Obstetric Medicine

Obstetric Medicine
2021, Vol. 14(2) 113–115

DOI: 10.1177/1753495X20936902
journals.sagepub.com/home/obm
SAGE

Adverse outcome of acute splenic sequestration crisis in pregnancy

Ann Kinga Malinowski[1,2,3], Jacqueline Parrish[1], Rohan D'Souza[1,2,3], Kevin HM Kuo[2,4], Richard Ward[2,4] and Nadine Shehata[2,5,6,7,8]

Abstract

Background: Acute splenic sequestration crisis, characterized by abrupt fall in hemoglobin, splenomegaly, hypovolemia, and often thrombocytopenia, occurs infrequently in adults with sickle cell disease and extremely rarely during pregnancy.

Case: A 25-year-old woman with HbSC presented at 33 weeks' gestation with vaso-occlusive pain. Sudden worsening of abdominal pain and non-reassuring fetal surveillance on day 3 of admission led to emergent delivery. Acute splenic sequestration crisis was the diagnosis of exclusion based on clinical presentation and intra-operative hemoglobin of 37 g/L. Five- and 10-minute Apgar scores were 4. Neonatal brain magnetic resonance imaging revealed significant diffuse white matter abnormalities.

Conclusion: Acute splenic sequestration crisis in pregnancy must be considered in the differential diagnosis for this patient population as it can evolve rapidly and lead to maternal and fetal compromise.

Keywords

Splenic sequestration, sickle cell disease, pregnancy complications, hematologic

Date Received: 20 January 2020; Revised 27 May 2020; accepted: 29 May 2020

Acute splenic sequestration crisis (ASSC) is a complication of sickle cell disease (SCD), whereby abrupt intra-splenic sickling entraps blood within the spleen, resulting in sudden and profound drop in circulating blood volume.[1] We describe a case of ASSC in the third trimester, with consequent neonatal white matter brain injury.

A 25-year-old G2P1 with HbSC presented at 33 weeks' gestation with a vaso-occlusive episode (VOE). Medical history revealed mild splenomegaly and chronic pain, with inconsistent hydroxyurea use and no chronic transfusion support. Baseline hemoglobin was ~100 g/L. Typical supportive measures were instituted and the pain gradually improved. On day 3 of admission, sudden severe abdominal pain was reported. Fetal cardiotocograph demonstrated abnormal fetal heart rate, with absent variability and shallow, unprovoked decelerations. During urgent transfer to Labor and Delivery, the pain became excruciating. The patient repeatedly declined phlebotomy attempts owing to her significant discomfort. In the absence of hemodynamic instability and persistent concerns about fetal well-being, blood work was deferred and caesarean delivery undertaken. A male infant was born, weighing 2380 g, with Apgar scores of 2 at 1 min and 4 at 5, 10 and 15 min, with poor tone and respiratory effort and absent suck, gag, and grasp reflexes. Intraoperative maternal hemoglobin was 37 g/L (no excessive blood loss); 1500 mL of crystalloid and two units of red blood cells (RBCs) were administered. Table 1 presents laboratory indices and vital signs.

Two days after delivery, the patient described worsening of her upper abdominal pain. She was also noted to have a fever, hypoxia, tachycardia, and tachypnoea; thus, an acute chest crisis was suspected and piperacillin-tazobactam commenced. Abdominal ultrasound revealed splenomegaly (spleen measuring 21 cm). Clinical improvement was noted the following day. Computerized tomography scan was negative for pulmonary embolism or parenchymal disease, leg Dopplers were negative for deep vein thrombosis, and maternal echocardiogram was normal.

Four days after delivery, the hemoglobin was 49 g/L and increased to 67 g/L after transfusion of one unit of RBCs. The patient continued to improve and was discharged in stable condition on day 11.

[1]Division of Maternal-Fetal Medicine, Department of Obstetrics & Gynaecology, Mount Sinai Hospital, Toronto, Canada
[2]Department of Medicine, University of Toronto, Toronto, Canada
[3]Lunenfeld-Tanenbaum Research Institute, Mount Sinai Hospital, Toronto, Canada
[4]Division of Haematology, Department of Medicine, University Health Network, Toronto, Canada
[5]Institute of Health Policy, Management, and Evaluation, University of Toronto, Toronto, Canada
[6]Division of Haematology, Department of Medicine, Mount Sinai Hospital, Toronto, Canada
[7]Division of Medical Oncology and Haematology, Department of Medicine, University Health Network, Toronto, Canada
[8]Department of Laboratory Medicine and Pathobiology, Mount Sinai Hospital, Toronto, Canada

Corresponding author:
Ann Kinga Malinowski, 700 University Ave, Rm 3-909, Toronto, ON M5G 1Z5, Canada.
Email: ann.malinowski@sinaihealthsystem.ca

Table 1. Summary of the evolution of laboratory indices during admission with eventual diagnosis of acute splenic sequestration crisis.

Parameters	AD0	AD2	AD3 Intra-Op (14:24)	AD3 POD 0 (16:07)	AD4 POD 1	AD7 POD 4	AD8 POD 5	D/C POD 11	Normal range
Red blood cell transfusion			2 Units			1 Unit			
Hemoglobin (g/L)	104	103	37	69	69	49	59	91	110–150
Reticulocytes ($\times 10^9$/L)	184		90	151	116	188	163	275	10–90
Platelets ($\times 10^9$/L)	151	147	73	137	85	74	92	217	130–400
Bilirubin (umol/L)	24			29	26			11	3–20
LDH (u/L)	207			743	1080	617	582	380	135–225

AD: admission day; D/C: discharge; Intra-Op: intra-operative; LDH: lactate dehydrogenase; POD: post-operative day.

An interval splenectomy was carried out, remote from this admission, according to the patient's wish, given the profound impact of ASSC and desire for future pregnancies.

The neonate underwent magnetic resonance imaging (MRI) on day 4 of life, revealing significant diffuse white matter abnormalities, extending into the optic tracts. MRI one month later demonstrated persistent white matter injury, though less pronounced than on original imaging. The parents were advised regarding ongoing concern for cognitive, motor, and speech development, potential for cerebral palsy, and risk of seizures. Neurodevelopmental follow-up at 13 months of post-term age demonstrated a normal neurological examination and normal growth. Communication skills met age expectations. Ongoing neurodevelopmental follow-up is in place.

In absence of active bleeding, the differential diagnosis of acute exacerbation of anemia in context of SCD includes hyperhemolysis, aplastic crisis, and ASSC.[2] ASSC is characterized by splenomegaly and hemoglobin decrease of 20 g/L or more, often accompanied by hypovolemia and thrombocytopenia,[1,2] with platelet count nadirs a mean of three days following admission;[3] consistent with our case. The pathogenesis of ASSC is unknown; however, its development is hypothesized to be initiated by a "triggering event," such as acute obstruction of venous flow from the spleen, with consequent sequestration of RBCs and platelets.[4]

While ASSC is well-documented and frequently diagnosed in young SCD patients,[1,3,4] with prevalence in children with SCD of 7–30%,[1] ASSC is often overlooked as a potential diagnosis in adults due to its rarity in post-adolescent years.[1,3] Its infrequency in adulthood is likely secondary to relative splenic fibrosis and auto-infarction, characteristic of the natural history of SCD.[4] Unsurprisingly, its frequency is relatively higher in adults with HbSC or HbS/Beta-thalassemia, as 50% have baseline splenomegaly.[3,4] There are only three published reports of ASSC in pregnancy; with a neonatal death in 1972, and no reported adverse fetal outcomes in the subsequent two cases.[5] Three further reports describe ASSC postpartum.[5,6]

Presenting symptoms may include abdominal pain, fever, tachycardia, and hypotension.[3] In adulthood, most patients first present with acute VOE.[3] Aside from a previous episode, there are no known predictors of ASSC[1]; however, contemporaneous symptoms such as fever or infection are seen in up to two-thirds of patients.[1] The "classic" presentation with fever and left-sided abdominal pain has been challenged by a report addressing ASSC in 16 adults with SCD,[3] in which only one patient presented with fever, while five others developed fever during admission.[3] Similarly, only 2 of 16 reported left-sided abdominal pain and only four described any abdominal pain, whereas the typical pain of VOE was far more frequently seen on initial presentation.[3]

Sequelae of ASSC may include hypovolemic shock and death, and efforts to promptly restore circulating blood volume are paramount.[1,3] The acute decrease in oxygenation associated with an abrupt and often profound hemoglobin drop has the potential to result in fetal death or fetal brain injury due to hypoxia,[4,5] particularly if maternal hemoglobin acutely falls below 60 g/L.[5]

Watchful waiting or RBC transfusion, aiming to promptly restore circulating blood volume and oxygenation, remain the standard for ASSC management,[1,5] and their choice depends on the degree of hemodynamic compromise.[7] Caution must be exercised with respect to transfusion, given risk of hyperviscosity, which may be encountered once sequestered RBCs re-enter the circulation.[7] Close collaboration of obstetric and hematology teams is paramount in these circumstances.

Recurrence rates of ASSC are high[1] and elective interval splenectomy is sometimes recommended for secondary prevention.[1,3] While splenectomy is typically suggested following multiple episodes,[1] given potential for significant maternal and fetal compromise, its consideration may be reasonable following a single episode of ASSC in pregnancy, particularly if future pregnancies are desired.

Our case illustrates the potential for rapid evolution of ASSC during pregnancy, leading to maternal and fetal compromise, which can advance despite the ready availability of expertise and resources, and despite quick actions on the part of the multidisciplinary team in a quaternary teaching hospital familiar with SCD in pregnancy. The case further highlights the necessity of retaining this potentially catastrophic complication high on the list of differential diagnoses when assessing an abrupt change in maternal or fetal clinical status of pregnant women with SCD, as prompt intervention is key to restoring maternal and fetal well-being.

Acknowledgements

None.

Declaration of conflicting interests

The author(s) declared the following potential conflicts of interest with respect to the research, authorship, and/or publication of this article: AKM: Lecture Honorarium – Alexion; Advisory Board – Alexion. KHMK: Grants/Research Support – Thalassemia Foundation Canada, Peter Munk Cardiac Centre, UofT, Cincinnati Children's Hospital Medical Center, Canadian Hematology Society, Pfizer; Honoraria: Alexion, Novartis; Consultancy – Agios, Alexion, Apellis, Aruvant, Bluebirdbio, Celgene, Novartis, Pfizer; Chair of DSMB: Bioverativ; Scientific collaboration: Abfero, Phoenicia Biosciences. RD: Grants/Speaking Honoraria: Ferring Canada. JP, NS, RW disclose no relevant conflict of interests.

Ethical approval

Mount Sinai Hospital does not require ethical approval for reporting individual cases or case series.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

Informed consent

Written informed consent was obtained from the patient for their anonymized information to be published in this article.

Guarantor

AKM is the guaranteeing author for the manuscript.

Contributorship

AKM, RD, KHMK, RW, and NS participated in the clinical care of the patient described; AKM and JP wrote the initial manuscript. AKM, JP, RD, KHMK, RW, and NS critically appraised and revised the manuscript.

ORCID iD

Ann Kinga Malinowski https://orcid.org/0000-0002-3466-7570

References

1. Brousse V, Buffet P and Rees D. The spleen and sickle cell disease: the sick(led) spleen. *Br J Haematol* 2014; 166: 165–176.
2. Ballas SK, Lieff S, Benjamin LJ, et al. Definitions of the phenotypic manifestations of sickle cell disease. *Am J Hematol* 2010; 85: 6–13.
3. Naymagon L, Pendurti G and Billett HH. Acute splenic sequestration crisis in adult sickle cell disease: a report of 16 cases. *Hemoglobin* 2015; 39: 375–379.
4. Solanki DL, Kletter GG and Castro O. Acute splenic sequestration crises in adults with sickle cell disease. *Am J Med* 1986; 80: 985–990.
5. Maia CB, Nomura RM, Igai AM, et al. Acute splenic sequestration in a pregnant woman with homozygous sickle-cell anemia. *Sao Paulo Med J* 2013; 131: 123–126.
6. Noreldeen SA, Oppenheimer C, Chapman C, et al. Postpartum acute splenic sequestration in sickle cell disease. *J Obstet Gynaecol* 2008; 28: 440–441.
7. Davis BA, Allard S, Qureshi A, et al. Guidelines on red cell transfusion in sickle cell disease part II: indications for transfusion. *Br J Haematol* 2017; 176: 192–209.