Case Report

Acute myeloid leukemia presenting as polyserositis and leukemia cutis

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ABSTRACT

Acute leukemia generally present with nonspecific symptoms such as fatigue, weakness, malaise, anorexia, weight loss, fever, bone pains, bruising, or bleeding that begin gradually and are the consequence of associated cytopenias. Polyserositis with predominant pericardial effusion clinically manifested as heart failure as the presenting feature of acute myeloid leukemias (AMLs) has been rarely described. In this report, we describe a case of a 21-year-old male, who presented with symptomatic serositis and leukemia cutis and was subsequently diagnosed as AML-myelomonocytic type (AML-M4).

Key words: Acute myeloid leukemia, extramedullary leukemia, leukemia cutis, pericardial effusion, pleural effusion

INTRODUCTION

Acute myeloid leukemias (AMLs) are malignant disorders of the hematopoietic stem cell compartment. Patients with AML usually present with bone marrow failure that causes symptoms of anemia, bleeding from thrombocytopenia, and neutropenic infections. Extramedullary disease and cutaneous manifestations are rare presenting features of the disease.[1] Extramedullary manifestations can manifest simultaneously with, or precede bone marrow involvement. Usual sites of extramedullary involvement include bone, joint, periosteum, gums, skin and lymph nodes, and less commonly the orbit, oropharynx, paranasal sinuses, intestine, mediastinum, breast, central nervous system, epidural region, uterus, and ovary.[2,3] Sweet syndrome and chloromas are the other rare extramedullary presentation of AML.[3] Sludging of blood flow as a result of leukostasis can cause pulmonary manifestations (severe dyspnea and diffuse lung shadowing), retinal hemorrhages, or neurological manifestations (altered mental status, ocular muscle palsy, etc.). Pleural or pericardial effusion in a patient of AML usually develops during the course of leukemia as a result of drug toxicity, underlying infections, leukemic infiltration, or other disseminated solid tumors.[4-6]

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Access this article online

Quick Response Code:

Website: www.ccij-online.org

DOI: 10.4103/2278-0513.182063

CASE REPORT

A 21-year-old male, nonsmoker, was admitted with a history of progressive breathlessness and dry cough for15 days. His breathlessness had progressed from breathlessness on exertion to breathlessness at rest and orthopnea. He developed swelling over both lower limbs for last 7 days. He also noticed multiple raised nodules all over his body, which were not associated with itching. He denied any history of fever, anorexia, night sweats, weight loss, bone/joint pains, visual disturbances, or bleeding.

On physical examination, he was conscious, oriented, afebrile, and anicteric. He had pallor, resting tachycardia, tachypnea, blood pressure of 100/60 mmHg, raised jugular venous pressure, and bilateral pitting pedal edema. There were multiple violaceous nodules or papules distributed all over his body [Figure 1a and b]. They were about 3–20 mm

Cite this article as: Saxena A, Bodkhe S, Kulkarni AR, Jain AP. Acute myeloid leukemia presenting as polyserositis and leukemia cutis. Clin Cancer Investig J 2016;5:284-6.

in size, firm in consistency with smooth external surface, and nontender. Gum hypertrophy, lymphadenopathy, petechiae, ecchymosis, mucosal bleeding, and sternal tenderness were absent.

His cardiovascular system and respiratory system examination revealed signs of pericardial effusion and bilateral pleural effusion, respectively. His abdominal examination revealed the presence of flank dullness, shifting dullness, and tender hepatomegaly.

His hemogram revealed a total leukocyte count of 44,200/mm^3, hemoglobin of 8.2 g/dl, and platelet count of 180,000/mm^3. His peripheral blood smear showed myeloblasts and monoblasts comprising a total of 70% of the differential leukocyte count [Figure 2a]. Bone marrow aspirate and biopsy showed hypercellularity with a diffuse infiltration by myeloblasts or immature myelomonocytic cells (75%) [Figure 2b]. The blast cells were positive for CD13, CD14, CD33, CD64, and CD117 antigens and negative for CD34, CD3, CD5, CD7, CD10, and CD19 antigens on immunophenotyping based on which a diagnosis of AML-myelomonocytic type (AML-M4) was made. His liver, renal, thyroid, and coagulation profile were all within normal limits. Serum lactate dehydrogenase was raised, 1110 U/L (normal, 90–221 U/L). His serological tests were negative for antinuclear antibodies, hepatitis B virus, hepatitis C virus, and human immunodeficiency virus.

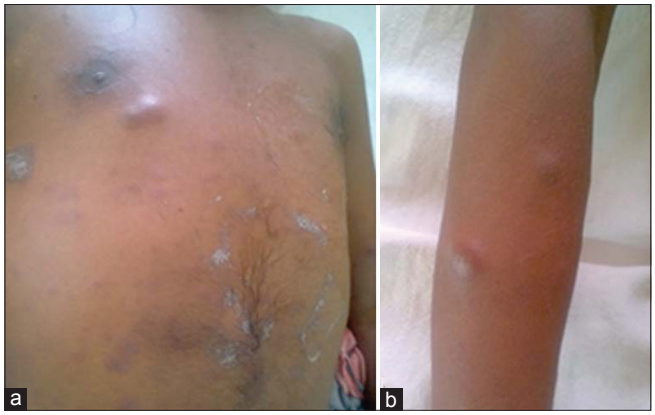

Figure 1: (a) Multiple violaceous nodules/papules distributed over the trunk. (b) Multiple nodules present over his right forearm

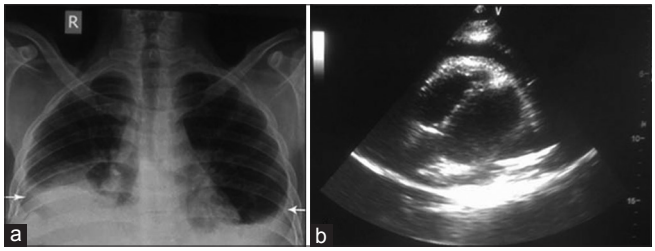

Figure 3: (a) Chest radiograph showing cardiomegaly with bilateral pleural effusion. (b) Echocardiogram (apical view) showing pericardial effusion

His chest radiograph revealed enlargement of the cardiac silhouette with bilateral pleural effusion [Figure 3a]. Echocardiogram revealed moderate pericardial effusion with the dilatation of the inferior vena cava [Figure 3b]. Ultrasonography of abdomen revealed hepatomegaly and free fluid in the abdomen with no evidence of any mass or lymphadenopathy. A computerized tomographic scan of his chest with contrast revealed bilateral pleural effusion and pericardial effusion with no evidence of any mediastinal or intrathoracic mass.

Diagnostic pericardiocentesis revealed hemorrhagic fluid which was exudative by Light's criteria. The cytopathology specimen obtained from pericardial fluid was positive for myeloid blasts [Figure 4]. The pleural fluid tapped was also hemorrhagic and exudative in nature. However, the pleural fluid block cytology was negative for any malignant cells on two consecutive occasions. His pleural fluid adenosine deaminase was 14 U/L (normal <40 U/L). Pleural fluid cultures were negative for bacteria, fungi, and mycobacterium tuberculosis. His ascitic fluid was non-hemorrhagic and transudative by Light's criteria. Fine needle aspiration cytology from the skin lesions showed infiltration by myeloid blasts.

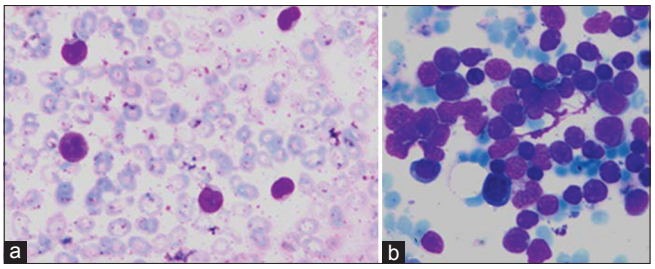

Figure 2: (a) Peripheral blood smear showing the presence of myeloblasts and monoblasts (Leishman, ×400). (b) Bone marrow aspirate showing the presence of plenty of myeloblasts (Giemsa, ×400)

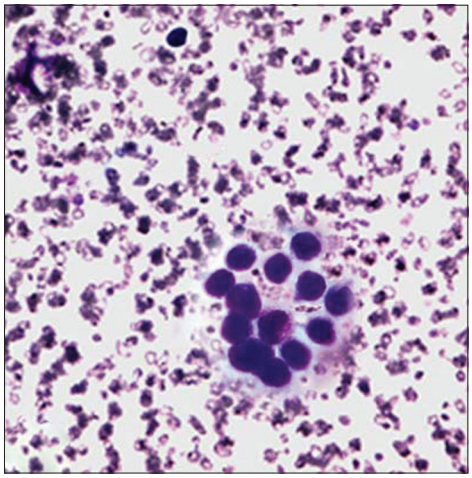

Figure 4: Pericardial fluid revealing the presence of myeloid blasts (Giemsa, ×400)

The patient was treated with standard induction chemotherapy for AML including cytarabine (100 mg/m^2/day; 7 days infusion) and daunorubicin (45 mg/m^2/day on day 1–3). A repeat bone marrow aspiration and biopsy performed on day 15 showed complete remission of leukemia. His repeat echocardiogram and chest radiograph on day 15 revealed a significant reduction in the size of pericardial effusion and pleural effusion, respectively. His cutaneous lesions had also regressed.

DISCUSSION

Malignant pleural effusions are more commonly seen with solid malignancies and among the hematolymphoid group, it is common in Hodgkin's and non-Hodgkin's lymphomas.[7] AML presenting with pleural effusion or pericardial effusion is a rare entity. The possible etiologies of pleural effusion in patients of leukemia as reported in the literature are leukemic infiltration into the pleura, pleural reaction secondary to intrapleural bleeding, underlying infections, drug toxicity, or by metastatic or rarely autoimmune causes. In most of the cases, resolution of the pleural fluid occurs with the treatment of leukemia, whereas the resistant or the relapsing cases may require a pleurodesis.[7]

Pericardial effusion as an extramedullary involvement at the initial presentation of AML is extremely rare.[8] It is usually observed in the course of patients undergoing treatment for leukemia as a result of chemotherapy toxicity or infections.[9]

The prognostic significance of pleural effusion or pericardial effusion in cases of AML is uncertain. Some authors are of the view that it does not affect the rate of remission and survival while others relate it to a worse prognosis.[10] Cytogenetic abnormalities with poor prognosis in AML include complex karyotype, -5, del (5q), -7, del (7q), or 3q21, 3q26, 11q23 abnormalities. Unfortunately, the cytogenetic analysis could not be done in our patient due to financial constraints.

Cutaneous solitary or multiple lesions of varied morphology, manifesting as leukemia cutis occurs as a result of neoplastic leukocytic infiltration of the skin. In leukemia, a wide spectrum of skin lesions including papules, macules, plaques, or nodules which may be skin-colored to erythematous or violaceous colored, generalized urticaria, exanthematous eruptions, purpura, Sweet's syndrome like lesions, leukocytoclastic vasculitis, and granuloma annulare-like lesions have been described in the literature.[11,12] Like other forms of extramedullary disease, leukemia cutis is generally associated with poor prognosis in patients with AML.[13]

CONCLUSION

We present this case so as to increase awareness among physicians that pericardial effusion with/without polyserositis manifesting clinically as heart failure may be the only initial presentation of acute leukemia. Its early recognition is essential for proper evaluation and management of disease.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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