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Original Research Paper

Anaesthesiology

ANAESTHETIC MANAGEMENT OF A CASE OF MARFAN SYNDROME POSTED FOR BENTALL OPERATION: A CASE REPORT

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ABSTRACT Marfan syndrome is a connective tissue disorder that is inherited as an autosomal dominant trait.[3] These patients have tubular long bones giving 'Abe Lincon'[3] appearance. Cardio-vasular anomalies are responsible for early deaths in patients of Marfan syndrome. Defective connective tissue in the aorta and heart valves can lead to aortic dilatation, dissection, rupture and prolapse of cardiac valves.[3] Bentall procedure is a type of cardiac surgery involving composite graft replacement of the aortic valve, aortic root and ascending aorta, with the re-implantation of the coronary arteries into the graft.

KEYWORDS : Bentall, Aorta, Valves. [5]

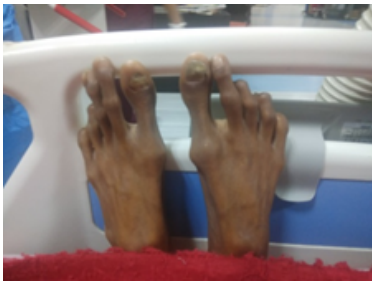
INTRODUCTION

Marfan syndrome is a connective tissue disorder that is inherited as an autosomal dominant trait. These patients have tubular long bones giving 'Abe Lincon' [3] appearance. Additional skeletal abnormalities include a high arched palate, pectus-excavatum, kyphoscoliosis, hyper-extensibility of the joints, early development of emphysema, high incidence of spontaneous pneumothorax, ocular changes such as lens dislocation, myopia, retinal detachment occur in more than half of the patients of Marfan syndrome. Defective connective tissue in the aorta and heart valves can lead to aortic dilatation, dissection, rupture and prolapse of cardiac valves. Mitrial gurgitation and mitrial valve prolapse are common. Valvular anomaly increases risk of bacterial endocarditis. Prophylactic beta blocker therapy is indicated for patients with dilated thoracic aorta. [3] Surgical replacement of aortic valve and ascending aorta is indicated when diameter of aorta exceeds 6cm[3] or if significant aortic regurgitation is present.

Pregnancy poses a unique risk for dissection of aorta in women with Marfan syndrome. Pre-op evaluation of patients with Marfan syndrome should focus on cardiopulmonary abnormalities. In most cases skeletal abnormalities have little impact on airway.[3] Care should be taken to avoid temporo-mandibular dislocation. In view of possibility of aortic dissection, any sustained increase in blood pressure is avoided; as can occur during laryngoscopy and surgical stimulation. High suspicion must be maintained for the development of spontaneous pneumothorax.[5]

CASE REPORT

A 20 yr old male patient with marfanoid habitus presented to our hospital with progressively increasing dyspnoea with chest discomfort on mild exertion, palpitations and orthopnoea since 2 years. Symptoms were aggravated acutely since 2 months. Further elicitation of history revealed that the patient was diagnosed case of Marfan syndrome though no significant past medical or surgical history warranting hospital admission was present. On general physical examination the patient was g, 188cm tall with long tubular extremities, hyper-extensible joints, long face, arm span greater than height, long neck, protruding jaw. Patient had a heart rate of 102 beats/ min, blood pressure 156/52mm Hg, SpO2 97% on room air, respiratory rate of 14/min. Airway examination- protruding teeth, high arched palate, increased horizontal mandibular length, more than 4 fingers mouth opening with modified mallampati grade 1 and an abnormally long neck. On auscultation normal vesicular breath sounds were heard over all lung fields. A high pitched early diastolic murmur was heard with maximum intensity at end expiration on the 3rd left intercostals space(suggestive of aortic regurgitation.) On workup, routine blood investigations were within normal limits, massive cardiomegaly could be appreciated on chest skiagram, two dimensional echo-cardiography revealed presence of severe aortic regurgitation, trivial tricuspid regurgitation, LVEF 50%, maximum aortic diameter of 9.6cm. CT Angiography revealed large aortic aneurysm of aortic root at sino-tubular junction with no evidence of dissection, narrowing of coronary ostia with aberrant origin of left circumflex artery from right aortic sinus. Patient was started on Tablet Enalapril 2.5 mg BD, Tab Metoprolol 50mg OD, Tab Aspirin 75 mg OD. Patient was counseled for Bentall operation. High risk consent for surgery and anaesthesia was obtained.



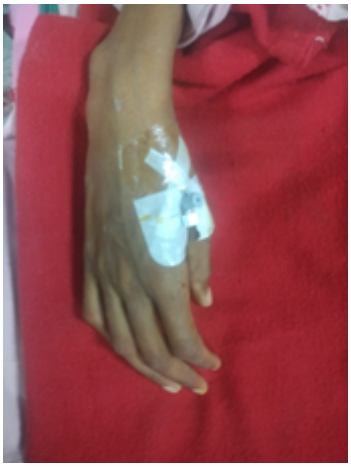
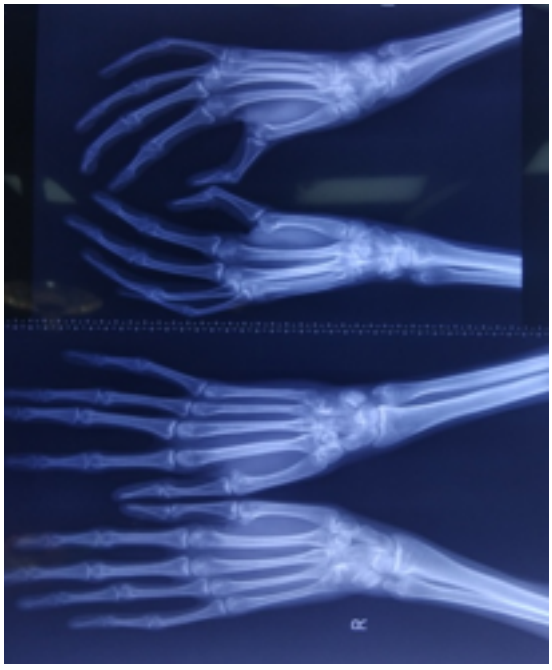


Image 1- Marfanoid Habitus Of Patient



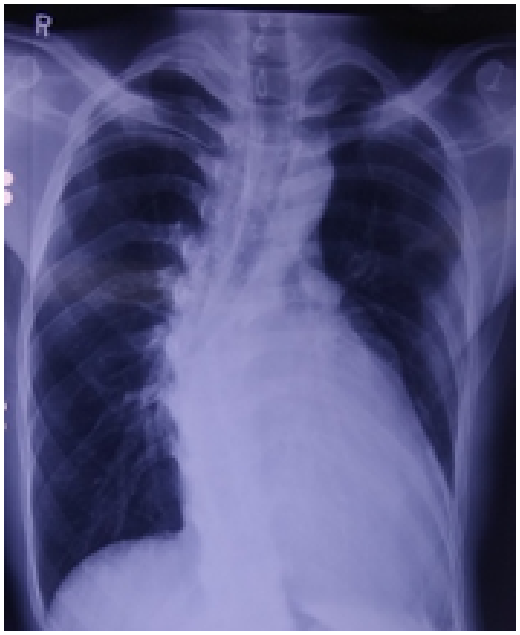
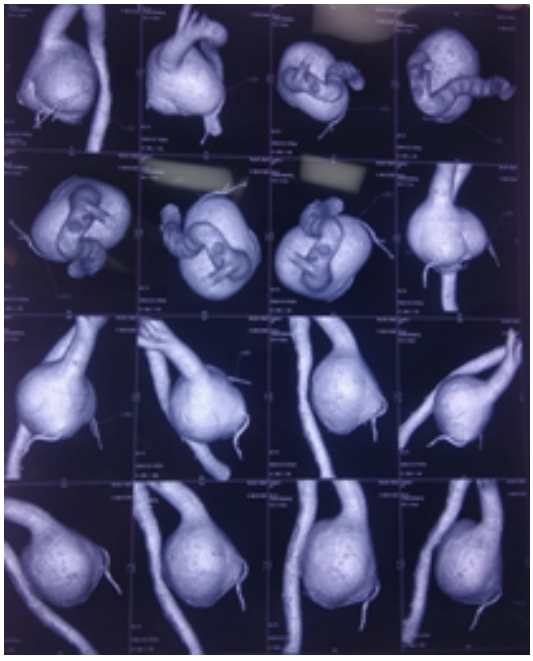


Image 2 – Skiagrams Of Patient Demostrating Long Tubular Bones And Massive Cardiomegaly.



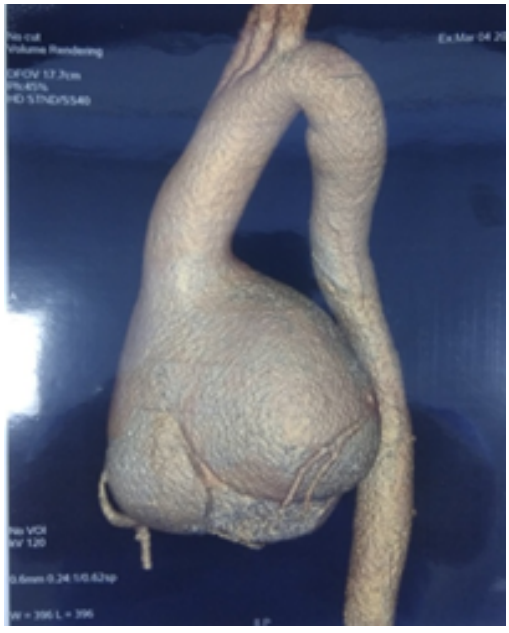


Image 3- CT ANGIO Showing Dilatation Of Aortic Root

ANAESTHETIC MANAGEMENT

Patient was evaluated thoroughly in the preoperative visit. Patient was pre-medicated with Inj. Fentanyl 250 mcg 30mins prior to induction of anaesthesia. On arrival in the operation theatre a 18 G peripheral intravenous access was secured. ECG electrodes were applied, NIBP cuff was tied, SpO2 probe was applied and baseline vitals were recorded. Radial artery and Right internal jugular vein were cannulated with aseptic precautions under local anaesthesia and ultrasonographic guidance. Induction was done using Inj. Etomidate 0.3mg/kg and Inj. Vecuronium 0.1 mg/kg followed by intermittent positive pressure ventilation for 3 minutes and endotracheal intubation. Other standard monitoring like $EtCO_2$, nasal temperature probe, urinary catheter, entropy, TEE were instituted after induction of anesthesia. Maintainance of anaesthesia was done with 0.6-1% Isoflurane in 100% Oxygen, Inj Fentanyl 2mcg/kg/hr and Inj. Vecuronium 0.1 mg/kg/hr. Inj. Heparin 4mg/kg was given before aortic cannulation. ACT was kept above 400sec. Patient was operated by median sternotomy.

Image 4- Opeeration Theatre Setup Showing Workstation, Tee, Entropy Monitors, Multipara Monitors And Various Drug Infusion Pumps In Place.

PERFUSION MANAGEMENT

Cardiopulmonary bypass was performed using heart lung machine with membrane oxygenator. Circuit was primed with 1000ml Ringer's Lactate, 500 ml plasma expander,5000IU Heparin, 20 gm Mannitol, 50 ml 5%Sodium Bicarbonate, Inj. Methyl Prednisolone 30mg/kg and Inj. Furosemide 10mg. Pump flow was set according to patients body surface area. Haematocrit was maintained around 25% and temperature of patient was maintained around 24-28 degree Celsius, PO_2 >300 mmHg and PCO_2 35-45 mmHg. During CPB ananesthesia was maintained with Inj Fentanyl 2mcg/kg/hr and Inj Vecuronium 0.1 mg/kg/hr. Serial blood sugar, arterial blood gas analysis and ACT measurements were done during procedure, in addition continuous arterial BP, temperature and urine output monitoring was done.

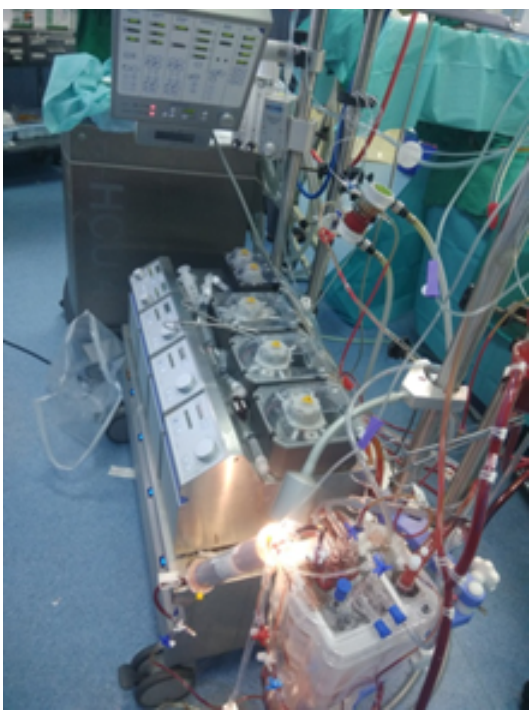


Image 5: The CPB Circuitry

SURGICAL PROCEDURE

After sternotomy, heart and the great vessels were dissected, anatomy was studied. Aorta was cross-clamped and opened after venting the heart through right superior pulmonary vein. The coronary ostial cannulae were used to infuse the cardioplegia solution. Aortic valve conduit was sutured first followed by coronary button implantation. Distal aortic anastamosis was performed after which re-warming was started. Before releasing the proximal aortic clamp, de-airing was done vigorously by providing head low position and blowing of the lungs. Separation from CPB was done and Inj. Protamine was given to keep ACT <120sec. Haemostasis was achieved and patient was shifted to ICU for weaning and extubation upon conclusion of the surgery.

Post operative course

During the postoperative stay in ICU patient had an episode of sudden desaturation, acute fluctuation in haemodynamics on postoperative day2. Development of spontaneous pneumothorax that was evident on clinical findings was confirmed by USG chest. Bilateral intercostal drains had to be inserted on urgent basis. This warranted need for prolonged ventilation and gradual weaning. Patient was tracheostomised on day 4 post-op to facilitate weaning. By post-op day 7, pneumothorax had resolved, patient was given spontaneous breathing trial and successfully extubated. Tracheostomy decannulation was done on post- op day 10. Patient was discharged successfully on post-op day 14.

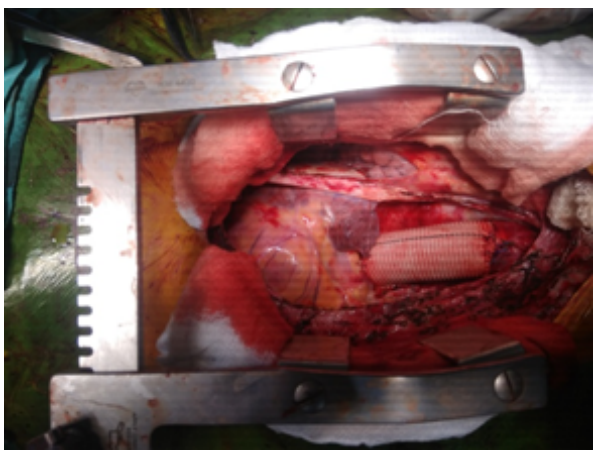


Image 6- The Surgical Site

CONCLUSION

Aortic valve replacement, aortic dissection surgery and Bentall operation are frequently performed cardiac surgeries in patients of Marfan syndrome. The surgeries on aorta have to be meticulously done. Myocardial and Neurological protection are chief concerns. Blood conservation and haemostasis play a pivotal role. Post operative period may be complicated by respiratory complications such as spontaneous pneumothorax as in this case. Meticulous surgery and extremely vigilant anaesthesia care improve the outcome.

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