Keywords: Cutis laxa, Monoclonal gammopathy of renal significance, Chemotherapy

Anahtar Sözcükler: Kutis Laksa, Renalin monoklonal gammopatisi önem, Kemoterapi

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Recurrent Pregnancy-Related Pure Red Cell Aplasia Responsive to Combined Corticosteroid and Azathioprine Therapy: A Management Dilemma

Kortikosteroid ve Azatioprin Kombine Tedavisine Yanıt Veren Tekrarlayan Gebelik-İlişkili Saf Kırmızı Hücre Aplazisi: Bir Yönetim İkilemi

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To the Editor,

Pure red cell aplasia (PRCA) is associated with normocytic normochromic anemia, severe reticulocytopenia, and reduced erythroid precursors in the bone marrow [1]. PRCA may be congenital or acquired. Among the acquired cases, pregnancy has been associated with PRCA [2].

A 25-year-old primigravid patient presented at 10 weeks of gestation with severe anemia with breathlessness for 2 months with no history of fever, jaundice, or weight loss. On examination, she had pallor but no icterus, peripheral lymphadenopathy, or hepatosplenomegaly. Laboratory investigations showed a hemoglobin level of 5.5 g/dL, red blood cell (RBC) count of 1.53 million/mm^3, and white blood cell (WBC) count of 5280/µL. Her mean corpuscular volume, mean corpuscular hemoglobin, and mean corpuscular hemoglobin concentration were 81.2 fL, 25.8 pg, and 31.8 g/dL, respectively. A peripheral smear showed normocytic normochromic RBCs with occasional spherocytes with reticulocytes of 0.3%. Hence, the absolute reticulocyte count was 200/µL and the reticulocyte index was 0.15, which was suggestive of hypoproliferation. Ultrasound showed a single live intrauterine pregnancy corresponding to 9 weeks. On admission, packed RBCs were transfused. Hemoglobin electrophoresis was performed, which ruled out thalassemia. Other investigations for liver, renal, and endocrine functions were normal. Bone marrow aspirate cytology (Figure 1) showed occasional basophilic erythroblasts and promegaloblasts with moderate lymphocytosis. The percentage of erythroid precursors was 0.8%

and the myeloid erythroid ratio was 85:1. The overall marrow cellularity was near normal. Serological tests for toxoplasma, rubella, cytomegalovirus, hepatitis B and C antibodies, and parvovirus IgM were found to be negative. Tests for antinuclear antibodies (ANA), ds-DNA, anticardiolipin antibodies, and lupus anticoagulant were also negative. The patient was started on oral prednisolone at 1 mg/kg body weight daily and was slowly tapered to 5 mg/day in the second trimester. At 23 weeks, her hemoglobin was 4.3 g/dL (Figure 2), although there was no history of bleeding; hence, oral azathioprine at 50 mg twice daily was started. Hemoglobin levels were checked periodically and corrected. The fetal growth and development were assessed regularly. Subsequently, her hemoglobin improved and she did not require further blood transfusions. At 38 weeks of gestation, she vaginally delivered a male baby weighing 2.6 kg. There were no congenital abnormalities observed. After delivery, hemoglobin was found to be 11.2 g/dL. The steroids were tapered and stopped after 4 weeks. She was in remission for 2 years when she presented with amenorrhea for 2 months associated with breathlessness. Ultrasound revealed intrauterine gestation corresponding to 8 weeks. Her hemoglobin was 4.3 g/dL with hematological findings as observed previously. Oral prednisolone was started and packed RBCs were transfused. As she was unwilling to continue the pregnancy, medical termination of pregnancy was performed, a copper intrauterine device was inserted, and prednisolone was tapered as before.

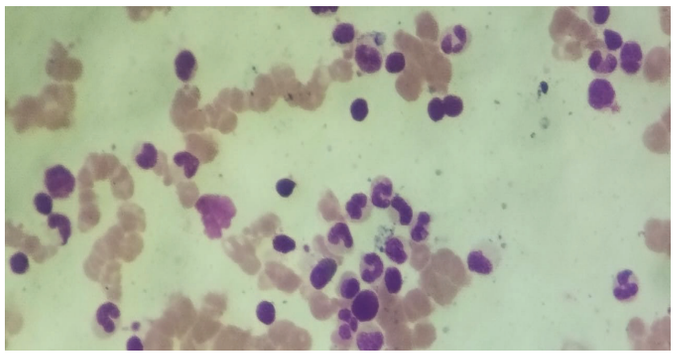

Figure 1. Bone marrow aspirate cytology showed occasional basophilic erythroblasts and promegaloblasts with moderate lymphocytosis.

PRCA rarely occurs during pregnancy but it should be considered as a cause of refractory anemia in pregnancy. It can manifest in any trimester and is diagnosed by bone marrow

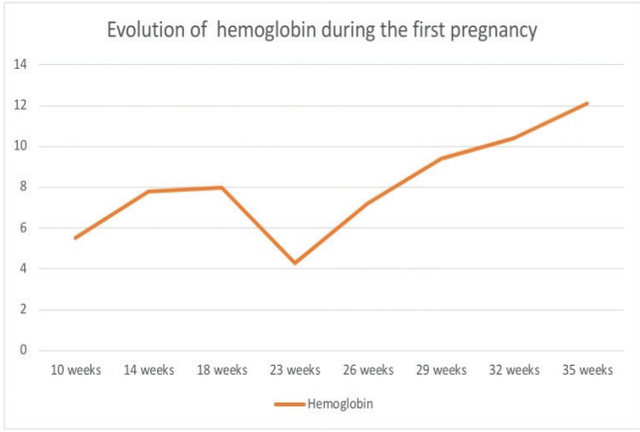

Figure 2. Hemoglobin values during first pregnancy.

Table 1. Different modes of treatment used in pregnancy-related pure red cell aplasia by different authors.

Author	Year	Diagnosis at presentation	Mode of treatment	Gestational age at delivery	Mode of delivery	Conditation of infant at brith	Congenital anomalies	Time taken for normalization of hemoglobin postpartum
Miller and Rashid [3]	2008	Primigravida with anemia	Blood transfusion alone	34 weeks	Vaginal delivery	Alive	None	4 weeks
Kashyap and Pradhan [4]	2010	Fourth pregnancy with anemia	Blood transfusion with corticosteroid therapy	37 weeks	Vaginal delivery	Still born	None	4 weeks
Ito et al. [5]	2012	Primigravida with anemia	Blood transfusion alone	38 weeks	Vaginal delivery	Alive	N/A	5 weeks
Aggarwal [6]	2013	Primigravida -15 days post cesarean section with anemia	Blood transfusion alone	N/A	Cesarean section	Alive	N/A	8 weeks
Edahiro et al. [7]	2019	Primigravida with anemia	Blood transfusion with corticosteroid therapy	37 weeks	Vaginal delivery	Alive	None	28 weeks
Our case	2019	Primigravida with anemia	Blood transfusion with corticosteroid and azathioprine	38 weeks	Vaginal delivery	Alive	None	4 weeks

Hb: Hemoglobin; N/A: not available.

aspirate cytology. In pregnancy, management is performed with prednisolone, although immunosuppressive therapy has also been used [2]. In Table 1, the different modes of treatment used in cases of pregnancy-related PRCA by different authors are summarized [3,4,5,6,7]. PRCA in pregnancy has a better prognosis compared to pre-existing PRCA and aplastic anemia in pregnancy. In general, PRCA that develops during pregnancy spontaneously resolves postpartum [8]. Although it rarely recurs in subsequent pregnancy, recurrence was observed here and hence the permanence of PRCA is not known [3].

Keywords: PRCA, Acquired pure red cell aplasia, Anemia in pregnancy, Hypoproliferation, Erythroid progenitors, Promegaloblast

Anahtar Sözcükler: PRCA, Edinsel saf kırmızı hücre aplazisi, Gebelikte anemi, Hipoproliferasyon, Eritroid öncüller, Promegaloblast

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A Case of Burkitt's Lymphoma Mimicking Peritonitis Carcinomatosa

Peritonitis Karsinomatozayı Taklit Eden Bir Burkitt Lenfoma Olgusu

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To the Editor,

A 30-year-old man was admitted to the hospital with fatigue, fever, nausea, and abdominal distension in August 2019. Laboratory analyses were as follows: white blood cell count, 13,400/µL; absolute neutrophil count, 9,700/µL; absolute lymphocyte count, 2,100/µL; hemoglobin, 14.5 g/dL; platelets, 442,000/µL; C-reactive protein, 15.6 mg/L; lactate dehydrogenase, 186 U/L; ferritin, 927 ng/mL; alanine transaminase, 108 U/L; aspartate transaminase, 245 U/L. Abdominal ultrasound showed massive ascites. Cytospinning of the ascites revealed B-cell non-Hodgkin's lymphoma. PET-CT showed increased FDG uptake of the whole peritoneum, omentum, and small intestine (Figure 1). Peritonitis carcinomatosa was considered in the differential diagnosis. The patient underwent tru-cut peritoneal biopsy; the findings were consistent with Burkitt's lymphoma. In immunohistochemical analysis, CD20, CD10, bcl6, and c-myc were positive; CD5, bcl2, CD23, MUM1, and TDT were negative. The Ki-67 index was 99%. FISH analysis for myc/IGH translocation was positive. Bone marrow was normocellular with no sign of lymphoma involvement and conventional cytogenetics showed a normal karyotype: 46, XY [20]. Cerebrospinal fluid cytospinning