Single stage bilateral total knee arthroplasty in patient with G6PD deficiency

Sinukumar Bhaskaran, Hemant Wakankar

Abstract:
Glucose 6-phosphate dehydrogenase deficient patients when undergoing a major lower limb surgery would present with their own set of perioperative issues. A 74 years old male with bilateral osteoarthritis of knees underwent sequential bilateral total knee replacement at our institute. Inspite of avoiding all the offending drugs in this condition the patient still developed hemolysis. He recovered well with supportive management and further recovery was uneventful. We inferred that tourniquet or to a lesser extent the surgery itself might have acted as a stressor since all drug inducers were avoided.

Key words: Glucose 6-phosphate dehydrogenase deficiency, sequential bilateral total knee Arthroplasty, hemolysis.

Introduction
Glucose 6-phosphate dehydrogenase deficiency is the most common enzyme disorder worldwide presenting with either acute or chronic hemolysis. It is a X linked recessive disorder with the homozygotes being the more affected. There is a worldwide prevalence of 400 million distributed mainly around the middle eastern, south east asian and central asian countries (1).

The RBC’s depend solely upon this enzyme as a source of antioxidants. Any oxidative stress in G6PD-deficient patients can lead to hemoglobin denaturation and precipitation with subsequent acute hemolysis leading to anemia, jaundice, and reticulocytosis. If inadequately managed, patients may develop complications including acute renal failure, permanent neurologic damage, cardiovascular collapse, or even death (2).

Total knee arthroplasty is becoming an increasingly accepted procedure by patients for pain relief for severe arthritis of knees. Hence in the practice of arthroplasty one is going to be faced with cases of G6PD deficiency.

Case Report
A 74 years old male diagnosed with G6PD deficiency presented with bilateral end stage arthritis of his knees. He never had a hemolytic episode previously. His family physician had provided him with a list of medications which were to be avoided. At our centre we routinely practice single stage sequential bilateral total knee arthroplasty and patients have had satisfactory outcomes with minimal morbidity rates. After detailed deliberations with our anesthetist and physician we offered the patient a single stage bilateral total knee replacement. We decided to avoid drugs like paracetamol and tranexamic acid which were medications in our post operative protocol and additionally we also tried to avoid sulpha drugs and Furesemide if possible.

He was given a combined spinal epidural anaesthesia. The surgery was performed under tourniquet control. We used a cemented posterior stabilized implant. The total surgical time was 129 minutes and the tourniquet times were 51 and 46 minutes for the right and left sides respectively. The intraoperative blood loss of both the knees were about 300 cc. The wound was closed after ensuring adequate hemostasis over a suction drain. Post operatively we avoided Tranexamic acid as planned and the drains were left clamped to achieve a tamponade effect within the knee.

Two hours after shifting out the patient in the post op recovery ward the wounds began to ooze significantly on either side and his dressings were completely soaked. This prompted us to release his drains and apply a pressure dressing on both the sides. We attributed this soakage to the non usage of tranexamic acid. Within an hour his total drain volume from either side was 1000cc. His vital parameters remained within normal limits initially following which his hourly urine output began to drop. But all this while there was no sign of hematuria. A decision was made to transfuse him with RBC concentrate and shifting him to ICU for monitoring.

Overnight he developed hematuria and his laboratory investigations revealed a steep fall in the haemoglobin (14 gms% to 8.7gms% ).But we maintained adequate hydration and transfused him further 2 units of RBC concentrate over the next two days. Further investigations revealed an elevated indirect bilirubin count and increased reticulocyte count.
He responded to the supportive management, maintaining adequate volume and hydration, and his hemolysis gradually settled. The urine cleared out in the next two days and lab parameters returned to normal as well. He was shifted out of the ICU by the post operative day 5. His rehabilitation protocol proceeded normally and his operative sites healed well. His remaining post operative course though turbulent due to severe urinary tract infection and gastroenteritis from which he recovered, he did not have any further episodes of hemolysis. He developed UTI following the prolonged catheterization he required for monitoring hematuria. But he responded well to antibiotics and was discharged by the 12th post operative day. Post discharge he has resumed all his activities and is asymptomatic at 8 weeks.

**Discussion**

Apart from drugs, infection, tourniquet and surgical stress are also known to induce hemolysis in G6PD deficient patients. Inspite of avoiding all the potentially offending drugs in G6PD deficiency our patient developed hemolysis. There was an unknown factor that induced oxidative stress during the perioperative period leading to hemolysis.

Extensive research of the literature did not reveal any reports of total knee arthroplasty in G6PD deficiency patients. There were studies by anesthesiologists which concurred that drugs like lidocaine, midazolam and anesthetic gases could precipitate hemolysis (3). They advised that Prilocaine was a safer drug as compared to lignocaine. Our patient had received Bupivicaine for combined spinal epidural block which has not been mentioned in either of the studies.

It is known that use of tourniquet provokes a state of metabolic lactic acidosis (4) within the limb and this in turn could have triggered the precipitation and hemolysis cascade. Following the release of tourniquet a significant amount of acidic metabolic end-products are discharged into the systemic circulation. These in turn could have stimulated the precipitation-hemolysis cascade. Retrospectively evaluating the perioperative sequence of events this forms the most likely cause. The condition was self limiting as we know it is the aged RBC’s that are susceptible to this oxidative stresses and it resolves once the newly synthesized RBC’s are introduced into the circulation.

Surgery itself too could be incriminated as a potential stressor in these conditions and one cannot rule out its role in inducing hemolysis. Both the surgeon and patient would have to weigh the risk-benefit ratio in such instances.

We concurred that tourniquet was the most likely cause and in future to avoid its use in a similar scenario. We would also prefer to stage such a procedure considering the likely possibility of the surgery itself acting as a stress inducer.

**References**