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Research Article

Anesthetic management of a patient with thalassemia major for a deformity correction of upper limb

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ABSTRACT
Thalassemia is a group of hereditary disorder which causes the body to produce less hemoglobin. ß thalassemia major is a severe variant requiring frequent blood transfusion and have perioperative implications due to iron overload, extramedullary hematopoiesis and multiple organ involvement. We report an 18-year-old female, a diagnosed case of ß thalassemia major, posted for radial corrective osteotomy and ulna plating. She was receiving packed blood cell transfusion every two weeks and on oral deferasirox and intravenous deferoxamine for 6 days in a week after every transfusion. She had a past history of ASD device closure. Investigations revealed normal ferritin levels, normal iron and total iron binding capacity. Recent echocardiography showed normal left ventricular function with no residual shunt across the device. Oral deferasirox was continued till the day of surgery. She was anethetised with ultrasound guided right brachial plexus block with 15ml 0.5% bupivacaine through supraclavicular approach and 10 ml 0.5% bupivacaine through interscalene approach. Procedure was uneventful with minimum blood loss. There have been only few reports of ß thalassemia major patients with extramedullary hematopoiesis with bone deformity for corrective surgery. Hence this article is presented to give an overview of managing such patients under regional anesthesia technique.

INTRODUCTION:

Thalassemia is a group of hereditary disorder that causes the body to produce less hemoglobin, a protein in red blood cells that helps in carrying oxygen to all the cells in the body. It is classified into two major categories – alpha and beta thalassemia. Alpha thalassemia is caused by missing or altered genes responsible for making the alpha-protein chains of haemoglobin, which is located on chromosome 16. Beta thalassemia is caused by gene mutation affecting the beta protein chains of haemoglobin. Beta thalassemia is again classified into major, minor, intermedia based on the severity of symptoms.

Thalassemia major has two mutated genes, which causes severe anemia in early stages of life, requiring multiple blood transfusions[1,2]. Patients with thalassemia major have perioperative implications because of chronic hemolytic anemia, iron overload, extramedullary hematopoiesis and multiple organ involvement. Around 15million people are affected by thalassemia worldwide. The major cause of mortality in thalassemia major is cardiovascular complications related to iron overload. With multiple blood transfusions, iron accumulates in the body (hemosiderosis). Iron gets deposited in cardiac muscle causes iron induced cardiomyopathy, which results in congestive cardiac failure, arrhythmias, and sudden cardiac arrest[3]. But clinical outcome has become better over the few decades because of blood transfusion, iron chelation therapy, dedicated centers to treat such patients and awareness about the treatment. The anesthetic management of patient with thalassemia major is challenging, which requires coordinated efforts of hematologist, anesthesiologist and cardiologist, to balance the thrombotic and bleeding effects in a chronic anemic patient[4,5]. We describe a case of thalassemia major with hand anomaly (bilateral Madelung deformity), posted for right distal radius corrective osteotomy and ulna plating.

Case description:

A 18yr old female weighing 41kg, a diagnosed case of thalassemia major presented with chief complaints of bilateral wrist pain, inability to flex the wrist. She was a diagnosed case of thalassemia major from the age of 3 years, receives one packed blood cell every 15days at a dedicated thalassemia and sickle center near her native place. Also, she was on oral deferasirox and intravenous deferoxamine for 6

days in a week after every transfusion. Past history of atrial septal defect closure was done at the age of 5years. Radiology of her both wrists revealed Madelung deformity and then she was posted for distal corrective osteotomy and ulna plating of her right hand. She visited our hospital on the second day of last blood transfusion.

On admission, her hemoglobin was 11.1 gm/dL, serum ferritin was 628 ng/mL, serum TIBC (total iron binding capacity) was 364 µg/dL, serum iron was 113 µg/dL, coagulation profile normal, total bilirubin was 1 mg/dL. Xray of the right wrist showed uneven growth in the wrist with shortened radius and overgrown ulna (Madelung deformity). Chest x ray showed mild cardiomegaly with normal lung fields. Echocardiography showed normal left ventricular function with no residual shunt across the ASD device. Her effort tolerance was adequate. She was posted for distal radius corrective osteotomy and ulna plating under American society of Anesthesiologist grade 2. Her blood group was B positive and one unit of packed red blood cell was reserved. She was kept nil per oral 6hours before for solids, as per ASA guidelines. After shifting to operation theatre, peripheral venous access was secured with 18G iv cannula. Standard monitors – ECG, non-invasive blood pressure (NIBP), saturation probe (SPO_2) was attached. Under sterile aseptic precautions, ultrasound guided supraclavicular block was given using 15ml 0.5% bupivacaine and interscalene block with 10ml 0.5% bupivacaine. After checking the adequacy of block, tourniquet was applied to her right arm and the procedure was done uneventfully without any hemodynamic changes. After the procedure was over, she was shifted to postoperative icu and the orals were allowed after 2hours of shifting. Postoperative analgesia was given with intravenous paracetamol 1g. oral deferasirox was continued from the first postoperative day.

DISCUSSION:

Madelung deformity is a rare wrist condition caused by abnormal growth or premature closure of the inner, lower (volar-ulnar) portion of the radius growth plate in the forearm. This uneven growth leads to a shortened radius, an overgrowing ulna that protrudes at the back of the wrist and a V-shaped misalignment of the carpal bones. Even though it is congenital, it becomes noticeable during the early adolescent age. It occurs more commonly in female than male. But in thalassemia, deformity occurs as a secondary skeletal complication of iron chelation therapy and premature closure of growth plates. That is why, the deformity in thalassemia patients is often being referred as pseudo-Madelung deformity. Chronic anaemia causes bone marrow expansion, which thins out the cortical bone, weakens the metaphyseal region near the wrist joint, making the epiphyseal growth plate to close prematurely [6,7,8]. Common symptoms are pain, decreased wrist grip and restricted range of motion in the wrist. Initially these symptoms will be treated by adjusting the chelation doses. If the pain persists with severe restricted wrist movements, then these patients need corrective osteotomy of radius. Our patient also admitted with similar complaints of severe wrist pain and restricted wrist movement, requiring corrective osteotomy of radius with ulna plating.

Patients with beta thalassemia major receive regular blood transfusion to maintain adequate haemoglobin levels and will be on oral or intravenous chelation therapy to reduce the iron load in the blood. Our patient also was receiving packed blood cell transfusion every 15days, on intravenous deferoxamine for 6days after the blood transfusion and oral deferasirox for 6days in a week, which was continued till the day of surgery. There are no specific guidelines about when to accept the patient for surgery after the blood transfusion. Our patient had a blood transfusion 2 days before the admission, her hemoglobin was 11.1 gm/dL, normal iron profile and the proposed surgery was an intermediate risk surgery, so we accepted her for surgery. There are no specific recommendations to stop the oral iron chelating agents before surgery. So, we asked her to continue the drugs till the previous day of surgery. Chelating agents can cause transient abnormal liver function tests and transient increase in serum creatinine[9,10,11], but our patient had normal liver function test values and normal serum creatinine. Beta thalassemia major patients very rarely are associated with congenital cardiac anomalies. They generally may develop iron induce cardiomyopathy due to repeated blood transfusions. Our patient had atrial sepal defect, for which she underwent device closure at the age of 5years. Recent transthoracic echocardiography showed no shunt across the ASD device and normal left ventricular function. Prolonged oral chelation therapy can cause neutropenia, making them prone for infection. Our patient showed total count of 5100 cells/cu mm with 42%

neutrophils,40%lymphocytes, eosinophils 10%, monocytes 08%. Her coagulation status was normal. So, we decided to proceed for surgery under regional anesthesia.

Regional anesthesia is preferred over general anesthesia in thalassemia major patients so as to avoid hemodynamic instability associated with general anesthesia[12,13]. Mild iron induced cardiomyopathy can exaggerate this hemodynamic instability. Chronic bone marrow expansion can cause extramedullary hematopoiesis, which can cause craniofacial abnormalities with prominent maxilla and prominent upper teeth, which can make the airway difficult to access. Our patient had slightly prominent upper teeth.

We proceeded with ultrasound guided supraclavicular and interscalene block as surgeons did the procedure under tourniquet. Even though the hemoglobin was 11.1 gm/dL, we reserved one unit of packed blood cells. We maintained adequate hydration by giving ringer lactate solution up to 1.5litre. Blood loss was minimal and there was no hemodynamic instability in the intraoperative period. After the procedure got over, she was shifted to icu and the postoperative analgesia was maintained with intravenous paracetamol. We avoided NSAIDs as oral iron chelating agents are slightly nephrotoxic.

CONCLUSION

Perioperative management of patients with thalassemia major is challenging because of the nature of the disease, effects of repeated blood transfusions and also the effects of oral chelating agents. In this case, surgical intervention under ultrasound guided regional anesthesia has improved the patient's quality of life with respect to hand movement and with adequate postoperative analgesia. Therefore, a multidisciplinary approach is required to assess the patient's physical condition, to plan the perioperative period and to achieve best possible outcome in these patients.

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