Cesarean delivery in a patient with hypoplastic anemia with a very low refractory platelet level

Renal onkositom ile eş zamanlı endometrioid over ve endometrium kanseri

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Abstract

To describe the successful management of a patient with preexisting aplastic anemia by Cesarean section.

A 31-yr-old female patient with a hypoplastic myelodysplastic syndrome diagnosed six years back presented in this hospital was admitted with signs of pre-eclampsia (blood pressure of 140/90 mmHg, heavy proteinuria and moderate bilateral ankle edema), gum bleeding, and intrauterine growth restriction (IUGR) at 37 weeks amenorrhoea. Laboratory studies revealed pancytopenia (hemoglobin 6.4 $g \cdot dL^{-1}$, white cell count 2.7x $10^9 \cdot L^{-1}$, platelet count 10 x $10^9 \cdot L^{-1}$). Ultrasonography, however showed 35 week gestation with moderate IUGR. Patient had premature onset of labour and a Cesarean delivery was performed at 35 weeks with prophylactic platelet transfusion and meticulous blood pressure control. The procedure was uneventful, conducted under general anesthesia with an estimated blood loss of around 400 mL and a live female baby was delivered. Postoperatively her gum bleeding, pancytopenia improved but thrombocytopenia remained at discharge in spite of several units of platelet transfusion.

Hypoplastic anemia is rare in pregnancy and it is risky for both mother and fetus. The mother is at risk of aggravated nature of disease because of poor control, life-threatening episodes of bleeding and infection. A multidisciplinary team approach (obstetrician, anesthesiologist, hematologist, and pediatrician) is essential. While accurate assessment and close monitoring of the hematological condition should be made, it is very difficult to correct abnormalities before surgery. Emergent cesarean section proved to be life saving in this case.

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Key words: Aplastic anaemia, cesarean section, thrombocytopenia

Özet

Önceden aplatik anemisi olan bir hastanın sezeryan ameliyatının başarılı bir şekilde yönetiminin tanımlamak.

31 yaşında bayan hasta 6 yıl önce bu hastanede hipoplastik miyelodisplastik sendrom tanısı ile izlenmiş olup 37 haftalık amenoresinde preeklampsi belirtileri (kan basıncı 140/90mmHg, ağır proteinüri, bilateral yumuşak ayak bileği ödemi), dişeti kanaması, intrauterin gelişme geriliği (IUGR) ile hastaneye başvurdu. Laboratuar çalışmalarında pansitopeni (hemoglobin 6.4 $g \cdot dL^{-1}$,beyaz hücre sayısı 2.7x $10^9 \cdot L^{-1}$,trombosit sayısı10 x $10^9 \cdot L^{-1}$) ortaya kondu. Ultrasonografi, bununla birlikte orta seviyede IUGR ile 35 haftalık gebeliği gösterdi. Hastanın erken doğum sancıları mevcuttu ve 35. haftada trombosit transfüzyonu ve çok dikkatli kan basıncı kontrolu altında sezeryanla doğum yapıldı. Prosedür olaysızdı, doğum genel anestezi altında tahmini 400 mL kan kaybıyla yürütüldü ve canlı bir kız bebek doğurtuldu. Postoperatif diş eti kanaması ve pansitopenisi düzeltildi, fakat trombositopenisi birçok ünite trombosit transfüzyonu verilmesine rağmen düzeltilemedi.

Gebelikte hipoplastik anemi seyrektir ve hem anne hem de fetus için risklidir. Hastalığın doğası gereği anne güçsüzlük, hayatı tehdit edici kanama ve infeksiyon dönemlerinden ötürü şiddetli risk altındadır. Multi disipliner bir ekip tutumu (obstetrisyen , anestezist, hematolog, pediatrist) gereklidir. Ameliyattan önce anormalliklerin düzeltilmesi çok zordur, hematolojik koşulların iyi bir şekilde gözlemlenmesi ve doğru değerlendirilmesi gerekmektedir. Bu vakada acil sezeryanla doğumun hayat koruduğu ispatlanmış.

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Anahtar kelimeler: Aplastik anemi, sezeryanla doğurtma, trombositopeni

Introduction

Hypoplastic anemia is a grave hematological disorder with pancytopenia in the peripheral blood and hypocellularity in the bone marrow. Fortunately pregnancy is rare in it. There is risk of life-threatening episodes of bleeding and infection. Intra-uterine growth retardation and intrauterine death of the fetus is very common, while maternal infection can cause chorioamnionitis leading to preterm delivery. We describe a patient with both hypoplastic anemias with IUGR and pre maturity. A team of obstetricians, anesthesiologists, hematologists, and pediatricians managed the case. The patient showed absolute refractoriness and worsening of thrombocytopenia in spite many units of platelet transfusion and had premature onset of labour. Cesarean section was performed at 35 weeks and with a platelet count of 10 x 10^9 L^{-1}. Cesarean delivery in this low uncorrected refractory platelet level has not been previously described in the literature

Case Report

A 31-yr-old female patient with a hypoplastic myelodysplastic syndrome diagnosed six years back presented in this hospital in early second trimester and was admitted with signs of

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gum bleeding, and intrauterine growth restriction (IUGR), pre-eclampsia (blood pressure of 140/90 mmHg, proteinuria and moderate bilateral ankle edema), at 37 weeks amenorrhoea. She was diagnosed in Christian Medical College, Vellore and had a more or less stable disease with high dose of danazol. She had an elective abortion three years back and was in early second trimester when she first reported in this hospital. She had worsening of symptom and aplastic anaemia because of very low platelet, which required platelet transfusion several times. At 37 weeks amenorrhoea she had to be admitted for severe gum bleeding when laboratory studies showed hemoglobin (Hb) of 6.4 $g \cdot dL^{-1}$ (normal range: 11.5-16.5 $g \cdot dL^{-1}$), white cell count (WCC) 2.7 x $10^9 \cdot L^{-1}$ (normal range: 4-11 x $10^9 \cdot L^{-1}$), platelet count 10 x $10^9 \cdot L^{-1}$ (normal range: 150-400 x $10^9 \cdot L^{-1}$), prothrombin time (PT) 10.5 sec (normal range: 11.3-13.2 sec), albumin 28 $g \cdot L^{-1}$ (normal range: 42-54 $g \cdot L^{-1}$), aspartate aminotransferase (AST) 36 $U \cdot L^{-1}$ (normal range: 12-28 U·L-1), lactate dehydro-genase (LDH) 473 $\mu moL \cdot L^{-1}$ (normal range: 200-360 $\mu moL \cdot L^{-1}$), and 5.7 g of protein in a 24-hr urine collection. The platelet count could not be increased in spite of seven units of platelet transfusion. Ultrasonography, however showed 35 week gestation with moderate IUGR. Her Blood pressure could be checked with 10 mg nifedipine. When she had premature onset of labour in 35 week gestationwith very low and refractory platelet count a case conference was held and she was scheduled for a emergent Cesarean section at 37 weeks after two doses of I M betamethasone 12 mg to hasten fetal lung maturity. In view of pre-eclampsia and co-existent thrombocytopenia, general anesthesia was chosen.

One unit of packed red blood cells had been transfused. Inj methyl prednisolone was used preoperatively. The patient was premedicated with oral ranitidine 150 mg induction was performed with thiopentone 375 mg. Anesthesia was maintained with oxygen, nitrous oxide. Infraumbilical vertical midline incision of abdomen was done. Lower segment section was performed. Inj. Methergin was used on birth of anterior shoulder. Uterus contracted well and placenta separated on mild traction. There was no gross bleeding on separation while suturing uterus in two layers. Even suture points did not bleed and suturing could be done smoothly and uneventfully which is in sharp contrast to sutures of abdominal wall where continuous profuse oozing was just very difficult to manage. Cautery would not help and four layers of very close suturing with extra suturing for subcutaneous fat had to be used to make a bloodless exit. Total two units of blood were used for completion of operation. A live female baby weighing 2000g was delivered three minutes after skin incision, with Apgar scores 8 and 10 at one and five minutes respectively. Due to prematurity and intra-uterine growth retardation, an experienced neonatologist was present at delivery but the baby did not require intubation and transfer to the neonatal intensive care unit. Intravenous pethidine 50 mg was administered and the patient's intraoperative blood pressure was stable until wound closure. Four more units of platelets were transfused close to the end of surgery in order to minimize the amount or risk of postoperative oozing or hemorrhage. The operation took 30 min from skin incision to wound closure. The blood loss was estimated to be around 400 mL.

The pancytopenia improve after four more units of platelet and one unit of whole blood and her rate of recovery by repeat transfusion was significantly better than by transfusing before operation. Having received 2 U of blood, as suggested by the hematologist, the Hb increased from 10 $g \cdot dL^{-1}$. The other laboratory results at discharge were WCC 4.7 x $10^9 \cdot L^{-1}$, platelet count 8 x $10^9 \cdot L^{-1}$, albumin 25 $g \cdot L^{-1}$, AST 44 $U \cdot L^{-1}$, LDH 530 $U \cdot L^{-1}$. Though she had depressed platelet count again she could be managed and after an otherwise uneventful postoperative recovery she could be discharged on 9th postoperative day. In total 24 units of platelet (8 U before operation, 8 U immediate postoperative, 8 U in post operative period and five units of whole and packed blood was used in this case.

Her baby needed routine care of prematurity and IUGR including early fat restriced feeding and Vit. K injection and did well up to discharge.

Discussion

Hypoplastic anemia is characterized by diminished hematopoietic precursors in the bone marrow and results in a deficiency of circulating erythrocytes, granulocytes, and platelets. In aplastic anemia, the precursors are absent or severely reduced, Hb can easily drop to around 3 $g \cdot dL^{-1}$ and, in severe cases, the neutrophil and platelet count drop to <0.5 x $10^9 \cdot L^{-1}$ and <20 x $10^9 \cdot L^{-1}$ respectively. (1) The presented patient had a Hb level of 6.4 $g \cdot dL^{-1}$ on admission and probably already had hypoplastic anemia resulting in thrombocytopenia before pregnancy. Pregnancy seems to have a detrimental effect on the disease process (2). A parturient with hypoplastic or aplastic anemia is at increased risk of hemorrhage and infection and there are a number of case reports and reviews of aplastic anemia in pregnancy (2-4) It has a very variable clinical course and outcome, (5) and termination of pregnancy is usually advised for those with severe disease. There are case reports on successful management of pregnancy in bone marrow failure syndromes such as Diamond-Blackfan anemia and Shwachman-Diamond syndrome. (6) In these reports, the patients required Cesarean section, but none was reported in such refractory and low platelet count.

Close monitoring of hematological variables and appropriate replacement therapy is though important not always helpful. White cell transfusion may be required only for serious infection. Thrombocytopenia in hypoplastic anemia is treated with platelet transfusion, but repeated platelet transfusion may become ineffective due to platelet refractoriness and cross-immunization (3).

Since platelet transfusion is indicated for major surgery such as Cesarean section, 10 U of platelets were transfused and intraoperative blood loss was moderate (around 600 mL).

Thrombocytopenia has been regarded as a contraindication for neuraxial regional techniques and general anesthesia was the best option. The other platelet function tests include bleeding time, Bleeding time is however, highly operator-dependent, lacks sensitivity and specificity and has poor diagnostic value even when platelet function is altered (7). It is generally accepted that patients with a platelet count lower than 50 x $10^9 \cdot L^{-1}$ need platelet transfusion before major surgery (8). The same threshold is also recommended for patients with aplastic anemia and myelodysplasia (9). There are no agreed guidelines on dose of prophylactic platelet transfusion in thrombocytopenic patients. Studies on platelet dose for prevention of major bleeding complications are lacking (10). Norol et al. demonstrated the dose-effect of platelet transfusions and suggested a platelet dose of 1.5 U for each 10 kg body weight in patients without clinical factors favouring platelet consumption and receiving random platelet concentrates (11).

Thus 10 U of platelets were transfused before the start of surgery. But the instant case was found to be refractory to any amount of platelet transfusion with very low count and situation changed only after delivery of the fetus. There is no record till date of emergency cesarean section in such a situation in Aplastic anaemia.

In summary, we have presented a parturient with hypoplastic anemia and severe pre-eclampsia. A multi-disciplinary team approach involving the obstetrician, anesthesiologist, hematologist, and pediatrician was important. Cesarean section is possible in such a low platelet count under platelet and blood transfusion.

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