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Case Report

Mucormycosis And Hemophagocytosis Syndrome in A Patient Following Acute Myeloid Leukemia

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Abstract:

Acute Myeloid Leukemia (AML) is a malignant disease that affects the bone marrow and may affect red blood cells, white blood cells(myeloblasts), and platelets. Some cases of disease may be asymptomatic and can be diagnosed from random blood tests. AML mostly affects cells that are not fully developed, causing these cells to fail to perform their normal functions. If this disease is left untreated, it gets worse quickly and can be fatal. Genetic factors may cause the development of AML, but the exact cause is not known. There are environmental factors in AML etiology (eg, chemicals, radiation, tobacco, chemotherapy), in some patients, AML may occur due to clonal hematopoiesis manifest as a myelodysplastic syndrome, myeloproliferative neoplasms, paroxysmal nocturnal hemoglobinuria, aplastic anemia and more. Here we present a case report of 59 years old male patient presenting to the clinic with malaise and referred to haematology department due to pancytopenia. However, further investigations confirmed AML.

Keywords: Acute Myeloid Leukemia (AML),Mucormycosis, Hemophagocytic lymphohistiocytosis (HLH), case report, mucormycosis, hemophagocytosis

Introduction

Acute Myeloid Leukemia (AML) is a malignant disease that affects the bone marrow and may affect red blood cells, white blood cells(myeloblasts), and platelets. Some cases of disease may be asymptomatic and can be diagnosed from random blood tests. AML mostly affects cells that are not fully developed, causing these cells to fail to perform their normal functions. If this disease is left untreated, it gets worsely quickly and can be fatal. Genetic factors may cause the development of AML, but the exact cause is not known. There are environmental factors in AML etiology (eg,chemicals, radiation, tobacco, chemotherapy), in some patients, AML may occur due to clonal hematopoiesis manifest as a myelodysplastic syndrome, myeloproliferative neoplasms, paroxysmal nocturnal hemoglobinuria, aplastic anemia etc.

Clinical presentation: anemia (eg, shortness of breath, weakness, dyspnea), thrombocytopenia (excess bleeding or bruising), and neutropenia (fever, infections),hepatomegaly, splenomegaly are common but lymphadenopathy is rare.

Mucormycosis usually occurs in immunosuppressed hematology patients and is fatal. Since this disease does not have a specific clinical method, it is often not diagnosed. Mucormycosis is caused by opportunistic fungal pathogens and is more common in immunosuppressed hematology patients receiving chemotherapy or in patients who have undergone bone marrow transfer. These microorganisms are found everywhere in nature and in decaying

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plants and soil. All people encounter this fungus every day. The only reason why this disease is rare in humans is the immune system

RISK FACTORS:

- Diabetes mellitus, particularly with ketoacidosis
- Treatment with glucocorticoids
- Hematologic malignancies
- Hematopoietic cell transplantation
- AIDS
- Injection drug use
- Trauma/burns
- Malnutrition

The most common occurrence is nasal mucus, which occurs when the fungus enters the sinuses through inhalation. The most common symptom we encounter in the clinic is nosebleeds. Headache, nasal sores and nasal congestion may be other clinical symptoms. The second most common form of mucormycosis is lung infection after inhaling the fungus. Pulmonary infection occurs most frequently in patients with neutropenia.

Hemophagocytic syndrome is a hyperinflammatory state caused by overstimulated but dysregulated and often ineffective immune responses.In HLH, NK cells fail to eliminate activated macrophages. This results in excessive CD8+ T cell and macrophage activation and elevated levels of interferon gamma and other cytokines leading to HLH pathology. In addition to antigen presentation and cytokine production, macrophages can also phagocytose host cells. Hemophagocytosis is part of the sepsis syndrome caused by severe infection. Hemophagocytosis may be seen in tissue biopsies (lymph nodes, spleen, liver) or bone marrow aspirates/biopsies.

clinical findings:

- Fever
- Splenomegaly
- Bicytopenia
- Hypertriglyceridemia or hypofibrinogenemia
- Ferritin >500 mcg/

Case Report

A 59-year-old male patient applied to our hematology department due to pancytopenia in the blood results obtained from an external center. The patient was diagnosed with AML as a result of the examinations and bone marrow aspiration.

The patient followed as AML 7+3 treatment protocol has been planned. The patient received antibiotic treatment for various reasons while receiving chemotherapy. After receiving treatment, the patient was deemed suitable for bone marrow transplantation. Approximately 1 month after the bone marrow transplantation, the patient started to complain of skin rash, fever, diarrhea and vomiting. As a result of the biopsy taken from the patient's skin by dermatology, Graft versus host disease grade 2 was determined. The patient was routinely followed up by the infectious diseases department and the stool tests were positive for CMV PCR and treatment was started. Hepatomegaly was detected in the patient's abdominal USG results. The patient had neutropenia and received antibiotic treatment due to infection during this period. The patient complained of nosebleeds and the ear, nose and throat department was consulted. During the examination, a lesion was observed in the nasal septum and a biopsy was taken. The biopsy result was urgently evaluated by pathology and the result was

mucormycosis.The patient was taken into surgery. He continued to be intubed in the ICU. mucor treatment was started The patient was intubated and monitored in intensive care for a while. A peripheral blood smear was taken from the patient and phagocytosed macrophages were observed in the smear. The patient's peripheral blood smear and laboratory results were compatible with hemophagocytosis syndrome disease. Extubation of the patient was planned, but the patient did not wake up. The patient was followed in the intensive care unit and exitus within a few days.

LAB:

Hdl cholesterol:6 mg/dl
Cholesterol:5 mg/dl
Ldl cholesterol:164 mg/dl
Vldl cholesterol:76 mg/dl
Triglyceride:381 mg/dl
Fibrinojen:439,69 mg/dl
Ferritin:27781 pg/ml
Creatinine 1,5 mg/dl
Sodium (na) 141 mmol/l
Potassium (k) 3,76 mmol/l
Chlorine (cl) 99 mmol/l
Calcium (ca) 6 mg/dl
Phosphorus (p) 6,5 mg/dl
Wbc 0.47 10^3/ul
Hgb 9.00 g/dl
Ly# 0.020 10^3/ul
Mcv 82.50 fl
Mo# 0.02 10^3/ul
Ne# 0.43 10^3/ul
Plt 95.00 10^3/ul

Discussion and conclusions

Acute Myeloid Leukemia (AML) is malignancy of the white blood cells that leads to bone marrow failure and organ infiltration. Untreated, AML is fatal and life-threatening.[3]

Mucormycosis, caused by opportunistic pathogenic fungi, is a difficult-to-diagnose with high mortality that commonly occurs in patients with impaired immune status, particularly those with diabetes mellitus, hematological malignancy, and neutropenia.[4]

If patients with pencetopenia and low immune system have fever of unknown reason, hemophagocytosis syndrome may be considered.[8] In HLH, NK cells fail to eliminate activated macrophages. This results in excessive CD8+ T cell and macrophage activation and elevated levels of interferon gamma and other cytokines leading to HLH pathology.

After our patient was diagnosed with AML, chemotherapy treatment started. The patient had entered pancytopenia due to chemotherapy. Patient who developed GVHD after transplantation ,the extended duration of hospitalization caused a patient with a low immune system to be mucor. our patient had both mucor and hemophagocytosis syndrome and both are mortal. Unfortunately he passed away.

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