Major Surgery in A Jehovah Witness with Sickle Cell Disease: Case Presentation

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Abstract

Introduction: A Jehovah’s Witness belongs to the religious group that does not accept blood transfusion in any form, while a sickle cell disease patient has abnormal haemoglobins that do not last in circulation predisposing one to anaemia and other systemic complications. Performing a major surgery in a Jehovah’s Witness who has sickle cell disease is tasking for a surgeon.

Case presentation: This case reports a 28-year-old African female with sickle cell disease who outrightly refused any form of blood transfusion as being a Jehovah’s Witness and having a complex primary hip that required total hip replacement. This work highlighted the complexity and difficulty encountered by virtue of the fact that patient had orthopaedic complications of Sickle Cell Disease and measures taken to prevent sickling crisis.

Conclusion: It is possible to carry out major surgery in a sickler who has durable power of attorney not to receive blood, but optimum preparation, meticulous and fast surgery and adequate monitoring must be instituted to avert morbidity and mortality seen in this group of patients.

Keywords: Jehovah’s Witness, Sickle cell disease, total hip arthroplasty.

Introduction

A major surgery is one that elicits a major fluid shift and causes a metabolic response to trauma. Most major surgeries will require blood transfusion following blood loss from the operation and so planning a major surgery in a Jehovah’s Witness (JW) with the comorbidity of sickle cell disease is a dilemma to the surgeon. Jehovah’s Witness are a religious group that do not accept blood transfusion in any manner even in life threatening situations. This belief is not borne out of their desire for martyrdom but faith which is tied to the expectation of eternal salvation [1]. Sickle cell disease (SCD) is an autosomal recessive disorder in which abnormal haemoglobin is formed which results in sickled red blood cells that can cause a variety of systemic complications among which is avascular necrosis of the head (AVN) of the femur. Management of AVN of femoral head can range from conservative treatment to different surgical modalities amongst which is total hip replacement (THR) for severe cases of AVN or where other modalities have failed. THR is considered a major orthopaedic surgical procedure in which mean blood loss has been reported to be above 1000 ml in different studies [2, 3].

This article reports a case of a patient with Sickle Cell Disease with severe AVN of head of femur who is a Jehovah’s Witness with a legal document refusing blood transfusions in any form and has need of a total hip replacement on affected hip.
Case presentation

A 28-year-old female nursing student who is a Jehovah’s Witness from Akwa Ibom state, Nigeria presented with a three year history of pain in the right hip which had been progressively worsening with noticeable shortening within one year duration. Pain was worse while walking and restricted home chores and school activities. She is a known SCD patient diagnosed in childhood who had never attended a sickle cell clinic and did not know her steady state haemoglobin. Her last crisis which was that of bone pains was ten years prior to admission. Examination showed an underweight patient with body mass index of 17.9 kg/m², with an anhalgic gait mobilizing using a walking stick. She had a tender right hip with hip flexion of 50 degrees, abduction and adduction of 10 degrees with zero internal rotation, external rotation and extension due to pain. Her Harris hip score was 53 and she had a limb length discrepancy of 2 cm. Her haemoglobin was 10 g/dL, ESR 45 mm/L hr, WBC 16000/mm³, neutrophils of 72%, lymphocytes of 28%. The liver function test, serum electrolytes, urea and creatinine, fasting blood sugar, urinalysis, were all within normal ranges. X-ray of the hip is as shown in (Fig. 1).

A diagnosis was made of avascular necrosis of the head of femur in a SCD and Jehovah’s Witness patient who will not accept blood in any form and had a durable power of attorney to the effect. She was thoroughly counseled and started on erythropoietin injections 4000 units three times a week prior to surgery. The anaesthetist decided on a blood conservation technique by way of acute hypervolemic hemodilution in theatre with general anaesthesia in order to achieve a close control of desired blood pressure using hypotensive anaesthesia. The blood pressure ranged between 85/50 mmHg and 100/60 mmHg from a baseline of 120/80 mmHg. Measures to avoid hypothermia, hypoxia, and detrimental hypotension were instituted to avoid complications of sickling. The lead surgeon was the most experienced and fastest surgeon. Diathermy and bone wax were used during surgery. Perioperative nurses and health attendants were also prepared to avoid undue delays in providing required materials and instruments. The radiographer was at hand to operate the image intensifier for locating the medullary canal. The duration of surgery was 4 hours 32 minutes with an estimated blood loss of 1200 ml with urine output of 1600 ml. Difficulty was encountered locating the medullary canal as following the neck cut, complete absence of cancellous bone was noted with only sclerotic bone seen and patient had femoral perforation. A size 6 stem was all the created canal could take and acetabulum received a size 48 mm cementless cup and neutral liner, the head size was 28+1.5. Bone graft from the head of femur was packed into acetabulum for the protrusio and two screws used for added stability to the shell. Post operative haemoglobin was 7.7 g/dL, ESR 35 mm/L hr, WBC of 16600/mm³, neutrophil of 66%, lymphocyte of 34% and the patient did not have any crises post operatively. She was placed on extended antibiotic prophylaxis for 10 days, commenced on antithrombotic day one post operative, pain control and haematinics. Her post operative X-rays were satisfactory (Fig. 2). She was allowed to sit up on post operative day one and mobilized on partial weight bearing on third day post operatively. She has had three months of follow up and is doing well with a Harris hip score of 89 at six weeks.

Discussion

Blood loss following total joint replacement can be substantial [4]. Performing a THR in a SCD patient with their inherent systemic problems, low haemoglobin and propensity for complications is daunting particularly in a patient refusing blood transfusion. It is a combination most surgeons will be unwilling to take on. It is very challenging and requires adequate perioperative preparations to avert morbidity and mortality, optimize red blood cell levels and reduce blood loss. Proper planning with the anesthetist and haematologist is crucial in the management of these patients. Various options proposed by different authors as blood conservation strategies in surgery include allogeneic blood programs, preadmission donation programs, techniques or pharmacologic agents that reduce blood loss, perioperative blood salvage, hemodilution techniques, bipolar sealers, use of perioperative recombinant erythropoietin to promote stimulation of bone marrow and increase red cell production [4, 5, 6, 7]. Of these options, the JW will only accept the use of erythropoietin, techniques or pharmacologic agents that reduce blood loss and hypervolemic hemodilution technique. They do not accept any form of autologous blood transfusion once the blood has left the body and is bagged.

In the preoperative work up, the haemoglobin level should be determined and optimized, clotting profile and bleeding disorders corrected and antithrombotic or antiplatelet drugs stopped if possible. Our patient had a haemoglobin of 10 grams and it has been reported that patients with 10-13.5 grams will have 40-60% chance of requiring blood transfusion with those less than 10 grams having a 90% chance [8]. As a SCD patient, they usually have a lower transfusion trigger than patients without and so are likely to cope with lower red cell volumes since the body has adapted to constant drops in red blood cell counts. It is important to know the steady state haemoglobin of SCD patients to know how much red cells are required to build up the patient preoperatively. Erythropoietin increases red cell production from the bone marrow and should be given weeks before intended surgery as it increases haemoglobin by 1 g/dL every 4-7 days [9]. Other means of building up the haemoglobin include administration of folic acid, Vitamin B12, B6, C, multivitamins and nutritional support.

Intraoperative measures to minimize blood loss include quick but
meticulous surgical technique and haemostasis. Diathermy should be available as well as topical adhesives like bone wax and oxidized cellulose. Use of tranexamic acid intraoperatively has been reported to reduce blood loss in THR [10] and so will be beneficial for JW patients but should be used with caution in SCD because of the thrombotic complications of tranexamic acid. The role of anesthesia is very vital in blood conservation and this includes measures to avoid hypothermia, hypoxia, and detrimental hypotension. Acute hypervolemic hemodilution is done in theatre to dilute the blood thereby reducing the number of red cells lost during the procedure. Hypotensive anaesthesia is done to achieve a close control of desired blood pressure. Hypoxia is prevented using 100% supplemental oxygen, intravenous fluids warmed before use to avoid hypothermia. These measures along with hypervolemic hemodilution, hypotensive anaesthesia, physical means of haemostasis, use of diathermy, use of bone wax and meticulous surgery were specific intraoperative steps to reduce blood loss in the patient. Post operative measures include steps to avoid infections and prompt treatment in case of an infection and in addition, extend prophylactic antibiotics as SCD patients are more prone to infections. Unnecessary phlebotomies should be avoided and monitoring carried for frank bleeding. Frequent movements which may dislodge clots and trigger off bleeding at the surgical site should be avoided during first 24 hours. Prophylaxis for deep vein thrombosis should be instituted with caution and patient should be adequately hydrated.

Our patient had femoral perforation while trying to locate the canal. Femoral perforation was reported is the most common complication in this group of patients [11] and use of guide wire in the medullary canal followed by enlargement using incrementally sized conical reamers have been advocated [11]. We did impaction bone grafting of the acetabulum using autologous graft from the head before a cementless cup was inserted. Autologous impaction bone grafting for protrusio acetabuli has been reported with good success [12].

Conclusion
Major surgery in a sickle cell disease patient who is a Jehovah's Witness requires exceptional perioperative preparation as surgery is expected to last long and may involve significant blood loss. Patients should not be rejected but adequately counseled, prepared and monitored. Difficulty in locating and reaming femoral canal should be anticipated and measures put in place to overcome this challenge. We recommend that staged surgery can also be done in the event where blood loss is beyond control.

Clinical Message
A Jehovah’s Witness with a durable power of attorney not to receive blood with sickle cell disease can safely undergo a major surgery like total hip replacement with meticulous planning and team management. Adequate peri-operative preparation, management and monitoring cannot be over emphasized.

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