Case Report

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Splenic rupture and intracranial haemorrhage in a haemophilic neonate: Case report and literature review

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ABSTRACT

Splenic rupture and intracranial haemorrhage are life-threatening conditions infrequently encountered in neonates without history of birth trauma. External manifestations of birth trauma; namely, capput succadeneum and cephalhematoma, when present raise suspicions for more serious intracranial or visceral damage. Rupture of normal spleen without an obvious source of trauma in haemophilic neonate is a rare event. The concurrence of both conditions and the unusual presentation make this case a rare one that is seldom encountered in the literature. Additionally, when splenic rupture occurs, the consensus is to employ all non-operative techniques aimed at salvaging the spleen, thus avoiding the immune-compromised state associated with splenectomy. However, in this case, we present a 3-day-old male with family history of haemophilia A, who was diagnosed with splenic rupture and bilateral subdural haematomas and underwent splenectomy, albeit with post-operative complications, in light of haemodynamic instability and high ongoing transfusion requirements.

Key words: Hemophilia, intracranial hemorrhage, neonate, splenic rupture

INTRODUCTION

Spontaneous rupture of the normal spleen is an extremely rare event.[1] Most cases of spontaneous splenic rupture occur in pathologic conditions that cause splenomegaly such as haematologic malignancies, infections, metabolic disorders, erythroblastosis fetalis and portal hypertension.[1,2] Rarer associations include primary splenic disorders or malignancies, and derangements in haemostasis.[1-3] In our patient, we presumed that unreported or unnoticed minor trauma was the likely cause acting in concert with the haemophilic state leading to haemorrhages. Delivery through the birth canal alone is enough trauma to cause a subcapsular haematoma initiating the rupture process, which manifests three days later.[4-6]

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CASE REPORT

A 3-day-old baby, born at term, to a 23-year-old primigravid mother, discharged home from labour and delivery on day 2, presented through the Emergency Department three days post delivery with severe pallor, lethargy, poor feeding and vomitting.

Birth weight and Apgar scores were 3.45 kg and 9, respectively. Prenatal course was devoid of maternal infections, drug use, trauma or chronic maternal disease. Labour lasted approximately 8 hours and was uneventful with spontaneous vaginally delivery and no concerns for dystocia. Family history is significant for haemophilia A in three first degree maternal uncles. At delivery PTT was >260 seconds, PT 11.1 seconds and INR 1.1.

On presentation, the patient was found to be extremely pale with cold extremities, poorly responsive with a feeble cry, weighing 3.4 kg, saturating 99% on room air, but no obvious cranial signs of birth trauma or of any external source of bleeding. Abdomen, however, was found to be distended and tender especially in the left lower quadrant. Laboratory work-up revealed haematocrit of 14%, with few schistocytes and tear drop cells, WBC 10,300, platelet count was 19,3000. Resuscitation was immediately started with packed red cell as well as Factor-VIII transfusions (FVIII) with monitoring in the intensive care unit. Computed tomography (CT) scan of the head, abdomen and pelvis showed bilateral subdural haematomas with intact ventricular system and no mass effect [Figure 1], and a large haemoperitoneum with fluid in the paracolic gutters, a spleen with unclear contours surrounded by

a large haematoma [Figure 2]. The diagnosis of splenic rupture and intracranial haemorrhage was made.

Following initial resuscitation, the infant showed clinical improvement with improved responsiveness and decreased pallor. However, haematocrit remained low at 25% despite repeated packed red blood cell (PRBC) and FVIII transfusions suggesting on-going haemorrhage. The patient's neurologic status remained stable and the small subdural haematomas were managed expectantly.

On day 2, the patient became paler, slightly jaundiced, with increased abdominal distension and tachycardia. Haematocrit had dropped to 21%. A decision was made to take the patient for exploratory laparotomy. On exploration, the abdomen was filled with about 500 mls of un-clotted blood, and a huge rupture through the corpus of the spleen involving the hilum was noted. Splenectomy was performed and no other abdominal visceral injuries were noted. Post-operatively, patient remained haemodynamically stable and was extubated a few hours later in the ICU. Haematocrit then was 27% and patient continued to receive FVIII transfusions, IVF hydration and antibiotics with nasogastric tube in place. Pathology confirmed the specimen to be normal splenic tissue.

Post-operative day (POD) 1, the patient remained haemodynamically stable, and passed adequate urine. Transfusions continued with F-VIII, PRBC and platelets with marked decline in PRBC transfusion requirements; haematocrit was 35%.

Upon consultation with neurosurgeon, no surgical management was indicated for the asymptomatic small subdural haematomas.

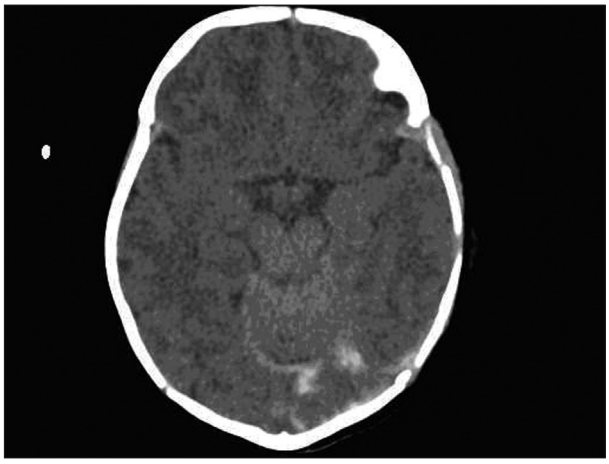


Figure 1: Computed tomography (CT) scan of the head showed bilateral subdural hematomas with intact ventricular system and no mass effect

POD 2, abdomen was found to be slightly distended but soft. Nasogastric tube remained in place. Abdominal X-ray revealed free intra-abdominal air consistent with normal post-operative changes, with no evidence of bowel obstruction. The nasogastric tube was removed and oral feeds were started on POD 3.

Patient's post-operative course was complicated by chylous ascites, haematuria and mild right hydronephrosis. The ascites was managed by nil per os status, total parenteral nutrition and percutaneous drainage with drainage catheter left in place. The haematuria and hydronephrosis resolved spontaneously. By POD 23, the ascites had resolved, the drainage catheter removed, and the patient tolerated feeds. Haematocrit had stabilized and no further transfusions were required.

Patient was discharged on POD 31 with arrangements made for FVIII supplement transfusions through home care services. The patient was followed in outpatient clinics and found to be doing well without neurologic sequelae.

DISCUSSION

While conservative, non-operative management of a ruptured spleen in a haemophiliac is feasible;[7,8,9] in the fact of haemodynamic instability and on-going haemorrhage, we believe splenectomy was the right approach. Some cases in the literature report initial good responses with non-surgical approach and transfusions alone, but with later recurrent haemorrhages and even death in some cases.[3,9,10] The transient renal disorder could be attributed to compression of renal structures by ascites, but the exact cause of the chylous ascites in

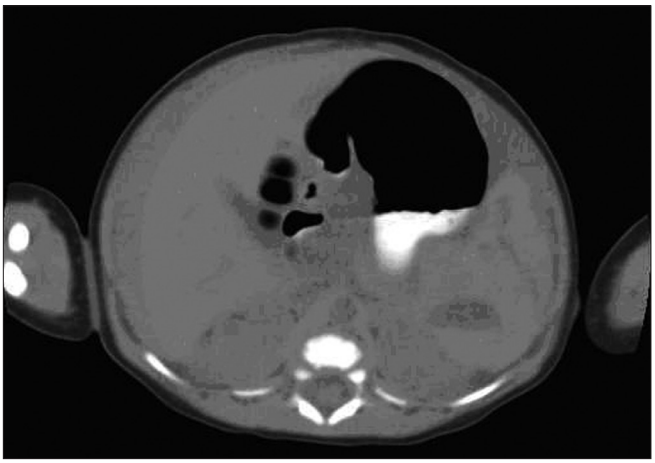


Figure 2: CT scan revealed large haemoperitoneum with fluid in the paracolic gutters, a spleen with unclear contours surrounded by a large hematoma

our patient remains obscure.

Intracranial haemorrhage in newborns, although recognized as the most serious bleeding complication occurs only in about 1-4% in haemophiliacs,[11-13] with at least 50% morbidity among survivors.[11] A more common presentation is bleeding at the umbilicus or post-circumcision.[1,13] Surgical management of subdural haemorrhages may be reserved for those associated with large haematomas, midline shifts or neurological deterioration.[12,14]

Thus, splenic rupture and intracranial haemorrhage may be unnoticed immediately following delivery. The presence of abdominal tenderness and distension, pallor and anaemia should alert the physician to the possibility of splenic rupture,[2,15] prompting early diagnosis using ultrasound, paracentesis, or CT scan if stable, and early intervention.

Perhaps, in known or suspected haemophiliacs, vigilant screening examinations, prophylactic factor replacements, shortly after birth may reduce the risk of haemorrhage,[13] consideration for elective Caesarean section should be given in such cases.

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