Transcatheter Aortic Valve Replacement (TAVR) in Thalassemic Patients

To the editor,

A 67-year female, known case of beta thalassemia intermedia and rheumatoid arthritis (RA) (since 50 years) was diagnosed to have severe calcific aortic stenosis, valve area of 0.7 cm^2, gradient 90/60 mm Hg, hypertrophic normal left ventricle without regional wall abnormality. Investigations revealed a sinus rhythm on ECG, hemoglobin 5.7 gm/dl, serum ferritin was 1650 ng/ml, indirect bilirubin 1.7 mg/dl, uric acid 9.6 mg/dl, severe restrictive airway disease, lumbosacral radiogram revealed multiple lobulated paravertebral masses with expansile lytic lesions, possible signs of extramedullary hematopoiesis, a normal renal function, coagulation profile and ultrasound abdomen. She was on oral diuretics, methylprednisolone, hydroxychloroquine, folic acid, amlodipine and deferoxamine. She had a history of blood transfusion one year ago. Airway assessment was normal and she was scheduled for a transcatheter aortic valve replacement (TAVR) under conscious sedation and local analgesia.

Pantoprazole and alprozolam were administered orally the night before surgery. Standard ASA monitors were placed and radial arterial and central venous line were inserted aseptically. Dexmedetomidine infusion at 0.5 ug/kg/hour, 50 ug fentanyl citrate, cefoperazone sulbactam for antibiotic prophlaxis and Heparin 100 IU/kg to maintain an activated clotting time of 250-300 seconds were administered. TAVR was carried out via the right femoral artery with core valve of 23 mm. A left bundle branch block developed which reverted to sinus rhythm within one hour of temporary pacemaker support. Transthoracic echocardiography (TTE) did not reveal any paravalvular leak, pericardial tamponade or new regional wall motion abnormality. Heparin was reversed with protamine in a 1:1 dose. Two units of packed red blood cells (PRC) were transfused, was shifted to the ICU with a hemoglobin of 6.5 gm/dl. She was administered dual antiplatelet drugs to be continued for six months. The temporary pacemaker was removed after 24 hours and the patient was ambulated. She was discharged on Day 5 and was well on 1 week follow up.

Though case reports are available in the literature for open heart surgeries in thalassemic patients but TAVR has not been reported till date.

Thalassemic patients undergoing cardiac procedures pose challenges to the cardiac anesthesiologist. They may have organ dysfunction due to haemochromatosis including cardiomyopathy, liver cirrhosis and altered renal profile.[1] Diabetes mellitus, thyroid dysfunction may be encountered in beta thalassemia from anterior pituitary dysfunction due to iron overload.[2] Hypercoagulability is commonly encountered thus perioperative measures to prevent deep venous thrombosis (DVT) need to be taken. Airway assessment is of vital importance as the probability of difficult intubation in the presence of maxillary hypertrophy, nasal bridge depression, dental protrusion is high.[3] Laryngeal mask airway insertion may be challenging due to a high-arched palate. Caution is needed during transfer and positioning of patients because of risk of pathological fractures due to osteoporosis. Health personnel need to avoid exposure to the patient's blood to prevent infections. Associated RA may be complicated by involvement of other organs lung involvement extending to pulmonary fibrosis, heart failure and cervical spine or temporo-mandibular joint arthritis may lead to a difficult

airway.[4] Anesthetic agents are chosen according to patient's clinical condition and extent of organ damage. Broad-spectrum antibiotics are generally administered since they are immuno-compromised.[1]

Use of conscious sedation for TAVR reduces deleterious effects of anesthetic agents, is associated with greater hemodynamic stability, reduced inotrope and vasoconstrictor requirement, neurological monitoring, respiratory complications, avoids urinary catheterization, early ambulation, shorter length of stay, all of importance in patients with comorbidities.[5] In case of conversion to open-heart surgery appropriate measures to prevent hemolysis is the use of PRC in the priming solution, a non-pulsatile flow pattern and low pressures for cardiotomy suction are required in thalassemia patients. Unstable alpha thalassemia hemoglobin chains may precipitate after prolonged exposure to relatively low temperatures which can be prevented by a short duration of bypass under normothermia or mild hypothermia (32-35 degrees).[6]

Thalassemic patients need to be worked up thoroughly. Close communication between an anesthetist, surgeon, pulmonologist and hematologist is essential to ensure optimum management of such patients undergoing cardiac procedures.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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