Naso Orbital Chloroma with Mucormycosis of Maxillary Sinus - A Varied Clinical Presentation

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Abstract

Rhino-orbito-cerebral mucormycosis is a rapidly progressive and fatal disease that mostly occurs in patients with diabetes mellitus and immunocompromised status. Antifungal therapy is the standard of care. Surgical debridement depends upon the hemodynamic status of the patient. Here we are reporting a case of mucormycosis in a 43 year female with type II diabetes mellitus and acute myeloid leukemia.

Case Presentation

A 43-year-old Mrs. S. came to the outpatient department of our hospital with complaints of cough with expectoration and hemoptysis for the past ten days (Figure 1). She had been taken to a local hospital where she underwent treatment and was given two units of blood transfusion. Three days after the transfusion, she developed a painful swelling over the right eye and cheek associated with loss of vision. The patient is a known case of bronchial asthma and type II diabetes mellitus for which she was not on regular medications. On examination, she was conscious and cooperative, and her vital signs were stable. She had no pallor, icterus, clubbing, cyanosis or lymphadenopathy. Purpuric rashes were present over her upper and lower extremities. Local examination of the right eye revealed blood stained discharge, proptosis and periocular edema. Swelling and induration were present over the right side of her face. Examination of the nose showed a deviated nasal septum to the right with a normal mucosa. On examination of the oral cavity, clots were seen over the palate and tongue. An impression of right orbital cellulitis with hemoptysis under evaluation with a possible transfusion reaction was made. Blood investigations revealed low hemoglobin (7.7 g/dL), raised total leucocyte count (122,460), and low platelet count (35,000). Peripheral blood smear was suggestive of acute leukemia probably of myeloid origin (Figure 2). Histopathological examination of the biopsy specimen from the right maxillary sinus showed a fungal granuloma. The leukocytic infiltrate found adjacent to the mucormycosis was suspicious of leukemic involvement (Figure 3). CT scan of the para-nasal sinuses showed sinonasal polyposis on the right side with cellulitis of the cheek, possibly due to an abscess in the right parotid gland (Figure 4). Also, proptosis of the right globe with orbital fat stranding and thickening of the inferior rectus muscle was noted (Figure 5). Video laryngoscopy showed edematous epiglottis, arytenoids and aryepiglottic folds. Both vocal cords were mobile with an adequate glottic chink. Patient was started on induction chemotherapy with Cytosine-160mg OD for 6 days and Daunorubicin-80 mg OD for 3 days, anti-fungals, Amphotericin-B-50 mg iv OD and Antibiotics inj. Piperacillin& tazobactam, linezolid. Insulin was given according to sugar values. Total counts gradually reduced over 20 days. Patient were started on G-CSF. Antibiotic changed to vancomycin for febrile neutropenia. Blood culture grew Klebsiela pneumonia and wound swab culture grew - E.Coli, Proteus mirabilis. Inj. Imipenem was added according to sensitivity pattern. Inj. Amphotericin-B was continued. Total counts improved to 3440 with time. During the course of hospital stay a total of 36 platelet concentrates and 10 packed cells were transfused. Surgical debridement was done as indicated. Patient is on regular follow up with improved clinical condition.

Discussion

Over the last decade, the incidence of fungal infections has increased considerably due to increase in illness like AIDS, immunosuppressive therapy due to organ transplant and other diseases like lymphomas and leukemias. Mucormycosis (also called zygomycosis) is a serious, relatively uncommon invasive fungal infection and one of the most aggressive and lethal invasive mycoses. Fungi from the order Mucorales are the etiologic agents of mucormycosis. Rhizopus and Rhizomucor are the genera usually cultured from tissue samples [1]. Mucorales are found commonly in the environment, and spores of these usually nonpathogenic fungi are likely to be inhaled daily [2]. In the normal human lung, spores are inhibited from germinating into hyphae by alveolar macrophages. Mucormycosis occurs in patients already having a preexisting immunocompromised disease eg., AIDS, or uncontrolled diabetes, or those under immunosuppressive therapy, patients receiving renal dialysis and desferrioxamine are also at increased risk for mucormycosis, but can infect healthy individuals as well [3]. In this case the patient was a case of uncontrolled type II diabetes mellitus and AML. Uncontrolled diabetes mellitus can alter the normal immunologic response of patients to infections. Such patients have decreased granulocyte phagocytic ability with altered polymorphonuclear leukocyte response. Reports have suggested that the ability of serum of immunocompromised patients to inhibit Rhizopus in vitro is reduced, which makes them suitable hosts to opportunistic fungal infections [4]. Mucormycosis is aggressive and potentially fatal in diabetic patients because of impaired host
defense mechanism and increased availability of micronutrients such as iron. This fungal infection usually originates from the paranasal sinuses. The fungus invades the blood vessels and subsequently spreads through them. Once fungal hyphae enter into the blood stream they can disseminate to other organs such as cerebrum or lungs which can be fatal for the patient. Mucor hyphae form thrombi within the blood vessels that reduce vascularity to the tissues and cause necrosis [5]. Usually mucormycosis occurs as a pulmonary, gastrointestinal, disseminated or rhino cerebral infection [6]. There is a close histopathological resemblance between mucormycosis and aspergillosis. Microscopically, aspergillosis has septate branching hyphae, which can be distinguished from mucormycotic hyphae by a smaller width and prominent acute angulations of branching hyphae. A definitive diagnosis of mucormycosis can be made by tissue biopsy that identifies the characteristic hyphae, by positive culture or both. Amphotericin B was administered parenterally as it is the drug of choice in treatment of mucormycotic infection. Blood urea & creatinine levels were monitored. Long-term treatment is required as the drug is fungi static rather than fungicidal. Clinical recovery like a febrile state, better wound healing and negative cultures from nose serves as guide for cessation of treatment. Once patient starts improving amphotericin- B is given on alternate days. Nephrotoxicity is seen in upto 80% of patients and is manifestated by azotemia, electrolyte wasting (hypokalemia) and decrease in urine concentrating ability [7,8]. Serum creatinine levels >3 mgm/dL and BUN 50 mg/dL requires temporary discontinuation of therapy.

Conclusion
Mucormycosis co existing with chloroma in the naso orbit region and an associated co morbid state like Type 2 Diabetes Mellitus must be treated aggressively with a very meticulous follow up protocol. Anti fungal drugs must be administered with caution, especially in compromised renal functions.

References