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ANAESTHETIC CHALLENGES IN A CASE OF A PATIENT WITH SICKLE CELL TRAIT WITH RELATIVE THROMBOCYTOPENIA POSTED FOR BIPOLAR HEMIARTHROPLASTY.

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ABSTRACT Sickle Cell Trait is a rare entity which poses a major risk perioperatively. There have been incidences showing the morbidity behind such traits. At times they pose a greater risk and threat to life. Patients often get to diagnosed only as an incidental finding. The primary goal and objective behind this study is to throw some light to the perioperative management for patients with sickle cell trait who for posted for various notable surgeries. The main aim for an anaesthetist is to prevent sickling crisis. The Strategies which are adopted by the anaesthetist should include right from an thorough preoperative assessment and to provide a safe and less complicated intraoperative period and an better postoperative care achieving a relatively pain free period.

KEYWORDS :

INTRODUCTION:

Sickle cell trait[1] is caused by abnormal hemoglobin called sickle hemoglobin or Hb S, secondary to a point mutation in the beta-globin chain[2]. This point mutation replaces A with T at codon 6 of the beta hemoglobin chain, causing the switch from glutamic acid to valine amino acid. The valine-type hemoglobin causes red cells to sickle when exposed to a low oxygen threshold.

Sickle cell trait is an inherited blood disorder that affects 1 million to 3 million Americans and 8 to 10 percent of African Americans. Sickle cell trait can also affect Hispanics, South Asians, Caucasians from southern Europe, and people from Middle Eastern countries.

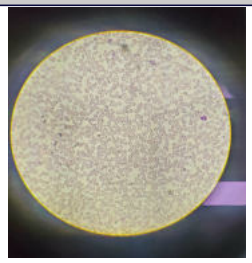
Most people with sickle cell trait have no symptoms and will not have any health complications. Occasionally people with sickle cell trait can have blood in their urine. Under extreme conditions such as high altitude, severe dehydration, or very high intensity physical activity, red cells can become deformed or sickled. Complications like Rhabdomyolysis, splenic ischemia/infarction, or Glaucoma[3]. Renal medullary carcinoma has been associated with sickle cell trait.

There is no sign of Vaso-occlusive crisis, unlike sickle cell disease. However, patients with sickle cell trait could have the same presentation as sickle cell anaemia if exposed to conditions that favour sickling, including severe hypoxia, dehydration, increased sympathetic outflow, hypothermia, hyperthermia, high 2,3-DPG levels, or released inflammatory cells. Decreased oxygen levels may cause red blood cells to transform from a normal disc shape to a sickle shape. As a result, the sickled cells begin to adhere to vascular walls, eventually blocking blood vessels in the muscles, kidneys, and other organs, which causes tissue death. Apart from the sickling of the cells, other cells interact to cause more adhesion of the red blood cells, including inflammatory cells and platelets. This could occur in multiple organs of the body and end up in constant ischemia.

Case Discussion:

A 75 year old female a came with complaints of alleged history of slip and fall at her residence and had injury over left hip, with difficulty in walking for 3 days, patient also gives history of bleeding gums for the past 1 month. She was then diagnosed to have left intertrochanteric fracture. She is also a known case of Coronary artery disease on regular medications and also a known wheezer on Inhaled Bronchodilators. Now patient has come to the Pre-Anaesthetic check up for undergoing left Bipolar hemiarthroplasty.

On general examination, patient was thin built, cachexic with a stable hemodynamics. Systemic examination were within normal limits. Upon airway examination her Mallampati grade was III and there were no restriction in neck movements with adequate mouth opening, Buck tooth present and partially edentulous.



(Peripheral Smear Study Showing Sickle Cells And Normal Cells)

Complete hemogram showed Hb-13.4%, TC- 16,330, Platelets-51,000.ECG showed left bundle branch block and AV block. 2D-Echo showed EF-60%, aortic valve sclerosis, grade-I Diastolic Dysfunction. Cardiologist advised moderate risk for non cardiac surgeries with a Major Adverse Cardiac Events intra-operatively. Physician advised moderate risk only if platelets counts>1,00,000 after adequate platelet transfusion[4].As per ESRA guidelines the minimum platelet count for performing spinal anaesthesia is 80,000[5].Preoperatively one pint of Single Donor Platelet was transfused and the platelet count on the day of surgery was 94,000. Since it was a quasi emergency and also the platelets were more than 80,000 patient was taken up under ASA-IIIE for the procedure.

High risk ,ICU and post-operative ventilator support were explained to the patient and the attendees. After ensuring Adequate blood and blood products reservation, patient was shifted to the Operating room. All ASA Standard monitors were connected and baseline vitals were recorded. Temperature monitoring was done as well. In view of difficulty in positioning for the subarachnoid block, a peripheral nerve block was performed.

Patient in supine position, under sterile precautions, ultrasound guided left femoral nerve block performed with 20ml of Inj.0.25% Bupivacaine with 4mg of Inj. Dexamethasone. Patient was then shifted to the operating table and in sitting position, Under sterile precautions, subarachnoid blockade at L3-L4 space done using 25G Quincke's needle with 10mg of Inj.0.5% heavy Bupivacaine + 25mcg of Inj. Fentanyl and level was fixed at T8.Patient was then shifted to Lateral position for the procedure with adequate pressure points coverage. After 20 mins of incision, patient started having fall in BP, which was initially managed with crystalloids. The further fall in BP was managed with vasopressor like Inj.Phenylephrine[6] 30mcg IV bolus followed by infusion. Patient was able to hold a mean arterial pressures of 75-80mmhg. The other hemodynamic parameters were found to be within normal limits.No other intraoperative events were noted. Total blood loss was 380ml and urine output was 200ml. Multi-modal analgesia was preferred for post-operative pain. Patient was shifted to

ICU for observation with stable hemodynamics. Repeat Hb-10.1 and platelets 37,000 following 4 random donor platelets transfusion. On post-op day1 ,CBC showed HB-8.8 , Platelets 45,000and TC - 17,680.Hematology opinion advised for repeat peripheral smear, CRP, ESR, LDH, and D-Dimers. One pint of packed red cells was transfused.

Repeat platelets on 2nd post-op day was found to be 19,000. Hematology opinion sought and advised for four fresh frozen plasma transfusion[7] and repeat platelet was found to be 39,000. Patient was symptomatically better and platelets were in raising trend, patient was shifted out of ICU.

Hemoglobin electrophoresis showed sickle cell trait. Patient was discharged on 9th post-op day. Repeat platelet at the time of discharge was 76,500.

DISCUSSION :

Sickle cell trait cases being posted for any surgery should involve a proper pre-operative evaluation including the possible causes of the current sickle cell trait and the confirmatory tests that are being deployed for the same, ensuring adequate reservation of Blood and blood products, adequate fluid therapy , preventing and managing intraoperative hypotension, providing normothermic environment, Adequate analgesics for mitigating pain involved in the surgery.

The plan of anaesthesia should be regional wherever applicable. Femoral nerve blocks or Fascia Iliaca block for positional pain so as to enhance the patient to sit for the Spinal Anaesthesia [8] and can be preferred even in thrombocytopenia cases as they are superficial blocks.

The notable triggers for sickling crisis would be hypoxia, hypothermia, hypovolemia, hypotension and the worsened cardiac status would further more complicate the outcomes.

Hypothermia was prevented by proper warming devices and blankets to avoid cold peripheries. Warm fluids should be given to maintain normothermia.

The most common trigger intra-operatively is hypoxia which should be given an special emphasis. It is essential Maintain the desired Fio_2in order to prevent hypoxia if general anaesthesia is to be considered. Regional techniques however would warrant minimal oxygen in order to avoid hypoxia.

Blood loss can also stimulate an adverse crisis in these cases owing to the considerable volume loss. It is always better to replace the lost blood with only blood products like packed red cells, platelet concentrates, fresh frozen plasma. Single donor platelet transfusion have shown better results in improving platelets[9].As per ESRA guidelines minimal platelets for spinal anaesthesia would be 80,000cells/mm^3 and for peripheral nerve blocks 50,000cells/mm^3[10].

The second most common factor for sickling which is hypotension which can be managed with crystalloids and colloids depending on the volume loss. Vasopressors like Phenylephrine causes rise in cardiac output by raising preload . Other Vasopressors like Noradrenaline, vasopressin can also be considered if persistent hypotension.

Dehydration has to be prevented by adequate fluid management and blood products in order to avoid an adverse sickling crisis. Post-operative period should also aim at providing a multimodal analgesia and prompt fluid therapy. Proper monitoring in the first few postoperative days is essential. Blood and blood products if deranged hemogram persists.

CONCLUSION:

The main aim of an anaesthetist in a case of sickling trait is to avoid sickling crisis. Since most of the time sickle cell trait is a hidden factor, careful pre-operative evaluation and intraoperative planning is essential. Regional anaesthesia offers more safety of margin as compared to General anaesthesia wherever applicable.

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