by use of steroid, elevated sugar and malnutrition, increased levels of available serum iron as a result of acidosis prevent elimination of fungus thereby promoting growth of fungal hyphae [9]. Usage of deferoxime predispose to mucor infection as supply of free iron to fungus is increased thereby promoting its growth [10].

Surgical debridement along with antifungal therapy are the main modalities of treatment. Mortality rate is higher than 50% with an incidence ranging from 62.5% in rhino-cerebral form to 100% in disseminated form [5]. Its high morbidity and mortality rates are because of rapid vascular invasions, with subsequent tissue necrosis and infarction.

Gastrointestinal Mucormycosis in Pregnancy

Pregnancy has been defined as a risk factor for mucormycosis by several authors. The first case of mucormycosis in pregnancy was reported by Lloyd., et al. in 1949 in a 26-year-old female presenting with intractable nausea and vomiting in the third trimester [11]. Several case reports have been published thereafter. Occurrence of mucormycosis in pregnancy could be postulated due to partially immunocompromised state in pregnancy. Also, increase free serum iron promotes fungal growth in pregnancy. Oral iron supplements given during pregnancy further increases free serum iron. Rhinocerebral mucormycosis is reported most commonly during pregnancy [12-14]. Rithvik., et al. have reported a case of gastric mucormycosis with perforation in a 29-year-old pregnant female who also had acute fatty liver of pregnancy (AFLP) [15]. We report second case report of similar association.

Symptoms of both AFLP and mucormycosis are nonspecific such as anorexia, nausea, vomiting, malaise, fatigue, headache and abdominal pain thereby delaying diagnosis. AFLP can cause maternal metabolic acidosis secondary to impaired clearance of serum lactate by damaged hepatocytes. Acidosis in turn increases serum free iron which acts as stimulant for growth of mucor.
Conclusion

Mucormycosis is a fulminant invasive disease with high mortality and morbidity. Cases could occur in pregnancy owing to increased availability of free iron and partial immunocompromised state. So, possibility of mucor should be thought in patient with abdominal sepsis for early diagnosis and prompt management.

Conflict of Interest

Nil.

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Bibliography


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