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Case Report

# Perioperative management of thalassemia intermedia patient posted for major spine surgery – A case report

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ABSTRACT

Thalassemia patients are known to have abnormal haemoglobin structure and function because of genetic suppression of chains in haemoglobin molecule. These tend to lead to abnormal coagulation pathway and can cause excessive bleeding in the intra operative period. Hence, thalassemia patients are difficult and often considered as high-risk patients. Vigilant monitoring, early diagnosis and prompt treatment plays a vital role in the management and improving patient outcomes. This case illustrates a successful T4-T8 laminectomy and excision of extradural lesion in a patient with Thalassemia intermedia, pointing out the pathophysiologic considerations and discussing the means to reduce the perioperative risk. There have only been a few reports in the literature for the same and hence this article is intended to provide an overview of the anaesthetic management for a patient with thalassemia, focussing specifically on the blood conservation strategy for major spine surgeries. The particularities described in this case report may help other anaesthesiologists choose the best strategy when facing challenging patients similar to the one described.



## 1. Introduction

Thalassemia is an autosomal recessive inheritance, is characterized by haemolytic anaemia caused by partial or complete deficiency of globulin chains compromising haemoglobin tetramer thereby affecting the function.[1] Globally, fifteen million people are affected by thalassemia.[2] In thalassaemia intermedia, there is a milder anaemia and the need for blood transfusion usually arises in the long term. Although the major cause of mortality is still cardiovascular complications in patients with thalassaemia, thromboembolic events secondary to a hypercoagulable state comprise an important cause of mortality, especially in patients with thalassaemia intermedia.[1] Clinical outcomes have improved due to advancement of treatment by means of blood transfusion and iron chelation therapy. The anaesthetic management of patients with thalassemia intermedia challenges clinicians and requires coordinated efforts of the haematologist, cardiologist and anaesthesiologist in order to balance between thrombo prophylaxis as well as risk of bleeding in an already anaemic patient. The level of preoperative preparation must consider both the surgical risk and the impact of the disease. We describe a case of a low dorsal spinal cord compression at level between T4-T8 posted for decompression surgery.

## 2. Case Description

A 54-year old male weighing 70 kg, suffering from thalassemia intermedia since the age of 24 presented with chief complaints of bilateral lower limb weakness along with decreased sensation below the chest for one and a half months. His past medical history revealed 4 blood

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transfusions till date and had also underwent Splenectomy 30 years back. On admission his haemoglobin (Hb) was 8.6 g/dl and platelet count was 5,40,000/cm3. On examination, he had frontal and maxillary bossing but no significant mandibular enlargement. Airway assessment showed 3 finger mouth opening and Mallampatti Grade 2 with short neck and crowded teeth. He was pale, afebrile with normal vitals and normal cardiovascular and respiratory system. His baseline oxygen saturation (SPo2) was 96% on room air. Systemic examination revealed severe spasticity of both his lower limbs along with numbness from T4 till the metatarsals. No other skeletal abnormalities were evident. In view of limited mobility on daily activities he was started on anticoagulant therapy and anti-embolic stockings. All routine investigations, serum electrolytes renal function tests, liver function tests and coagulation profile were within normal limits. Chest X Ray showed cardiomegaly and an increased cardiothoracic ratio with clear lung fields (Figure 1). ECG revealed normal sinus rhythm with left ventricular hypertrophy. Subsequently an echocardiogram demonstrated an ejection fraction of 62% and normal wall motion of ventricles. MRI dorsal spine showed multiple T2 hypo extradural lobulated lesions with mild post contrast enhancement seen in the posterior aspect of spinal cord from D4-D8 mildly compressing cord (Figure 2).

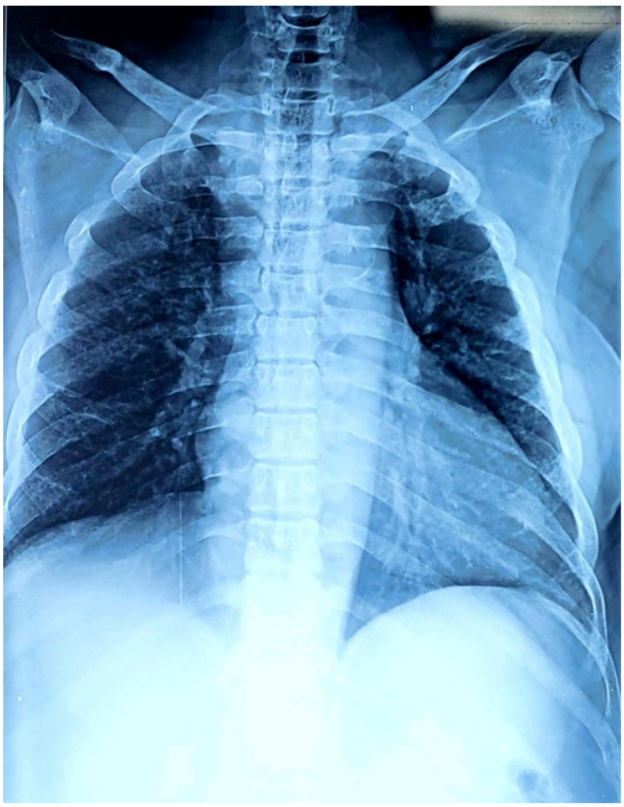

**Fig. 1:** Chest radiography showing Cardiomegaly

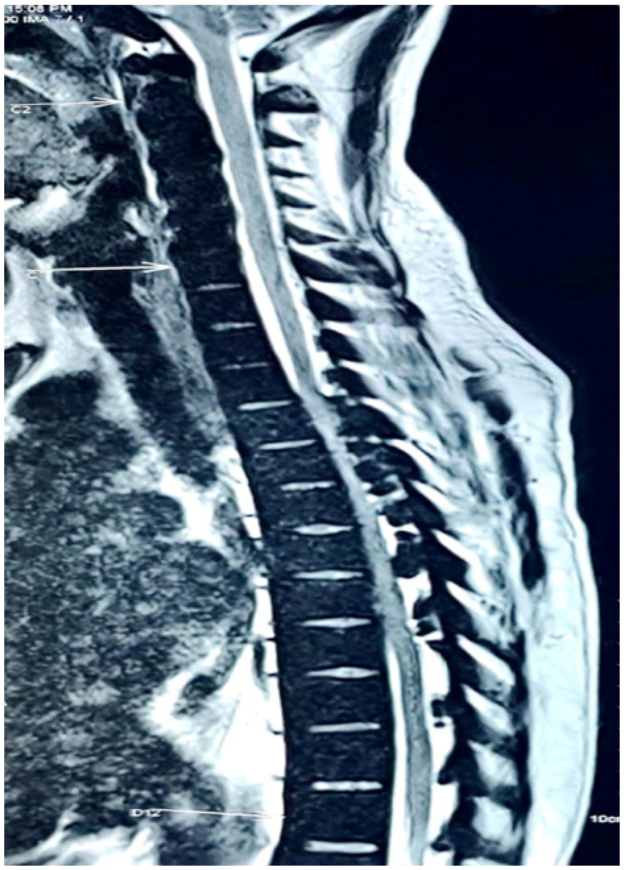

**Fig. 2:** D4-D8 extradural lesion

He was planned for T4-T8 laminectomy and excision of extradural lesion under American Society of Anaesthesiologists (ASA) physical status II and patient was kept nil per oral before surgery as per ASA guidelines. Upon arrival into the operating theatre, consent was checked, patient was laid supine, standard ASA monitors were attached. Peripheral venous access was secured with two wide bore cannulas. Baseline HR was 92bpm, NIBP of 126/84mmHg, SpO2 of 96% on room air and respiratory rate of 18 cycles/min. Aseptic precautions were adhered to and antibiotic prophylaxis was instituted. As a part of blood conservation strategy, bolus intravenous Tranexamic acid 1g was given prophylactically. Gradual compression stockings were placed on the patient's legs to prevent any thromboembolic events in the perioperative period. Preoxygenation was done with 100% oxygen for 3 minutes. Controlled general anaesthesia was achieved with Midazolam 1mg, Morphine 7.5mg, Propofol 100mg and was paralyzed with Vecuronium 6mg. Patient had Cormack Lehane Grade IIb during laryngoscopy and was intubated with 8mm cuffed ETT. Bilateral air entry was checked, found adequate and tube was fixed at 21 cms depth. Patient was maintained on volume controlled ventilation with O2: Air ratio of 60:40 along with Sevoflurane. Controlled

ventilation was facilitated with regular intermittent doses of vecuronium. He was cleaned and draped and the surgery commenced. Intraoperatively surgery proceeded for 5 hours with total blood loss of around 900ml and urine output of 600 ml. The hemodynamic variables remained stable throughout the intraoperative period. He received 1 unit of packed red cells intra operatively along with as part of perioperative fluid therapy. After surgery when patient's spontaneous efforts were equal and adequate, neuromuscular blockade was reversed with myo-pyrrolate. Postoperative pain was managed with multimodal analgesics. Patient was started on DVT prophylaxis on POD 3 during the hospital stay and then mobilised gradually. Postoperative ambulation physiotherapy was continued effectively. His postoperative course was uneventful and was discharged 8 days later from the hospital.

## 3. Discussion

Thalassaemia intermedia is characterized by haemoglobin concentrations of 70–100 g/L and usually with symptoms of anaemia, jaundice and hepatosplenomegaly.[3] They usually develop moderate anaemia with microcytic and hypochromic RBCs. Extramedullary hematopoiesis is a common complication which is a physiological compensatory phenomenon for almost all chronic haemolytic anaemias.[4] There may also be skeletal changes such as expansion of facial bones and obliteration of maxillary sinuses. Unlike patients with thalassemia major, thalassemia intermedia mainly occurs as a result of increased intestinal iron absorption rather than transfusion therapy. Clinical complications in thalassemia intermedia include gallstones, extramedullary hematopoiesis, leg ulcers, thromboembolic events and pulmonary hypertension which is the major cause of heart failure in these individuals.[3] A proper laboratory diagnosis is crucial for characterizing the different forms of thalassemia with important implications for prevention and treatment. The hematological parameters including red cell indices and morphology, followed by separation and measurement of Hb fractions are the basis for the identification of thalassemia carrier. Screening for thalassemia intermedia reveals Hb A 0%, Hb A2 3.5-5%, Hb F 90-96%. Molecular analysis remains the definitive diagnostic tool for thalassemia intermedia phenotype.[5,6] Recently MRI is used to evaluate iron accumulation in liver and heart to guide chelation treatment[7] Medical management of thalassemia includes blood transfusion and chelation therapy. Drugs used in chelation therapy include desferrioxamine, deferiprone and deferisarox. Potential side effects of these drugs are sensoneural deafness, visual disturbances, vertebral dysplasia, growth retardation (desferrioxamine), agranulocytosis (deferiprone), transient deterioration in renal function, skin rashes and gastrointestinal upset (desferasirox). Leucocyte depleted blood with extended antibody typing used to reduce risk of allo immunisation.[8,9] Splenectomy has been shown to be effective in reducing transfusion requirements and improving morbidity. However, splenectomy may cause increased risk of post-operative infections and thrombotic events. Other novel treatment modalities include stem cell transplantation, bone marrow transfer, foetal haemoglobin inducers and gene therapy.[10]

### 3.1. Preoperative assesment

Anesthesia challenge starts with difficulty in positioning due to extramedullary hematopoiesis occurring at vertebral canal, unanticipated difficult airway including difficult mask ventilation and intubation because of facial deformity caused by erythropoitein release and massive marrow hyperplasia.

The cardiovascular system should be carefully evaluated as these patients are at risk of developing pulmonary hypertension. It is important to prevent conditions worsening it, such as hypoxia, acidosis and hypercarbia. A full blood count should be done to identify baseline hemoglobin of the patients and opitimise them for surgery by transfusion preoperatively.[10] The laboratory criteria for transfusion include Hb of less than 70g/l on two occasions, more than 2 weeks apart. Other transfusion triggers include significant symptoms of anaemia, poor growth or failure to thrive, complications from excessive intramedullary haematopoiesis such as pathological fractures and facial changes. This thalassemia patients can be optimised preoperatively with hyper transfusion therapy thereby normalising the pre transfusion hemoglobin levels, reducing ineffective erthropoietic process and this preventing bone deformities.

### 3.2. Intraoperative

Blood salvage techniques plays a vital role in managing and controlling blood loss during intraoperative management of major spine surgeries.[11,12] Combination of preoperative blood donation, recombinant human erythropoietin and intraoperative use of boluses of tranexamic acid in addition to controlled blood pressure to prevent excessive blood loss have been found to be useful in these conditions.[11] And also Thalassemia patients are given broad-spectrum antibiotics coverage in the peri operative period to prevent infections.[13] In the presence of cardiomyopathy, pulmonary hypertension or congestive heart failure, close cardiovascular monitoring is necessarily indicated intra-operatively with transoesophageal or transthoracic echocardiography for real-time assessment of ventricular filling and contractility.[14] In addition, Hypercoagulability is commonly seen in thalassaemia, and thus appropriate measures to prevent deep venous thrombosis should be instituted.

### *3.3. Postoperative*

In the postoperative period, careful attention needed while transfer and positioning the patients as they are prone for pathological fractures because of osteoporosis and leg ulcers due to thin and fragile skin. Measures should be taken to reduce this risk by using compression stockings, low molecular weight heparin and mobilisation as Beta thalassemia inter media patients are at increased risk of thrombosis.[10]

## 4. Conclusion

Perioperative management of thalassemia patients appears challenging as multiple systems are involved due to the nature of the disease and also can be consequence of repeated blood transfusions leading to iron overload requiring chelating therapy. In this case, surgical intervention has improved the function, sensory and motor strength and ambulation thereby improving the quality of life dramitically. Thus, a multidisciplinary approach is required pre-operatively to allow thorough systematic investigation of pathological features, to optimise the patient's physical condition for readiness for such major surgeries and for careful planning of intraoperative period to achieve best possible outcome in these patients.

## 5. Source of Funding

None.

## 6. Human Subjects

Consent was obtained by participant in this study.

## 7. Conflicts of Interest

Nil.

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