Acute splenic sequestration crisis in an adult with sickle cell anaemia

Introduction

Sickle cell anaemia is the most common form of sickle cell disease. It is an autosomal recessive haemoglobinopathy characterized by homozygosity for a point mutation in the gene coding for the β-globin chain, creating haemoglobin S, leading to deformed and abnormally rigid erythrocytes. Subsequently the altered rheology leads to accumulation of these defective erythrocytes in capillaries causing a constellation of well-defined syndromes ('crises') in sufferers. Vaso-occlusive ('pain') crises are a common reason for hospital admission, usually requiring opiate analgesia in the acute phase. These may herald the onset of other sickle syndromes, in particular the chest syndrome, characterized by marked hypoxia, chest pain and bilateral lower zone infiltrates on chest X-ray.

Acute splenic sequestration crisis is characterized by engorgement of the spleen with blood, leading to a precipitous drop in haemoglobin concentration. This potentially fatal condition is rare among adult patients, as they undergo splenic autoinfarction during childhood. This article presents an unusual case of splenic sequestration and chest crisis in an adult woman homozygous for haemoglobin S (HbSS).

Discussion

The majority of homozygous (HbSS) paediatric patients progressively undergo recurrent splenic infarction. Adult sickle cell disease patients are therefore usually functionally asplenic and are prescribed phenoxymethylpenicillin prophylaxis against opportunistic infection by encapsulated organisms.

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Before splenic autoinfarction occurs, children with sickle cell anaemia are at risk of acute splenic sequestration crisis. This potentially life-threatening condition occurs when the spleen becomes acutely engorged with blood as intrasplenic sickling prevents venous drainage. Consequent erythrostasis can cause a precipitous drop in circulating haemoglobin level which may lead to hypovolaemic shock and ultimately, death (Michel et al, 1992; Koduri and Nathan, 2006). Other clinical features include the rapid development of tender splenomegaly, thrombocytopaenia (as a result of hypersplenism) and reticulocytosis. Patients with HbSS have little remaining distensible splenic tissue by adulthood and thus acute splenic sequestration crisis in adults with HbSS is exceedingly rare (Sarma, 1989; Moll and Orringer, 1996). In contrast, patients with other forms of sickle cell disease (such as HbSC or sickle/beta-thalassaemia) undergo less sickling and thus usually retain more splenic tissue into adulthood, and a number of cases have been described in adults with the HbSC genotype (Orringer et al, 1991;

Case Report

A 25-year-old Caribbean woman with known sickle cell anaemia (HbSS) presented with a 1-week history of lower back and limb pain. She had no history of chest crises nor any other major sequelae of her sickle cell disease. Aside from sickle cell disease she did not have any past medical history but had previously had recurrent painful crises for which she took hydroxyurea as preventative treatment. Her medications included regular folic acid and phenoxymethylpenicillin. She was afebrile and haemodynamically stable on examination, with oxygen saturations of 99% on air. Chest and abdominal examination were unremarkable. Her haemoglobin level was 11.3 g/dl. She was diagnosed with a vaso-occlusive crisis and treated with opiate analgesia, intravenous fluids, oxygen, oral co-amoxiclav and clarithromycin. Her pain settled after 5 days and she was discharged home.

She re-presented to the authors' service 3 days later with similar back and limb pain. She was also pyrexial (38.4°C) and tachycardic (102 bpm). Physical examination was again unremarkable. Her haemoglobin level was 10.2 g/dl, haemoglobin S 76.2% and platelets were 171×10^9/litre. She was commenced on a diamorphine patient-controlled analgesia pump and oral ciprofloxacin following growth of a sensitive strain of *Pseudomonas aeruginosa* on urine culture. Over the next 3 days her haemoglobin fell to a nadir of 5.1 g/dl with a corresponding decline in platelets to 56×10^9/litre. Three days later she continued to be pyrexial and reported some pleuritic chest pain. There were now fine bibasal crepitations on auscultation of the chest and an area of right lower zone consolidation on the chest radiograph. She was commenced on cefuroxime. The following day, she was noted to be tachycardic at 125 bpm and tachypnoeic at 25 breaths per minute. Examination of her abdomen revealed the acute development of palpable, hard, tender splenomegaly to the level of the umbilicus. Acute splenic sequestration syndrome was diagnosed and she received a 'top-up' transfusion of two units of packed red cells, increasing her haemoglobin level to 7.7 g/dl (*Figure 1*). Her spleen measured 17.3 cm on ultrasound.

A day later she became dyspnoeic and her oxygen saturations dropped to 90% on air, with a PaO_2 of 6.3 kPa. Examination of the chest now revealed bilateral coarse crepitations to the midzones, with bilateral lower zone infiltrates on chest radiograph. In view of this acute chest syndrome her antimicrobial treatment was changed to meropenem and she underwent an eight-unit autologous red-cell exchange transfusion, leading to resolution of her dyspnoea, a normalization of her blood count and a dramatic reduction in her analgesia requirement. Her haemoglobin level showed a rebound (to 14 g/dl) over the next few days and she required venesection to limit the risk of stroke. She was discharged on day 19 after her initial presentation at which stage her splenomegaly had settled.

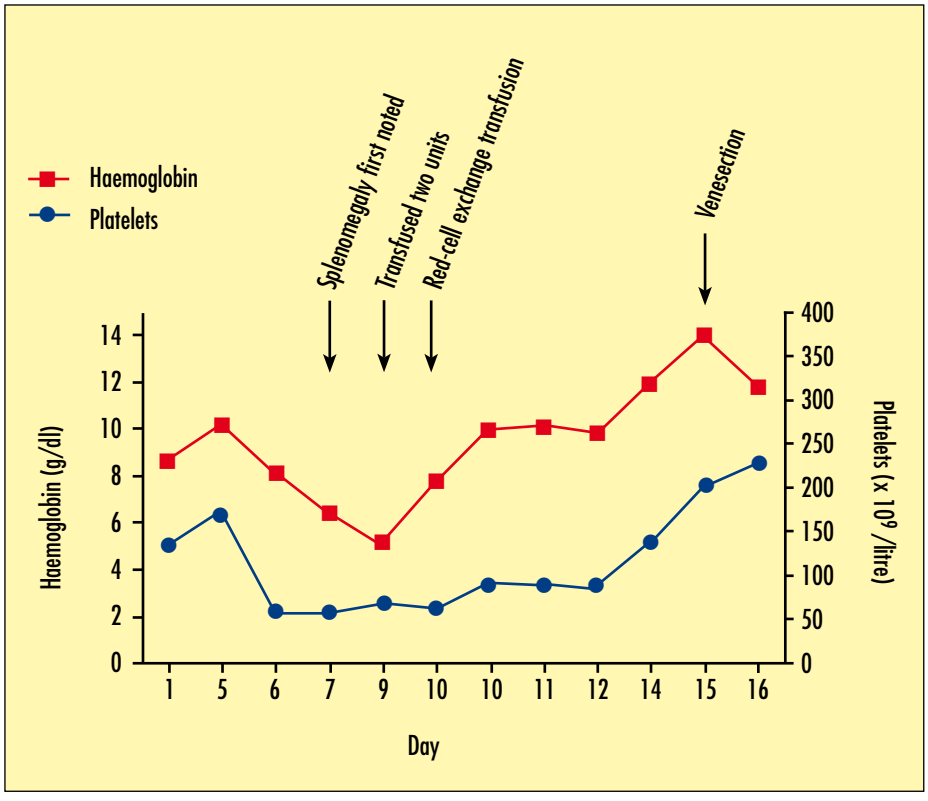

Figure 1. Changes in the patient's haemoglobin concentration and platelet counts during her admission. Two samples were taken on day 10 – one before and one after the red cell exchange transfusion.

Wang-Gillam et al, 2004; Rivera-Ruiz et al, 2008) and other heterozygous variants (Solanki et al, 1986; Koduri and Kovarik, 2006).

Treatment is focussed on replacing circulating volume with intravenous fluids and blood transfusion. The smaller circulating volume in paediatric patients renders them more susceptible to circulatory collapse following acute splenic sequestration crisis, with an estimated mortality of 12% (Topley et al, 1981). Owing to the rare nature of adult acute splenic sequestration crisis, the natural history of the condition has not been established, although serious morbidities and mortalities have been reported (Michel et al, 1992; Koduri and Nathan, 2006).

Patients being treated for acute splenic sequestration crisis can see large increases in haemoglobin concentrations to potentially dangerous levels, leading to increased risk of stroke. It is important to monitor for this complication and venesect as required to prevent further morbidity and mortality.

Conclusions

Acute splenic sequestration crisis in adult sickle cell disease patients represents an unusual but potentially life-threatening condition, emphasizing the importance of thorough physical examination in 'non-specifically' unwell patients. BJHM

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LEARNING POINTS

- Acute splenic sequestration crisis is common and potentially lethal in the paediatric population with sickle cell anaemia but can also occur in adult patients of all genotypes.
- Acute splenic sequestration crisis should be suspected when there is a rapid drop in haemoglobin accompanying the acute development of splenomegaly.
- Replacement of blood volume and exchange transfusion are cornerstones of management.
- Regular, thorough physical examination and close monitoring of haematological parameters are of paramount importance in patients with sickle cell disease.
- Haemoglobin levels may rebound to dangerously high levels during treatment; venesection is required to prevent the risk of stroke.