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Managing Complexities in Ascending Aorta Surgeries in Patients with Marfan Syndrome – A Case Series

Dr. Sandeep More; Dr. Prerana Dixit; Dr. Sanjeeta Umbarkar
Seth G S Medical College & K. E. M. Hospital, Parel, Mumbai

Abstract:- Connective tissue disorders like Marfan's Syndrome, Loeyz-Dietz syndrome and Ehlers-Danlos syndrome are associated with variable degrees of annulo-aortic ectasia and cystic medial necrosis of the aortic wall. These patients present to cardiac surgical units with a spectrum of disorders ranging from aortic insufficiency, aortic root dilatation, ascending aortic aneurysm and dissection and pose unique anaesthetic challenges for the cardiac anaesthesiologist. These cases need a systematic, multidisciplinary, case-based understanding for the perioperative management in view of increasing prevalence of aortic root disorders.

We came across 25 cases for ascending aorta repair surgeries in the past 1 year at our tertiary cardiac care centre, out of which we present here a case series of four patients who had associated Marfan Syndrome as diagnosed using the Ghent criteria. Through this series, we want to emphasize on a thorough understanding of various aspects of anaesthesia management including preoperative optimization, positioning, intraoperative monitoring and care as well as post-operative complications expected in this patient subset.

Keywords:- *Marfan's Syndrome, Connective Tissue Disorders, Aortic Root Disorders, Ascending Aorta Aneurysm, Aortic Dissection, Bentall Procedure.*

I. INTRODUCTION

Connective tissue disorders (CTDs) form a heterogeneous group of relatively rare disorders primarily involving the structural proteins of the body tissues (1). Marfan's syndrome with a reported incidence of 1 in 3000 to 5000 individuals occurs due to defect in the FBN1 gene of chromosome 15 responsible for formation and functioning of the structural protein, fibrillin (2). A spectrum of multiorgan involvement results along with cystic medial degeneration of the aortic wall with decreased collagen fibrils, reduced aortic structural integrity and annulo-aortic ectasia (3). Associated musculoskeletal abnormalities include arachnodactyly, thoracolumbar scoliosis and pectus deformities. Ocular association like ectopia lentis is also not uncommon. Overcrowding of ribs and associated pulmonary interstitial involvement along with the features mentioned above make these patients a unique anesthetic challenge for perioperative management (4).

At our tertiary cardiac care centre, we came across 25 cases for ascending aorta repair surgery in the past 1 year, out of which seven patients had associated connective tissue disorders. We present here a case series of four patients who had Marfan Syndrome as diagnosed using the Ghent criteria (4,5) and presented with ascending aorta involvement of varying degrees. Increasing prevalence has recently been noted for aortic disorders. (6) Through this case series we want to throw light on the various anaesthesic challenges including preoperative optimization, positioning, intraoperative monitoring and post-operative complications expected in this patient subset.

II. COMMON PARTS OF ANAESTHESIA MANAGEMENT

All patients underwent a detailed pre-operative anaesthesia evaluation. After written informed consent, the patients were wheeled in the operation theatre. The cases planned for discussion in this series did not have involvement of the aortic arch vessels hence left radial artery and right internal jugular vein were cannulated for all patients under local anaesthesia. This was followed by induction of general anaesthesia and endotracheal intubation followed by cannulation of the left femoral artery. The right femoral artery was spared in all cases if needed for establishing peripheral cardiopulmonary bypass at any step. Anaesthesia was maintained with oxygen, air and sevoflurane along with sedation mixture of Inj. Fentanyl, Inj. Midazolam and Inj. Vecuronium. Monitoring included standard ASA monitors along with invasive blood pressure monitoring, entropy and Near infrared spectroscopy (NIRS) for cerebral oximetry. Additionally, somatic NIRS sensors were attached for monitoring renal and spinal cord oxygen saturation.

❖ *Case based approach of the cases as follows:*

A. Case 1 -

40-year-old female patient presented with sudden onset sharp stabbing back pain and progressive dyspnoea on exertion since 1 month. She was diagnosed with Stanford type A aortic dissection with dissection flap originating from the root of aorta and extending upto the descending thoracic aorta however sparing the arch vessels. There was associated severe aortic regurgitation and prolapse of the anterior mitral leaflet causing moderate mitral regurgitation. The patient had a blood pressure of 180/60 mm Hg on presentation which was managed using Inj. Nitroglycerin infusion and beta blockers

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in the critical care unit, pre-operatively. The patient had severe double-S-shaped thoracolumbar kyphoscoliosis since birth because of which she was unable to lie down supine and always needed 45 to 60 degrees semi recumbent positioning. She had mild obstructive pulmonary function tests and received bronchodilators for pre-operative optimization after consultation with pulmonary medicine. In the operation theatre, the patient was given a ramp using 2 pillows under the neck and 1 pillow under the thoracic curve. Central line insertion in the internal jugular vein was challenging as Trendelenburg position could not be given because of the fixed spine deformity and the same was done using ultrasound guidance under local anesthesia. Anticipating difficulty in intubation due to the positioning, we used bougie guided video-laryngoscopy for intubation. Trans-esophageal echocardiography probe insertion was attempted once but could not be negotiated beyond the nasopharynx due to inability to move the neck of the patient. Surgical positioning for sternotomy was done using adequate pressure padding at scapulae, axilla, occiput and lumbar curvature. After sternotomy, complete heparinisation was achieved and anterograde perfusion was established using left common carotid artery with an 8mm Dacron graft. Aortic cross clamp was applied just distal to the innominate artery. The patient underwent Bentall procedure with St. Jude aortic valved conduit no. 21 and coronary buttons were implanted on the conduit. The intraoperative course was uneventful and the patient was shifted to intensive care unit with Inj. Nitroglycerine infusion 1 mg/ml @1 ml/hour. She was extubated on post operative day 2 being hemodynamically stable and conscious and oriented. However, over the next 5 days in the ICU, she developed type 2 respiratory failure and was put on non-invasive ventilation. Chest Xray showed right lower lobe consolidation on day 3 for which high antibiotics were started. Gradual desaturation developed and the patient had to be reintubated after which she succumbed to ARDS and respiratory collapse on post operative day 5.

B. Case 2 -

37-year-old female presented with progressive dyspnoea on exertion since past 5-6 months and was diagnosed with aortic root dilatation and severe aortic regurgitation, severe tricuspid regurgitation and severe pulmonary hypertension on echocardiography. She was known case of Marfan's syndrome with mild kyphoscoliosis and hypermobile joints since the past 20 years and had mild obstruction on pulmonary function tests. Chest X-Ray was suggestive of left pulmonary interstitial involvement as a consequence of the spine deformity. Pre-operative pulmonary optimization with bronchodilators was done. Hemodynamic optimisation was done using beta blockers and ACE inhibitors. Anaesthesia management included ramp positioning and adequate padding of pressure points. Intubation was done in a deep plane of anaesthesia followed by neuromuscular relaxation using Inj. Rocuronium 1.2 mg/kg. Gentle video – laryngoscopy with cervical spine stability was done after monitoring train of four count 0 on NMT monitor. Modified Bentall procedure was performed using St. Judes Aortic valved conduit no. 27 and coronary buttons were preserved by sparing a rim of aorta around the annulus. During the procedure, the native aortic valve was noted to be floppy with 6 x 6.5 cm dilatation of ascending aorta and transition zone at the ascending aorta and arch junction. Intraoperative hemodynamic remained stable and patient could be weaned off cardiopulmonary bypass with minimal inotropic supports using Inj. Adrenaline 0.05 mcg/kg/min and Inj. Nitroglycerine 1mg/ml @2ml/hour. Postoperative course was uneventful and gradual weaning from ventilatory support was done over 24 hours. After 3 days of ICU stay and follow-up in ward, patient was discharged home on postoperative day 9.

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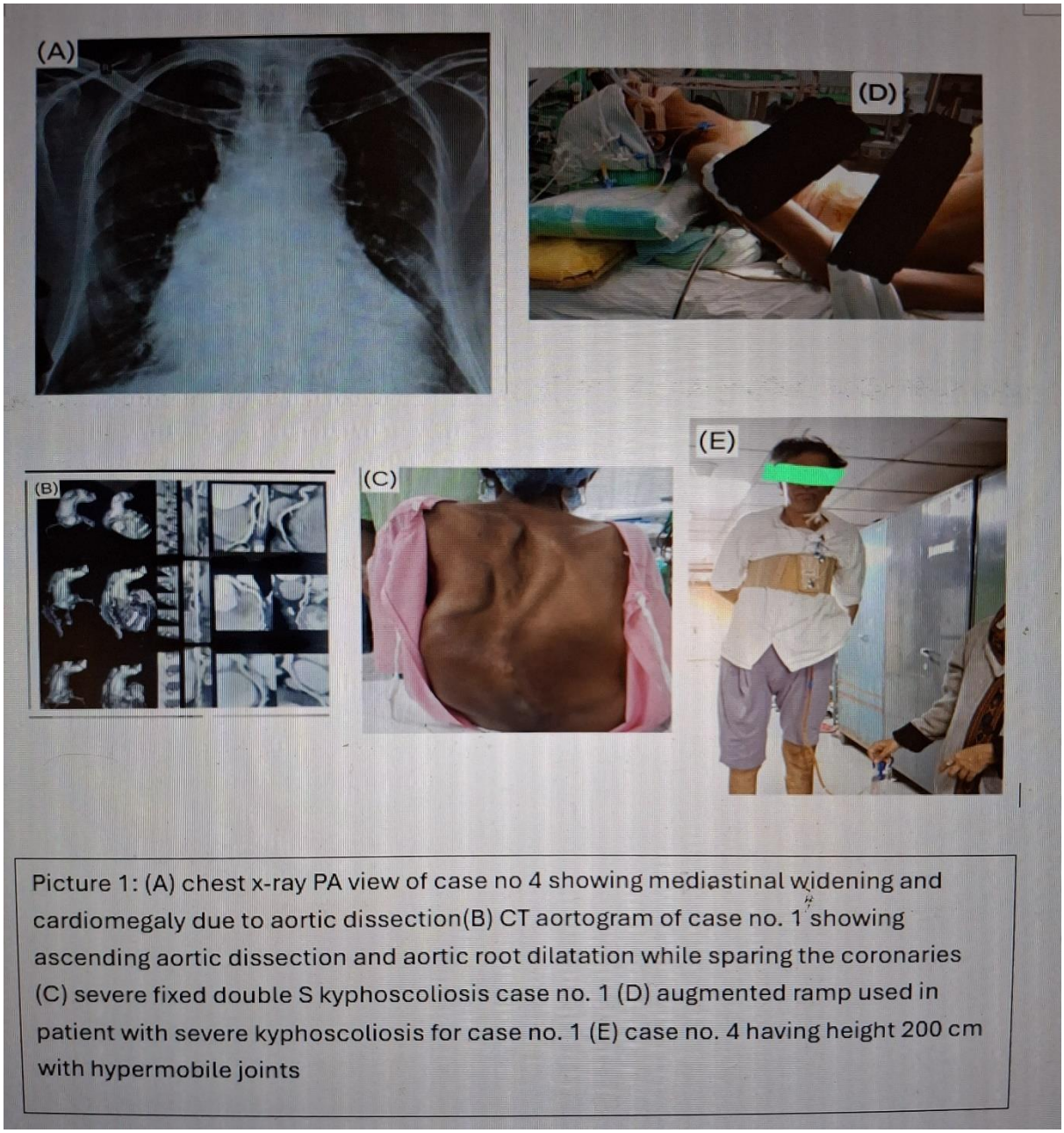

Picture 1:

C. Case 3 -

44-year-old male patient, known case of Marfan's syndrome and bicuspid aortic valve since last 17 years, presented with worsening dyspnoea on exertion from NYHA grade II to NYHA grade IV in the past 8 to 9 months. Transthoracic echocardiography revealed ascending aortic aneurysm with severe aortic regurgitation and severe mitral regurgitation and ejection fraction 30%. Patient was monitored in preoperative critical care unit after securing left radial artery cannulation for invasive blood pressure measurement and started on Inj. Dobutamine 5 mcg/kg/min via right internal jugular vein. Urgent surgical repair was planned. Intraoperative transoesophageal echocardiography revealed aortic annulus diameter 31 mm, ascending aorta diameter 51 mm and inter-sinus distance of 53 mm. There was associated severe mitral regurgitation attributable to prolapse of A2 P2 segments of mitral valve with severe tricuspid insufficiency. Cardiopulmonary bypass was established with aortic cannulation immediately proximal to the innominate artery. Bentall procedure was done using graft conduit made with TTK chitra mechanical valve no. 25 and dacron straight graft no. 24. Coronary buttons were implanted on the graft and alfieri stitch taken at the A2P2 segment for mitral valve repair. After weaning from cardiopulmonary bypass, there was significant blood loss due to oozing around the graft. The same was managed with transfusion of fresh frozen plasma and platelets concentrates supplemented with use of Inj. Fibrinogen concentrate 2 gm administered intravenously after test dose. Patient was shifted to ICU with Inj. Adrenaline 0.05 mcg/kg/min, Inj. Noradrenaline 0.05 mcg/kg/min and Inj. Dobutamine 5 mcg/kg/min. Elective ventilation was done for 24 hours and after noticing stable drain output the patient was extubated. Postoperative blood transfusion was done to maintain haemoglobin levels more than 9 g%. Inotropic

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supports were gradually weaned during the 6 days stay in ICU and patient was discharged home on POD 12.

D. Case 4 -

58-year male patient presented with intermittent anginal chest pain radiating to back for 1 year with increasing intensity in the past 1 month. He had associated dyspnoea on exertion NYHA grade II. Echocardiography revealed aneurysmal dilatation of ascending aorta with Stanford type Aortic dissection with severe aortic insufficiency. CT aortogram revealed dissection flap originating 1.7 cm distal to the sinus of Valsalva and extending till the left subclavian artery but sparing the arch vessels and coronary arteries. The patient had a height of 200 cm with hypermobile joints and arm span (205 cm) more than height. Standard operating room table was extended to give proper positioning to the patient with adequate padding of pressure points. General anaesthesia was induced and intubation done with video-laryngoscopy and bougie guidance in view of anticipated difficult intubation. Elective peripheral cardiopulmonary bypass was established using femoral artery and right atrial cannulation, guided by intraoperative transesophageal echocardiography. Bentall procedure was performed using St. Jude mechanical valved conduit number 25. True and false lumen of aorta were sandwiched surgically and adequacy of the same confirmed using TEE. Coronary button was done on the graft conduit and patient was weaned off cardiopulmonary bypass with Inj. Adrenaline 0.05 mcg/kg/min. Blood products were transfused to maintain haematocrit > 27. Patient was electively ventilated for 24 hours and had an uneventful ICU stay. He was discharged home on Postoperative day 10.

III. DISCUSSION

Thoracic aortic aneurysm due to cystic medial degeneration can occur as a result of connective tissue disorders, inflammatory disorders like Takayasu's arteritis, rheumatic valve disease with post-stenotic aortic dilatation, intramural hematomas or diffuse arteriosclerotic disease (6). Aneurysms in connective tissue disorders like Marfan Syndrome are unique due to deficiency of structural myofibrils in the aortic wall causing annulo-aortic ectasia. While elective surgical repair of the asymptomatic ascending aortic aneurysm is indicated at a diameter more than 50 mm in general population, the upper limit for a similar repair in patients with Marfan syndrome is reduced to more than 45 mm (7). When left untreated, aortic dissection can be a catastrophic and life-threatening event requiring emergency high risk surgery with increased morbidity and mortality. Pericardial tamponade, congestive heart failure, stroke, myocardial infarction due to coronary ostial dissection and malperfusion of the visceral organs are the most common causes of death after aortic dissection (6). Perioperative anaesthesia goals in patients with aortic aneurysms aims at preventing hypertension in order to minimize risk of aneurysm rupture or extension of an already started dissection.

Ascending aortic surgeries have evolved over the past few decades beginning with Bentall technique which involves aortic valve and ascending aorta replacement along with direct reimplantation of the coronary arteries. This was later modified by formation of coronary ostial "buttons" that are attached to the graft in the Modified Bentall procedure (9,10). Recent works have also focussed on valve sparing procedures such as David procedure and Wheat procedure in cases which do not affect the native valve significantly (11). Endovascular hybrid repair involving debranching and Thoracic Endovascular Aortic Repair (TEVAR) are the future of ascending aorta repair with much lesser perioperative morbidity (12).

The role of cardiac anaesthesiologist during all the above mentioned procedures in the operating room and catheterization laboratory is indispensable and has been further reinforced with the need to guide the procedures using transoesophageal echocardiography (11). Anaesthetic challenges are further exaggerated in special population groups like patients with connective tissue disorders with presence of skeletal malformations like long limbs, hypermobile joints, pectus deformities and kyphoscoliosis (4,8). Due to pectus deformities and as dilated aorta lies just beneath it, sternotomy may be crucial and there is a high chance of inadvertent injury to aorta causing hemodynamic collapse. Emergency peripheral bypass needs to be instituted and the patients should be thoroughly examined in the preoperative period for adequacy of femoral cannulation sites. Patient positioning holds utmost importance due to the presence of hypermobile joints. Excessive extension at a hypermobile atlanto-axial joint, may cause cervical cord injury and neurological damage during laryngoscopy and intubation (4) Presence of kyphoscoliotic deformities as discussed in the cases here, need special attention during long aortic repair surgeries to avoid musculoskeletal injuries under anaesthesia.

The systemic nature of connective tissue disorders is also associated with increased risk of bleeding complications due to increased vessel fragility, spontaneous rupture and prolonged bleeding times (13). This is further complicated with increased duration of aortic cross clamp time due to inherent nature of the surgical procedure.and the use of deep hypothermic circulatory arrest when needed (14). Surgical hemostasis after repair becomes crucial for successful outcome of patients. One patient in this study had fulminant intraoperative bleeding around the graft conduit which could be successfully managed using fibrinogen concentrate. More studies regarding the use of fibrinogen and other hemostasis strategies are warranted for aortic repair in specially in patients with CTDs.

IV. CONCLUSION

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Elective as well as emergency aortic repair surgeries are becoming a major proportion of routine cardiothoracic operating rooms. Increased awareness and improving health education is bound to bring patients with connective tissue disorders to the hospital at a time where aortic aneurysmal diseases can be salvaged with minimal risk. Detailed understanding and multidisciplinary approach involving cardiologist, cardiac surgeon, cardiac anaesthesiologist and radiologist comprising the "aortic team" can offer patients with connective tissue disorders, a safe hospital stay and an improved long term quality of life.

- *Conflict of Interest:*
  We declare there is no conflict of interest.

- *Consent:*
  Proper consent was taken from the patients included in this case series.

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