Impact Case Reports

Case Report

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Splenic rupture complicating sequestration crisis in a two-year-old with homozygous sickle-cell anaemia

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Abstract

Introduction: Splenic sequestration crisis (SSC) is a major cause of early mortality in sickle-cell disease, although rupture of the engorged spleen is rarely reported. Sickled erythrocytes obstruct splenic sinusoids, blood accumulates and intravascular volume collapses, with free intraperitoneal haemorrhage if the capsule fails.

Case report: A two-year-old boy with homozygous sickle-cell anaemia arrived after two weeks of fever, breathlessness, abdominal distension and pallor despite two transfusions. He was tachypnoeic at 66 breaths/min, tachycardic at 136 beats/min, and febrile at 38.2°C; the spleen projected 12cm below the costal margin and packed-cell volume was 14%. Abdominal paracentesis retrieved 250mL of fresh blood, indicating haemoperitoneum. Immediate transfusion, artemisinin therapy and supportive care led to a transient improvement. Within twenty-four hours oxygen saturation and blood pressure deteriorated; ultrasonography showed a ruptured spleen with free fluid. Emergency laparotomy revealed a macerated, actively bleeding spleen. Total splenectomy with two intra-operative transfusions controlled haemorrhage. Post-operative antibiotics, a pneumococcal conjugate vaccine and penicillin prophylaxis were commenced. The child left hospital on day seven with haemoglobin 10g/dL and remained well at two-week review.

Discussion: Rupture can complicate SSC before auto-infarction fibroses the spleen. Clinicians should suspect rupture when rapid splenic enlargement, precipitous anaemia and circulatory compromise coincide. Diagnostic paracentesis and bedside imaging can aid decision-making in resource limited settings, but survival depends on prompt transfusion and surgical control of bleeding. Multidisciplinary management, coupled with stringent post-splenectomy immunisation and parental education, should be standard when this rare complication is encountered.

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Introduction

Splenic sequestration crisis (SSC) is a potentially fatal complication of sickle-cell disease (SCD). Sickled erythrocytes obstruct splenic sinusoids; blood pools in the organ and haemoglobin falls by at least 2g /dL[1]. SSC chiefly affects children aged six months to five years, before progressive auto-infarction renders the spleen fibrotic[2]. In this age group it is the second most common cause of death after infection, with a fatality of at least 3.6%[3-5]. Splenic rupture during SSC is rare because most spleens have already lost function, yet rupture can precipitate sudden haemodynamic collapse[6]. Rapid identification and transfusion are therefore essential[7].

Case

A two-year-old boy, was transferred after two weeks of fever, breathlessness, abdominal distension and

marked pallor. He had already received two transfusions elsewhere without lasting benefit and had no family history of haemoglobinopathy or abdominal trauma. On arrival he was tachypnoeic at 66 breaths/min, tachycardic at 136 beats/min, pyrexial at 38.2°C and his oxygen saturation in air was 92%. The abdomen was tense, a tender liver lay 4cm below the right costal margin and the spleen extended 12cm below the left costal margin; blood pressure was 100/60mmHg. Packed-cell volume measured 14% and haemoglobin electrophoresis confirmed homozygous SCD. Paracentesis removed 250 mL of fresh blood, supporting a diagnosis of SSC complicated by haemoperitoneum.

Cross-matched red cells were transfused together with intravenous fluids, antipyretics, analgesics and artemisinin. Over the next twenty-four hours respiratory distress deepened, oxygen saturation fell despite supplemental oxygen and blood pressure declined. Ultrasonography demonstrated intraperitoneal blood arising from a ruptured spleen, prompting emergency laparotomy. The spleen was macerated and actively bleeding; total splenectomy and two intra-operative transfusions achieved haemostasis. Post-operatively he received broad-spectrum antibiotics, a pneumococcal conjugate vaccine and analgesia. By the seventh day haemoglobin and vital signs had stabilised and he was discharged on oral penicillin, folic acid and an outpatient appointment. At a fourteen-day review the wound was sound, haemoglobin was 10 g/dL and his parents were counselled about infection risk, vaccine boosters and early presentation for febrile illness.

Discussion

SSC may progress swiftly to hypovolaemic shock if unrecognised[2,8]. In the present child, splenic rupture converted a contained sequestration into free intraperitoneal bleeding. Although rupture is uncommon because infant spleens are still pliant, it can precede complete auto-infarction[6]. Early paracentesis provided diagnostic confirmation yet definitive control required splenectomy; published series advocate prompt surgical liaison whenever haemoperitoneum is suspected[6,8]. Where resources permit, partial splenectomy or splenic embolisation may preserve immune tissue, but total splenectomy remains safest when haemorrhage is brisk or facilities limited[9].

Management relied on coordinated action from emergency physicians, haematologists, surgeons and nursing staff. Such collaboration shortens decision times and optimises transfusion, antibiotic cover and peri-operative care[9]. Post-splenectomy, children with SCD remain vulnerable to encapsulated bacteria; pneumococcal, *Haemophilus* and meningococcal vaccination, penicillin prophylaxis and parental education reduce mortality[10]. Continuous surveillance detects late problems such as overwhelming post-splenectomy infection, chronic anaemia or vaso-occlusive crises[11].

Clinicians should palpate the spleen at each acute visit for SCD, monitor haemoglobin closely and treat any sudden fall as an emergency. Rapid imaging, transfusion and, when required, splenectomy improve survival. Close post-operative care, full immunisation and family instruction are essential. Research into biomarkers that predict impending sequestration, gene-therapy strategies to prevent recurrence and the role of partial splenectomy or embolisation in resource-limited settings is warranted.

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