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CASE REPORT

Granulocytic Sarcoma with AML in Pregnancy

Das Subrata · Hajra Jyotirmoy · Roychowdhury Joydeb

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Introduction

Granulocytic sarcoma (chloroma) is a mass of malignant early myeloid precursor cells in an extramedullary location [1, 2]. Rarely, the ovary may be the first site for clinical manifestation of granulocytic sarcoma. The estimated incidence of acute myelocytic leukemia (AML) in pregnancy is 1 in 75,000 [3]. Granulocytic sarcoma of the ovary in association with AML has been rarely reported in the literature. However, only a few such cases have been reported in association with pregnancy [2].

Case History

A 20-year-old primigravida presented to our obstetric department with severe pallor, lethargy, epistaxis, inability to walk, and unilateral leg pain. She had a single live fetus with cephalic presentation of 30-weeks 4 days gestation from her last menstrual period (LMP). She was a carrier of beta-thalassemia. On examination, she had severe pallor. Her blood pressure was 100/60 mmHg and pulse rate was 94/min. On admission, her hemoglobin level was 6.2 g%. Peripheral blood smear showed hypochromic, microcytic red blood cells, target cells, and plenty of immature blast cells. Her platelet count was 22,000/mm^3 and blast cells were 75 %. The bone marrow was markedly hypercellular with 40 % of blast cells. These cells showed few nuclear chromatins, prominent nucleoli, and basophilic cytoplasm (Fig. 1); flow cytometry showed CD13, CD33, CD 34 positivity. Cytogenetic study showed a normal XX pattern. Based on the clinical features and laboratory findings, a diagnosis of AML FAB M2 Subtype (acute myeloblastic leukemia with granulocytic maturation) was made. Ultrasonography showed single live fetus with cephalic presentation with gestational age of 32 weeks 5 days; the placenta was situated in the anterior part of upper segment of the uterus with grade II maturity, and liquor amnii was adequate in volume. She was transfused 10 U of packed RBC and 24 U of platelet concentrates. Her pregnancy was terminated at 33 weeks by lower segment cesarean section (as per advice of hematologist) in order to start chemotherapy. Corticosteroid was administered for lung maturity of the fetus 4 days before termination. The patient delivered a preterm female baby with 1.7 kg birth weight and good Apgar scores.

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Das S., Assistant Professor · Hajra J., Senior Resident · Roychowdhury J., Professor and Head of the Department
Department of Obstetrics & Gynecology, ESI Postgraduate Institute of Medical Sciences and Research & ESIC Hospital, Diamond Harbour Road, Joka, Kolkata 700104, India

Das S. (✉), Assistant Professor
J/6, Banerjee para, Kamdahari, Garia, Kolkata, 700084, West Bengal, India
e-mail: drsubrata01@yahoo.co.in

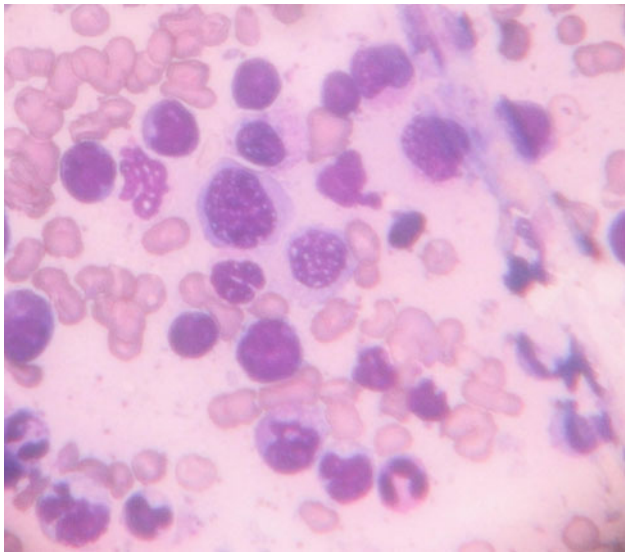

Fig. 1 Hypercellular bone marrow with depressed erythropoiesis, myeloid maturation shift to the left, occasional functioning megakaryocytes and blast cell. (Microscopic picture of bone marrow, Leishman staining, ×40)

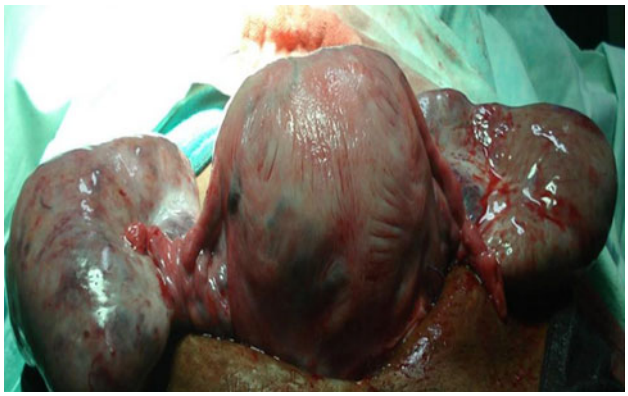

Fig. 2 Bilateral ovarian tumor (granulocytic sarcoma) detected during cesarean section

Per-operatively, we noticed two bilateral reniform ovarian masses each measuring 12.5 × 12.5 cm (Fig. 2). The masses were removed and sent for histopathological examination. Histopathology revealed that there was complete replacement of ovarian parenchyma by eosinophilic cells of myeloid origin (Fig. 3) and a diagnosis of granulocytic sarcoma was made. After an uneventful postoperative recovery, she was transferred to a regional cancer hospital for chemotherapy where she was treated with cytarabine and daunorubicin (7 + 3 days regimen). Cytarabine 100 mg/m^2 daily from day 1 to 7 and daunorubicin 60 mg/m^2 daily iv from day 1 to 3 were given.

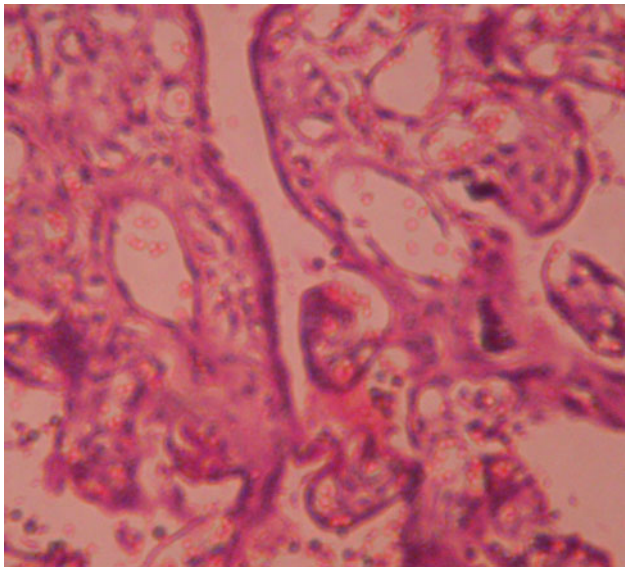

Fig. 3 Complete replacement of ovarian parenchyma with sheets of malignant neoplastic cells. Variable amounts of eosinophilic cytoplasm with round to oval nucleus are present within the neoplastic cells. (H and E staining, ×40)

However, the patient suffered from severe chest infections leading to septicemia which was non-responsive to intravenous ceftriaxone, metronidazole and amikacin antibiotics, and the patient died on the 8th day of completion of chemotherapy.

Discussion

Granulocytic sarcoma was also known as chloroma [1, 2]. This name was derived from the Greek word "chloros" (green) as these tumors often have a greenish tint due to presence of myeloperoxidase. Rappaport renamed it granulocytic sarcoma in 1967. Currently, any extramedullary manifestation of acute myeloid leukemia may be termed as granulocytic sarcoma or chloroma.

It occurs only in 32.3 % of patients with granulocytic leukemia, being clinically evident in less than 1 % of the patients. Granulocytic sarcoma may involve virtually any organ or tissue. The bone is the commonest site of involvement [2]. Involvement of the ovaries may occur rarely. Biopsy of the lesion followed by histopathological examination formed the basis of the diagnosis.

Association of granulocytic sarcoma in a patient with AML indicates poor response to treatment and poor prognosis [1]. The tumor is quite sensitive to antileukemic chemotherapy. If sarcoma is persistent even after completion of induction chemotherapy, local surgery or radiation therapy is often considered. In our patient, the tumor was

found during cesarean section and in order to establish a diagnosis, excisional biopsy was considered.

Conclusion

Leukemia along with granulocytic sarcoma in pregnancy is a very rare entity and often proves fatal. Our case also emphasizes the importance of controlling secondary infection in order to prevent the increased morbidity and mortality associated with such cases.

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