First Sickle Cell Crisis Postpartum in HbSB Thalassaemia

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BACKGROUND

HbSB thalassaemia refers to populations who carry both sickle cell trait and beta thalassaemia. This heterozygous condition falls in the spectrum of sickle cell disease. Sickle cell disease is the most common inherited autosomal recessive haemoglobinopathy, and increasing in prevalence due to global migration trends. A single amino acid change in the beta-globin subunit causes red blood cells (HbS) to polymerise and sickle in low oxygen states, resulting in haemolysis, vaso-occlusion and inflammation, ultimately causing complications like acute pain crises and end-organ damage.

CASE REPORT

A 30yo G2P1 had an emergency caesarean section at 40+1 weeks for obstructed labour on a background of previous caesarean section and HbSB thalassaemia. She had never had a sickle crisis and was on prophylactic clexane commenced by a haematologist. Intraoperative findings showed a partial dehiscence of the right uterine scar, and atonic uterus causing a 600ml postpartum haemorrhage. Day 1 post-operatively, the patient developed progressive bilateral shoulder, arm, and left knee pain. After exclusion of infection and venous thromboembolic disease, multidisciplinary management of the sickle crisis by anaesthetics, general medicine and haematology included analgesia, intravenous fluids, oxygen, prophylactic clexane, and monitoring for haemolysis. Her haemoglobin remained stable at ~90g/L and MRI showed bone marrow infarctions of the left humerus and femur (figure 1-2). Due to minimal improvement of her symptoms with supportive management, the patient began manual red blood cell exchange on day 8 as guided by the haematology team. She was eventually discharged on day 13 with ongoing prophylactic clexane.

DISCUSSION

Sickle cell disease in pregnancy is associated with increased risks of maternal and fetal morbidity and mortality, particularly preterm birth, fetal growth restriction, acute painful crises, and thromboembolic events[1-3]. Antenatally, women should be managed in a multidisciplinary team and counselled on complications, fetal monitoring, and avoidance of precipitating factors for pain crises including dehydration, hypoxia, and overexertion. Optimisation of organ function should be completed early antenatally, specifically cardiac, lung and kidney testing. Postnatally, women should receive 6 weeks of prophylactic anticoagulation. HbSB Thalassaemia and other heterozygous combinations are often overlooked and managed only as sickle cell trait. However as evidenced by this case, these patients can also develop acute sickle complications. Vaso-occlusive crises (VOC) are generally managed supportively with analgesia, and consideration of fluid and oxygen[3]. Blood transfusions are only indicated for severe anaemia, to reduce sickle cell complications and maintain oxygen supply to the fetus. Current guidelines do not support the use of prophylactic exchange blood transfusions in pregnancy, except in women with previous complications requiring transfusion and those who had a transfusion regimen pre-pregnancy[2].

Fig 1: T2 weighted image of humerus showing bone marrow oedema in the humeral shaft up to anatomical neck

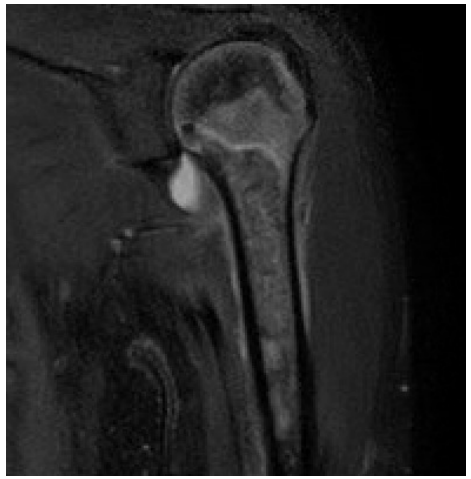
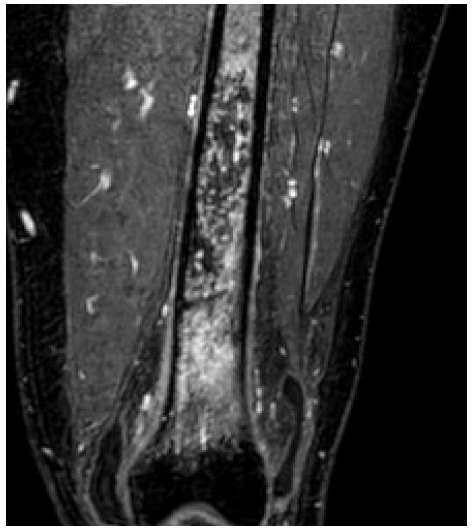


Fig 2: Gadolinium MRI of femur with features suggestive of bone marrow infarctions



Citations:
1.. Royal College of Obstetricians and Gynaecologists. Green Top Guideline No 61: Managing Sickle Cell Disease in Pregnancy. London: RCOG; 2011.
2. Society for Maternal-Fetal Medicine Consult Series #68: Sickle cell disease in pregnancy. Sinkey, Rachel G. et al. American Journal of Obstetrics & Gynecology, Volume 230, Issue 2, B17 - B40
3. Position statement on the management of pregnancy in sickle cell disease. Yue, M et al. Australian and New Zealand Journal of Obstetrics and Gynaecology, 2024 [Preprint]. doi:10.1111/ajo.13888.