CASE REPORT

Perioperative complications in a patient with sickle cell disease following penetrating trauma

Thomas V. Divinagracia a, Tim A. Emhoff a, Suresh K. Agarwal a, Peter A. Burke a, Karen Quillen b, Erwin F. Hirsch a,*

a Department of Trauma Surgery, Boston Medical Center, Dowling 2 South, 840 Harrison Avenue, Boston 02118, United States
b Department of Hematology, Boston Medical Center, United States

Accepted 18 June 2007

A 26-year-old male with documented SCD (Hgb SS) presented to the Emergency Department with multiple gun shot wounds. The patient had bullet wounds in the left lower quadrant and the left flank as well as two wounds to the left upper extremity (with an obvious deformity) and a single wound to the right lower extremity (with the bullet palpable in the subcutaneous tissue). The patient was in distress, tachycardic, diaphoretic with a rigid abdomen. A chest roentegram was reported as being normal, following which, an exploratory laparotomy was performed. Findings at laparotomy were four perforating small bowel injuries with a small serosal tear in the left colon. A small bowel resection (including all four injuries and comprising of ~50 cm of proximal ileum) with primary anastomosis was performed. The colonic serosal injury was also oversewn. Total estimated blood loss was ~300 cm³. The patient was haemodynamically stable throughout the case with good urine output. An intraoperative arterial blood gas was performed and was unremarkable. An intraoperative CBC was notable for a leucocytosis of 24 K/UL and a Hct of 36%. This procedure was followed by a washout of the left upper extremity wound for a comminuted ulnar fracture.

The patient was extubated post op and transferred to the PACU where he remained stable. He was admitted to a regular surgical floor, where his post op care was relatively standard. He was placed on an isotonic fluid initially with a narcotic patient controlled analgesia (PCA) for pain. Over the first 12 h his post-operative course was uneventful. At the initial post op check the patient’s vital signs were stable. His pain score was noted to be 5/10. He had acceptable urine output and his nasogastric tube was functioning well. A post op CBC revealed a WBC of 24 K/UL and a Hct of 34%. Approximately 4 h later, the patient was assessed by the surgical team. He was, again, relatively comfortable (pain 5/10) with stable vitals. However, in approximately 6 h the patient was complaining of 10/10 incisional pain, as well as chest pain. He also had severe extremity wound pain. His blood pressure was recorded as 78/50 mmHg with a pulse of 145 bpm and he had a low-grade fever of 100.9 F. An ABG revealed a pH of 7.04, PCO₂ of 58 mmHg, PO₂ of 50 mmHg and a HCO₃ of 15 mmol/L. Haemoglobin...
Sickle cell disease is a hereditary haemoglobinopathy resulting from the inheritance of a mutant version of the β-globin gene on chromosome 11, the gene coding for the β-globin chains of the protein haemoglobin A (α2β2). Individuals with this mutation have resultant defective haemoglobin S. Patients with SCD typically are homozygous for the abnormal β-globin gene (α2β²) (although there are other genotypes such as SC disease and S-beta-thalassemia). Deoxygenation of haemoglobin S leads to intracellular polymerization and distortion of the red blood cell (RBC) into the classic sickle shape. The major consequence of this sickle shape is that RBCs become much less deformable and obstruct the microcirculation. Tissue hypoxia, which promotes further sickling, results. The clinical hallmark of this process is the vasoocclusive crisis, causing ischemic injury to the organ supplied. Pain is the most frequent complaint during these episodes, and it is ischemic in origin. This may essentially involve any organ, leading to a variety of systemic problems.

Some of the notable clinical sequelae seen in SCD relevant to this case are acute chest syndrome, myonecrosis and multisystem organ failure. Acute chest syndrome is an acute pneumonia-like complication of SCD, which is typically defined by fever, a new infiltrate on chest X-ray, along with chest pain, hypoxemia and respiratory distress. Precipitating factors for acute chest syndrome include infection, fat embolism and surgery. Although the involvement of muscle and fascia is not often recognized as a complication seen sickle cell crisis, myonecrosis (and myofibrosis) has been clearly documented in the literature. The pathogenesis is, again, rooted in vascular occlusion.

Acute multiorgan failure has been documented in the literature as a catastrophic complication of sickle cell crises. The hallmarks of this complication include pain, which is unusually severe for the patient, and rapid declines in the haemoglobin and platelet counts. Relevant to this case, there is an association with rhabdomyolysis. In a report that characterized acute multimodular organ failure seen in sickle cell patients, there were objective criteria given to define dysfunction of the pulmonary, hepatic and renal systems. Our patient clearly met these criteria for all three systems.

Our patient developed many associated complications of SCD during his prolonged hospital course. It is critical to consider what might have been done differently during the first 24 h of the patient’s course that may have prevented the development of some of these complications. Aggressive hydration and meticulous pain control in sickle cell patients are always paramount in preventing crisis. In reviewing the literature there is significant evidence to support the practice of simple perioperative transfusions in sickle cell patients undergoing elective abdominal procedures such as cholecystectomy or splenectomy. Although the performance of routine exchange transfusions for larger surgical procedures (such as hip arthroplasty) has been documented, this practice remains controver-
sial. There have been efforts in the literature to try to stratify surgical risk categories into low, medium and high (based on the nature of the procedure) in order to determine the subset of SCD patients who would more clearly benefit from preemptive transfusions.1

In dealing with trauma and emergency surgery in sickle cell patients there is a paucity of precedents or guidelines in the literature. In general, a multidisciplinary approach to these patients is critical.3 Certainly, earlier involvement of the haematologic team (with closer and more specific monitoring of haematologic parameters) should have been exercised. A more detailed perioperative discussion with the anaesthesia team with more aggressive pain control and closer monitoring of oxygenation and acid-base status in an ICU setting would also have been beneficial.2 Our patient required an extensive open abdominal procedure and suffered two significant extremity wounds. According to the risk stratification scheme mentioned above, he clearly would have been categorized as high risk. More aggressive blood transfusions in our patient early in perioperative period might have averted the development of some of the severe sickling complications from which he suffered. Lastly, exquisite haemostasis with proactive recognition of potential sites of haemorrhage in areas such as the retroperitoneum and all raw surfaces should have been exercised.

There have been small, randomized trials using inhaled nitric oxide (NO) in sickle cell patients. These studies are based on the principle that the vasoocclusive crises in sickle cell patients are associated with abnormal nitric oxide-dependent regulation of vascular tone.2 They showed improved pain control, fewer vasoocclusive crises and decreased length of stay in the NO treated patients (as compared to controls).8 This certainly has implications in the treatment of sickle cell patients in a variety of clinical circumstances. Indeed, this case highlights the need to reevaluate and update the armamentarium we, as surgeons, have in treating sickle cell patients—particularly in the setting of trauma and emergency surgery.

References