



Research Paper

## Subarachnoid Block for Hip Hemiarthroplasty in Apatient with Sickle Cell Disease: a case Report

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**ABSTRACT:-** Sickle cell disease (SCD) is an inherited disorder of beta-chain of hemoglobin leading to multi system disease with life threatening complications like acute chest syndrome, stroke, blindness, osteonecrosis and end stage organ damage. Osteonecrosis of femoral head is a common musculoskeletal complication in SCD. SCD patients with orthopedic complications are at greater risk of developing stroke and acute pain crisis during perioperative period than individuals without this hematological disorder. Care during positioning, vigilant monitoring to avoid hypoxia, hypothermia, hypovolemia and acidosis followed by better postoperative analgesia, infection control and haematocrit management are mainstay in managing SCD patients posted for surgery. Here we report peri-anaesthesia management of 21 year male patient, who was diagnosed case of sickle cell disease having bilateral avascular necrosis (AVN) of femoral head and was posted for right hip hemiarthroplasty under subarachnoid block.

**KEYWORDS:-** Sickle cell disease, subarachnoid block, hip hemiarthroplasty

### I. INTRODUCTION

Sickle cell disease (SCD), a relatively common haemoglobinopathy is due to inheritance of mutated beta-globin gene on chromosome 11, forming haemoglobin S (HbS). HbS causes affected red blood cells to polymerize under condition of low oxygen tension which results into their sickling. Accumulation of sickle cells in circulation causes inflammation, endothelial damage and recurrent vaso-occlusion leading to ischemia of various body organs. [1] Ischemia of femoral head often leads to osteonecrosis requiring surgical correction. Stress of surgery, dehydration, acidosis, hypoxia, hypothermia and infection in perioperative period may trigger ischemic episode and sickle cell crisis. For SCD patients requiring hip replacement surgery, regional anaesthesia provides good muscle relaxation, post-operative analgesia along with prevention, identification and treatment of complications. Here we report, peri-anaesthesia management of a 21 year male SCD patient posted for right hip hemiarthroplasty under subarachnoid block.

### II. CASE REPORT

A 21 year male patient was presented with severe pain in both hip regions associated with difficulty in walking since 3 months. He was a diagnosed case of Sickle cell disease and was receiving treatment in form of analgesics and blood transfusions periodically. He was thin built, weighing 50kg and height of 164cm. His general examination and systemic examination findings were within normal range. He had restricted movements at both hip joints. Laboratory investigations revealed low haemoglobin (8.2gm %) and HbS concentration (92%). X-ray pelvis region and MRI of hip region showed bilateral femoral head avascular necrosis (AVN) with lateral subluxation and bilateral hip synovitis. He was posted for right hip hemiarthroplasty under subarachnoid block. His preoperative preparation included, improving his haemoglobin up to 10 gm%. He received 900ml of whole blood transfusion in preoperative period. HbS concentration after transfusion was 30%. Patient received vaccination against pneumococcus and H. influenza type B. To prevent megaloblastic erythropoiesis and hypocalcemia, patient received oral folic acid and calcium supplementation in perioperative period.

Procedure of anaesthesia and surgery and likely perioperative complications were explained to the patient. Written informed consent for anaesthesia and surgery was obtained from the patient and his relatives. Patient was kept nil by mouth (NBM) for 6 hours prior to surgery. To avoid dehydration in NBM period, he was administered maintenance intravenous fluid (dextrose normal saline - DNS) at the rate of 90ml/hr.

On the day of surgery, patient was re-assessed for any fresh complaints. NBM status was confirmed. All routine monitors like temperature, pulse oximeter, NIBP and ECG were attached and vital parameters recorded. His pulse rate, blood pressure and SPO<sub>2</sub> were 102/min, 110/70mmHg and 98% respectively. Intravenous line was setup on right forearm with 18G intracath. Antibiotic prophylaxis was given with inj. Ceftriaxone 1gm i.v., inj. Gentamycin 80mg i.v. and inj. Metrogyl 500mg i.v. Patient was premedicated with inj. Ondansetron 4mg i.v. Under all aseptic precautions and patient in sitting position, lumbar puncture was done in L<sub>4</sub>-L<sub>5</sub> interspinous space by midline approach, using 25G Quincke's Spinal Needle. After confirming clear and free flow of CSF, hyperbaric 0.5% Bupivacaine 2.0ml with Clonidine 30µg (0.2ml) injected into subarachnoid space. After confirming loss of pin-prick sensation and proprioception at T<sub>8</sub> dermatomal level, patient was given left lateral position.

To avoid pressure compression in lateral position, cotton padding was done at pressure points like shoulder, elbow, knee and ankle joints as well as in axilla and over chest and abdomen. Hypothermia was prevented by using warm blankets and fluid warmer. Oxygen supplementation (at 2 L/min) was given via nasal prongs. For conscious sedation, inj. Dexmedetomidine 25µg i.v. bolus followed by i.v. drip at the rate of 25µg/hr (0.5µg/kg/hr) intra-operatively was given.

During intraoperative period, pulse rate, oxygen saturation and systolic blood pressure ranged between 90-110/min, 95-98% and 90-110 mmHg respectively. There was no need of vasoconstrictors or atropine intra-operatively. Blood loss during surgery was around 500-600ml and was replaced with 600ml of whole blood. Patient received Ringer Lactate 1000ml, inj. Sodium bicarbonate 30ml in 500ml of Normal Saline. Towards the end of surgery, for postoperative analgesia inj. Diclofenac sodium 75mg slow i.v. bolus and inj. Tramadol 100mg i.v. in drip was given. Surgery lasted for 2 hrs and 45 min and then patient shifted to postoperative recovery room for further observation and management.

Postoperatively, Arterial Blood Gas (ABG) analysis showed, pH 7.38, bicarbonate level 22mmol/L and no correction of base deficit was needed. On second postoperative day, haemoglobin level was 9.4gm% and 300ml of whole blood transfusion was given. Care was continued in postoperative period, to maintain normothermia, oxygenation and proper positioning. For next 5 days, analgesia was provided with inj. Tramadol 50mg i.v. 8hrly and antibiotic prophylaxis was continued with inj. Ceftriaxone 1gm i.v. 12hrly. With early ambulation and physiotherapy, patient was discharged on 12<sup>th</sup> postoperative day. Then, he was called for follow up in Orthopedic OPD every 3 monthly.

### III. DISCUSSION

The HbS gene of SCD is found primarily in population of Africa and South-West Asia; but with fast moving population, it became a worldwide phenomenon.[2] Prevalence of sickle cell gene in Indian tribal population is nearly 1-40 %.[3]

SCD patients most commonly present with acute pain, chest syndrome, pulmonary hypertension, liver/renal cell ischemia, splenic sequestration, AVN hip, priapism and acute hemolytic or aplastic anemia. These complications contribute to significant morbidity and mortality in SCD patients. Due to vast variety of complications, persons with sickle cell disease often require surgical intervention for treatment and prevention of further complications.

In SCD patients, osteonecrosis of femoral head is, as a result of hyperplasia of bone marrow, sickling of red blood cells, increased blood viscosity, arterial occlusion and venous obstruction causing ischemia of femoral head due to stasis and lack of oxygenation. (4) Since the beginning of 20<sup>th</sup> century many surgical modalities like subtrochantric osteotomy or tantalum rod insertion were tried for managing osteonecrosis of femoral head. As one of the promising surgical modality to AVN hip joint, hip replacement arthroplasties were offered to SCD patients in last few decades.(5)

Perioperative risk is related to multiple factors like type of surgery, patient's general condition, disease activity and associated organ dysfunction. In 1995, Koshy *et al.* published a decade long retrospective review of 1079 procedures in SCD patients and showed 2.9% SCD specific complication rate in patients posted for hip surgery, mainly because of organ failure. (6)

Regional anaesthesia provides redistribution of blood flow and increase capillary and venous oxygen tension in blocked areas; but concomitant compensatory vasoconstriction in non-blocked areas may result in decreased venous oxygen tension exaggerated by hypotension and venous stasis resulting into sickling. Hence, careful monitoring to maintain the level of block and manage any signs of hypo-perfusion holds the key. Subarachnoid or spinal neuraxial regional anaesthesia fulfills the anesthetic requirements for hip replacement surgery which provides simplicity of technique, better muscle relaxation and postoperative analgesia by adding adjuvant to local anaesthetic. It decreases intra-op blood loss and decrease post-operative nausea, vomiting.(7) Epidural anaesthesia not only avoids severe hypotension by segmental blockage but also helps in post-operative pain management.(8) In this patient, we planned spinal anaesthesia for the surgery.

The subject of perioperative blood transfusion is controversial. Alone metacentric randomized controlled trial found conservative transfusion regimen as effective as aggressive transfusion regimen in preventing perioperative complications (9) and anaesthesiologist should be aware about risk of alloimmunization from blood transfusion. Transfusion aim should be to increase hematocrit to 30% than diluting hemoglobin S. Thereby; we maintained blood hemoglobin levels to above 10 gm% throughout perioperative period by multiple blood transfusions.

As acidosis fastens erythrocyte deformation on exposure to hypoxia, (10) bicarbonate alkalization was tried in this case to prevent post-operative sickle cell crisis. A sickle cell crisis is a painful episode that occurs due to deformed sickle-shaped RBCs blocking blood vessels, causing blood and oxygen deprivation. Although maintenance of adequate oxygenation is fundamental aim of anesthetic care in SCD, prolonged oxygen supplementation depresses erythropoiesis in a population of chronic hemolytic anaemia, whereas abrupt withdrawal of prolonged oxygenation has been reported to trigger vaso-occlusive crisis (VOC). (11) Hence, titrated well monitored dosing of anxiolytics and oxygenation is important to maintain saturation and avoid any sign of hypoxemia.

Exaggerated reflex vasoconstriction and shunting of blood from the bone marrow in response to skin cooling has been suggested to be the mechanism of hypothermia-induced VOC. (12) Therefore, maintenance of normothermia is the basis of care for SCD patients.

Anesthetist has to have vigilant approach to post-operative care by providing analgesia and facilitates early rehabilitation.

Greater the proportion of HbS in red blood cell, greater is the propensity to sickle thereby survival is rare beyond the fifth decade. [13] Thereby, prevention of such complications through early diagnosis by newborn screening and prophylactic antibiotics, hydration, pain management, disease modifying therapies holds the treatment part. Multidisciplinary advances in the management patients with SCD led to increased life expectancy in these individuals.

Advanced studies about knowledge of pathophysiology of sickle cell disease and newer modalities of treatment like hematopoietic cell transplantation continue to increase our understanding about this disease. But due to absence of definitive outcome data, the peri-anaesthesia management of these patients mainly depends on meticulous attention of hydration, oxygenation and analgesia and temperature control.

#### **Author contribution:**

AB: conduction of the case, manuscript writing and corresponding author

MP: senior anaesthetist, guide, manuscript editor

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