Cesarean section in a patient with sickle cell disease, mitral stenosis, pulmonary arterial hypertension, and thrombocytosis

Sir,

A 20-year-old primigravida of 32 weeks gestational age was admitted with a sickle cell crisis. She was a diagnosed case of rheumatic heart disease with moderate mitral stenosis (MS) during her second trimester and sickle cell disease (SCD). She had dyspnea on exertion during pregnancy. On examination, heart rate was 83 beats per minute, blood pressure 122/76 mmHg, 1st and 2nd heart sound and mid-diastolic murmur over the apical area was present. Two-dimensional echocardiography showed moderate MS, moderate mitral regurgitation, moderate pulmonary arterial hypertension (PAH), enlarged right atrium and right ventricle, and normal biventricular function.

Lower segment cesarean section was planned at the completion of 36 weeks of gestation. Preoperatively hemoglobin was 9.5 g/dL (12–15.8 g/dL), total leukocyte count was 22.06×10^9/L ($4–11\times10^9$/L), and platelet count (PC) was 790×10^9/L (normal $150–400\times10^9$/L), but coagulation profile was normal. In the operating room, the left radial arterial and the right internal jugular venous lines were inserted under local anesthesia. Her pre-operative vitals were within normal range. Oxygen supplementation was provided continuously. A 20G epidural catheter was inserted at the 3rd and 4th lumbar space, and local anesthetic was given in a graded manner. On delivery of baby, injection oxytocin 10 U was given intramuscularly and then continued as slow intravenous (IV) infusion. Apgar score of the baby was 10 at 5 min. She was hemodynamically stable throughout the surgery which lasted for 35 min. There was about 500 mL of blood loss, and total of 200 mL of IV fluid was infused.

Postoperatively, she was transferred to the intensive care unit for oxygen supplementation, epidural analgesia, monitoring of hemodynamics, pain crisis, or any thrombotic events. Pneumatic compression devices were used for thromboprophylaxis. She was maintained in positive fluid balance to prevent sickling crisis. The rest of her post-operative period was uneventful and discharged from the hospital after 10 days. Both the baby and mother were fine at 6 weeks of follow-up.

In pregnancy, there is an increased oxygen demand and decreased functional residual capacity of lungs. Hence, there is a risk of hypoxemia and sickling in pregnancy with SCD [1]. The aggravating factors for sickling such as dehydration, hypoxia, acidosis, and hypothermia should be avoided. The major causes of death are the acute chest syndrome, sepsis, and multi-organ failure [2]. In patients with MS, there is a risk of hemodynamic instability after spinal anesthesia, and in SCD patients, a risk of an acute chest syndrome after general anesthesia is present [3]. Hence, graded epidural anesthesia is preferred, and we administered this in our patient. Optimal hydration should be maintained to prevent sickling crises and acute pulmonary edema. Only 200 mL of IV was fluid infused intraoperatively in our patient.

PAH is defined as mean pulmonary artery pressure >25 mmHg at rest assessed by the right heart catheterization. Pregnancy with PAH is a very high-risk condition [4]. Hypoxia, acidosis, and hypercarbia should be avoided to prevent an increase in pulmonary vascular resistance. The PC remains in a lower side of the normal limit in pregnancy, but there is physiological hypercoagulable state due to an increase of coagulation factors. Thrombocytosis in our patient may be due to chronic inflammation (secondary) [2]. Thrombotic complications are more if PC $>1000\times10^9$/L. There is an increased risk of venous thromboembolism in pregnant patients with SCD than those without SCD [5]. Early mobilization and pneumatic compression devices were used to prevent venous thromboembolism.

Detailed knowledge about SCD, MS, PAH, and thrombocytosis in pregnant women and adequate perioperative care is needed for safe outcome in these patients.

Upendra Hansda, Chitta Ranjan Mohanty, Sangeeta Sahoo

Department of Trauma and Emergency, AIIMS, Bhubaneswar, Odisha, India

Correspondence to: Dr. Upendra Hansda, Assistant Professor, Department of Trauma and Emergency, All India Institute of Medical Sciences, Bhubaneswar – 751 019, Odisha, India.
E-mail: uhansdah@gmail.com

Received - 30 July 2018
Initial Review - 22 August 2018
Accepted - 18 September 2018

REFERENCES

1. Silva-Pinto AC, de Oliveira Domingues Ladeira S, Brunetta DM, De Santis GC, de Lucena Angulo I, Covas DT, *et al.* Sickle cell disease and pregnancy: Analysis of 34 patients followed at the regional blood center of ribeirão preto, Brazil. Rev Bras Hematol Hemoter 2014;36:329-33.
2. Oteng-Ntim E, Meeks D, Seed PT, Webster L, Howard J, Doyle P, *et al.* Adverse maternal and perinatal outcomes in pregnant women with sickle cell disease: Systematic review and meta-analysis. Blood 2015;125:3316-25.
3. Boga C, Ozdogu H. Pregnancy and sickle cell disease: A review of the current literature. Crit Rev Oncol Hematol 2016;98:364-74.
4. Chakravarty EF, Khanna D, Chung L. Pregnancy outcomes in systemic sclerosis, primary pulmonary hypertension, and sickle cell disease. Obstet Gynecol 2008;111:927-34.
5. Naik RP, Streiff MB, Lanzkron S. Sickle cell disease and venous thromboembolism: What the anticoagulation expert needs to know. J Thromb Thrombolysis 2013;35:352-8.

Funding: None; Conflict of Interest: None Stated.

How to cite this article: Hansda U, Mohanty CR, Sahoo S. Cesarean section in a patient with sickle cell disease, mitral stenosis, pulmonary arterial hypertension, and thrombocytosis. Indian J Case Reports. 2018;4(5):418.

Doi: 10.32677/IJCR.2018.v04.i05.031