



PHOTO CLINIC

Hepatosplenic Mucor Balls

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A 10-year-old male with a history of B-cell acute lymphoblastic leukemia (B-ALL) in first complete remission presented with severe headache and pancytopenia 3 months after completing chemotherapy. Bone marrow aspiration and cerebrospinal fluid analysis confirmed systemic and leptomeningeal relapse. He received systemic induction with oral dexamethasone, mitoxantrone and vincristine and intrathecal chemotherapy with methotrexate, cytarabine and hydrocortisone. After 2-weeks of treatment, the patient developed neutropenic fever which persisted despite negative blood cultures. Abdominopelvic computed tomography (CT) scan showed several hypodense nodules in liver; in different sizes, the largest one in left liver lobe measuring 72×50 mm bulging the liver capsule with perihepatic inflammatory changes

(Figure 1A) and also multiple hypodense nodules in the spleen (Figure 1B). Biopsy of the liver lesion revealed aggregations of thick-walled, non-septate fungal hyphae with irregular branches in right angles (Figure 1C). A diagnosis of hepatosplenic mucormycosis was made and liposomal amphotericin was initiated. However, hepatosplenic lesions increased in number on follow up imaging and caspofungin was added. Despite receiving optimal treatment and appropriate antifungal therapies, the patient was succumbed to death due to disseminated mucormycosis.

Mucormycosis is being recognized as an increasingly important cause of morbidity and mortality in immunocompromised patients. In 2002, a consensus group of the European Organization for Research



Figure 1: A) Several hypodense nodules in the liver, in different sizes, the largest one in left liver lobe measuring 72×50 mm bulging the liver capsule with perihepatic inflammatory changes. B) Multiple hypodense nodules in the spleen. C) Aggregations of thick-walled, non-septate fungal hyphae with irregular branches in right angles in favor of mucormycosis.

and Treatment of Cancer/Invasive Fungal Infections Cooperative Group (EORTC) and the National Institute of Allergy and Infectious Diseases Mycoses Study Group (MSG) published standard definitions for invasive fungal infections for clinical and epidemiological research (1). Corticosteroid use for at least 10 days, neutropenia of ≤ 500 cells/ μ l and lymphopenia as a lymphocyte count of ≤ 500 cells/ μ l were considered as the most important risk factors for developing mucormycosis (2). In the largest population-based epidemiological study of mucormycosis among children with hematological malignancies conducted by Israeli Study Group of Childhood Leukemia, most cases of mucormycosis (92%) had been occurred in children with acute leukemias. According to that registry, the incidence of mucormycosis was 2.2% in the children with ALL and 2.1% in the children with AML (3). Most of the underlying diseases or conditions for mucormycosis have been reported to be hematological malignancies and solid organ transplantation (4). The most frequent sites of infection are rhinocerebral, followed by pulmonary, disseminated, gastrointestinal, or cutaneous; however, sporadic cases of invasive mucormycosis of the liver have been reported (4).

Hepatic mucormycosis is rare and is usually thought to be arisen from gastrointestinal disease, although dissemination may occur from the other sources (5). An extensive literature review in 2019 identified 14 cases of hepatic mucormycosis without lung involvement. Three cases had splenic lesions and four had gastrointestinal lesions, suggesting the possibility of transmission to the liver and/or spleen from the gastrointestinal tracts (4). pure fungal abscesses are almost always seen in patients being treated for hematologic malignancies. The presence of fungemia along with fungal hepatic abscess is a poor prognostic factor with overall mortality approaching 80% (6). Hepatic mucormycosis is readily seen on abdominal imaging. Typical CT findings include the presence of hypodense hepatic lesions surrounding vessels without a mass effect, representing areas of liver necrosis due to fungal thrombosis. These findings are not specific but suggest the presence of an angioinvasive organism (7).

In a case report and review of the literature, all cases of splenic mucormycosis by searching PubMed publications till October 2020 has been reported. The search yielded 27 cases of splenic mucormycosis including their case. Many of the cases had underlying immunocompromised conditions, except for 6 cases. A male predominance was noted in 22 cases of splenic mucormycosis. Simultaneous involvement of stomach and spleen was seen among four immunocompromised patients. The diagnosis of splenic mucormycosis should be considered in front of splenic lesions suggesting abscess or infarction, especially among immunocompromised patients (8). Similar to our case, three children with relapsed ALL has been reported who developed fever and splenomegaly due to fungal

splenic abscess, two during the re-induction phase of chemotherapy and the third within one month after achieving a second remission (9).

Conflict of Interest: None declared.

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