Anesthetic management of a patient with Marfan syndrome and severe aortic root dilatation undergoing cholecystectomy and partial hepatic resection

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ABSTRACT

Due to high mortality associated with aortic dissection, anesthetic management of patients with Marfan syndrome with severe aortic root dilation is a challenging situation. We describe the anesthetic management of a patient with Marfan syndrome with severe aortic root dilation, who required major surgery like cholecystectomy with partial liver resection under general anesthesia. A 47-year-old female presented to pre-anesthetic clinic for cholecystectomy with partial hepatic resection for gall bladder carcinoma. Clinical features, transthoracic echocardiography and computed tomography of thorax supported a diagnosis of Marfan syndrome with severely dilated aortic root. Aortic dissection in patients with Marfan syndrome and severely dilated aortic root can be precipitated by major hemodynamic changes under anesthesia. Careful hemodynamic monitoring and avoidance of hemodynamic swings can prevent this life-threatening event.

Key words: *Anesthesia, aortic root, liver resection, Marfan syndrome*

INTRODUCTION

Marfan syndrome is an autosomal dominant multisystem connective tissue disorder usually associated with mutation in fibrillin protein with a reported incidence of one in 9800 individuals.[1,2] Among the triad of skeletal deformity, ocular manifestations and cardiovascular involvement usually present in the affected individuals, cardiovascular involvement being the most serious.[3] Aortic dilation and aortic dissection can cause up to 50-70% of deaths in Marfan syndrome.[1]

Cesarean deliveries in parturients with Marfan syndrome have been reportedly done safely under general and neuraxial anesthesia.[3,4] However, scarcity of reports of major surgery in patients with Marfan syndrome stimulated us to report a case of cholecystectomy with liver resection in a patient with Marfan syndrome with severe aortic root dilation. The patient gave us a written consent in local language for publication of this information.

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CASE REPORT

A 47-year-old female (wt. 49 kg) was scheduled for cholecystectomy with partial hepatic resection for gall bladder carcinoma. At pre-anesthetic clinic she complained of occasional palpitation and mild exertional dyspnea. On examination, she was tall (171 cm) having mild pallor, icterus and arachnodactyly [Figure 1] with arm span more than her height and mild kyphoscoliosis. Her exercise tolerance was near normal (NYHA II). Her airway examination showed high arched palate with Mallampati grade II.[5] On cardiovascular examination, we found wide pulse pressure with peripheral signs of aortic regurgitation (water hammer pulse, locomotor brachialis). An early diastolic murmur was also appreciated best on second right intercostal space close to sternum. A case of Marfan syndrome was suspected clinically and further investigations were requested. Transthoracic echocardiography (TTE)

showed aortic root dilatation (root diameter 52 mm) with aortic regurgitation (AR) and left ventricular eccentric hypertrophy with grade II mitral valve prolapse (MVP). Left ventricular ejection fraction was 50%. Chest X-ray showed mediastinal widening with cardiomegaly and normal lung parenchyma [Figure 2]. Aortic root dilatation was also vivid in computed tomography (CT) thorax [Figures 3 and 4].

High risk of surgery was explained to relatives and written consent was taken. The day before surgery the patient was admitted to high dependency unit. Her recent medications were reviewed and ramipril and frusemide were stopped. Her hemoglobin was 10.2 g%, prothrombin time (INR 1.4) and her liver function test and serum electrolytes values were normal. In the morning of surgery, patient was administered morning dose of metoprolol, aspiration prophylaxis in the form of ranitidine, intravenous amoxicillin for surgical prophylaxis, Vitamin K (for mildly prolonged prothrombin time) and anxiolytic (alprazolam) as per institutional protocol. Adequate blood products were arranged. We requested a multidisciplinary involvement of anesthesiologist, cardiologist, gastrointestinal and cardiac surgeon. In anticipation of requirement for cardiopulmonary bypass, a perfusionist was available for the case. We planned for general anesthesia with controlled mechanical ventilation along with epidural analgesia. Under basic monitoring (5 lead electrocardiogram (ECG), noninvasive blood pressure and pulse oximetry), epidural catheter was inserted in T8-T9 interspace. Epidural analgesia was started at the rate of 4 ml/h with 0.125% bupivacaine with 2.5 mcg/ml fentanyl. Right radial artery and right internal jugular venous cannulation was done under local anesthesia. Patient was induced slowly with titrated dose of fentanyl (3 mcg/kg), etomidate (0.2 mg/kg), midazolam (0.05 mg/kg) and vecuronium (0.1 mg/kg) after adequate mask ventilation. Intravenous magnesium sulfate in the dose of 50 mg/kg was injected slowly over 10 minutes to decrease intubation response.[6] The patient was intubated uneventfully using McCoy blade (Grade II a of Cormack-Lehane). A trans-esophageal echogram (TEE) probe was inserted post-intubation to monitor the patient's aortic root diameter. Central venous pressure (CVP), invasive arterial pressure, FloTrac continuous cardiac output (FloTrac/Vigileo™, Edwards Lifesciences, Irvine, CA,

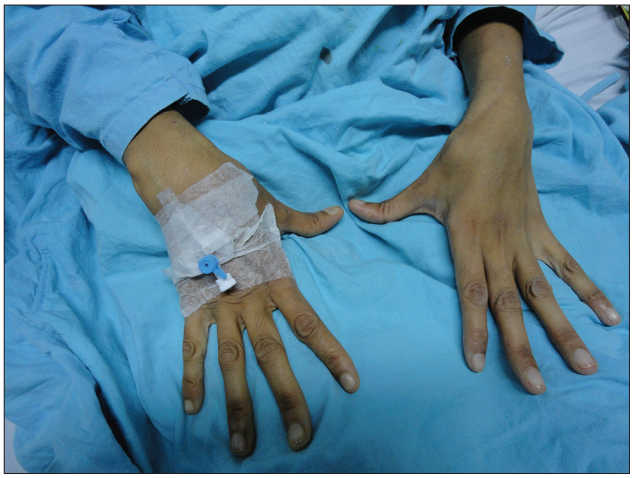

Figure 1: Arachnodactyly of the patient

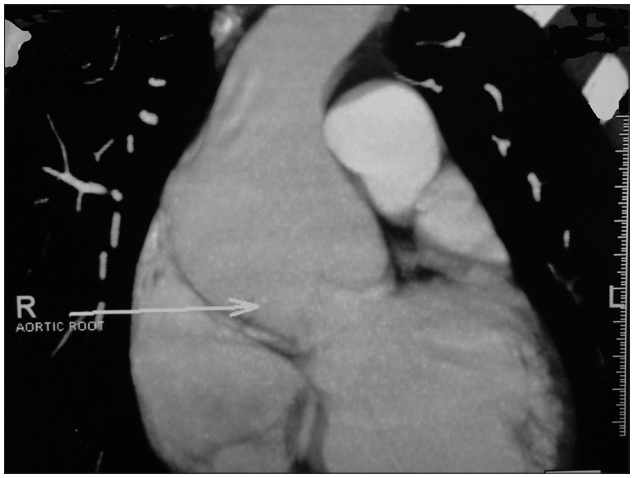

Figure 3: Computer tomography chest of the patient showing hugely dilated aortic root

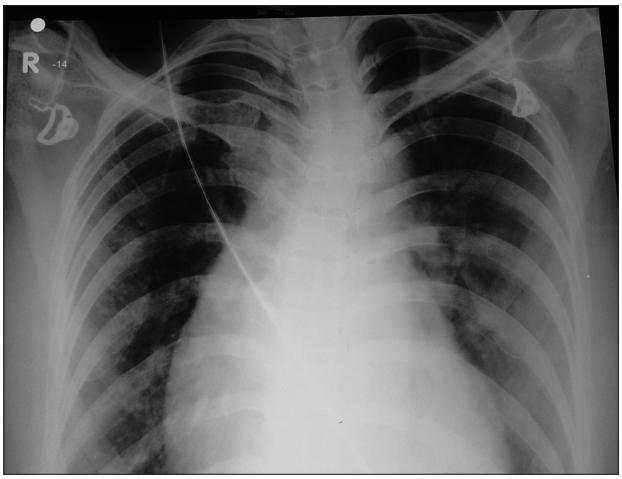

Figure 2: Chest X-ray of the patient showing scoliosis and cardiomegaly

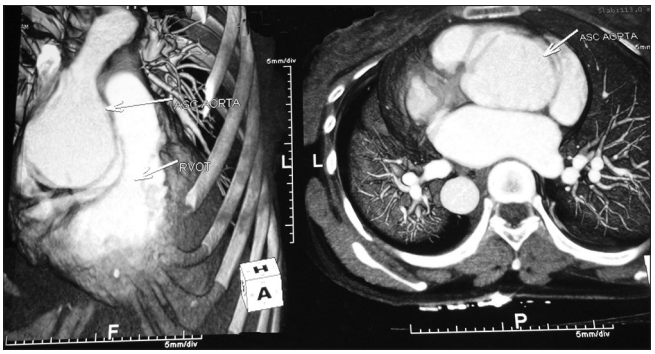

Figure 4: Computer tomography chest of the patient showing hugely dilated aortic root

USA) and neuromuscular monitor (TOF) and other basic parameters (ECG, pulse oximetry) were also monitored. Anesthesia was maintained with oxygen–nitrous (40:60), isoflurane, fentanyl and vecuronium. We were maintaining high normal heart rate (for regurgitant lesion like AR and MR) with sinus rhythm and cardiac contractility near preoperative status. We avoided any increase in afterload by vasopressor with pure alpha agonistic action like phenylephrine. We infused perioperative crystalloid and hydroxyl-ethyl starch (130/0.4) by monitoring CVP and stroke volume variation to maintain optimum preloading. Two units of packed cell (to maintain Hb around 8 gm %) and 4 units of fresh frozen plasmas were also transfused in view of blood loss (intraoperative INR was 1.52). However, it was a major surgery (duration 4h 30 min) with major fluid shifting and blood loss (total around 1.6 L) especially during hepatic resection (segment IVB and V) time. There were two episodes of mean blood pressure drop of more than 20% of baseline, but, they were managed by intravenous titrated mephentermine injection along with fluid. Intraoperative TEE showed mild change in aortic root diameter (52-53 mm). Perioperatively hypothermia was managed by using forced air warming (Warm Touch, Mallinckrodt, USA), and hypokalemia was corrected by potassium chloride infusion. At the end of surgery, the patient was reversed with neostigmine and glycopyrrolate, but was not extubated in view of prolonged surgery, blood loss, fluid shift and metabolic acidosis (pH 7.19, base deficit of -8). She was transferred to Post Anesthesia Care Unit (PACU) with monitors, where acidosis correction was given with sodium bicarbonate slow infusion. In PACU she was monitored as in operation theater. Euvolemia, eucapnia, normothermia, analgesia were maintained. Acid base and electrolytes disturbance corrected. TTE was arranged in post-op day one to evaluate cardiac contractility, left ventricular ejection fraction and diastolic function and was found favorable for extubation. She was extubated uneventfully 18 hours of PACU admission. Day 3 morning she was transferred to general ward from where she was discharged on post-operative day 7. Patient is now under follow-up of cardiovascular surgical team.

DISCUSSION

'Elastic fibre degeneration', lacking of smooth muscle cells in aorta and mucopolysaccharide deposition in between the cells of the media are the main features of aorta in Marfan syndrome.[1,2] This is known as 'cystic medial degeneration'.[1,2] Reduced distensibility of aorta with hemodynamic changes occurs as a result.[1] The goals of the anesthetic management of Marfan syndrome patients are thus first to decrease aortic pressure and to monitor the signs with preparations for possible aortic dissection.[3,4]

Our patient presented late with Marfan syndrome along with emergent condition (carcinoma gall bladder). Her medical management included reduction of arterial pressure with angiotensinogen receptor blocker (ramipril), beta-blockers and diuretics. Since cholecystectomy with liver resection is a major procedure, we did not consider prophylactic surgery for aortic root dilation at same setting; although, surgical correction is usually advised when aortic root diameter is 50 mm.[3]

We chose etomidate for induction due to its hemodynamic stabilizing action. Additionally, we have infused magnesium sulfate to decrease intubation stress and ultimately to reduce hemodynamic swings and chances of aortic dissection.[6,7] Inhalation agents decrease the force of cardiac contractility and as a result can decrease the chance of aortic dissection to some extent.[3] Epidural analgesia using a low dose of bupivacaine with opioids was administered to control intra and post-operative pain without causing hemodynamic alterations. In the event of aortic dissection, there would not be any neuraxial block to hinder our resuscitative approaches.[3] Hemodynamic instability was minimized with adequate preparation during intubation, intraoperative volume resuscitation and planned extubation with epidural analgesia.

To our knowledge, this is the first case report of a patient with Marfan syndrome and severely dilated aortic root undergoing major abdominal surgery. In conclusion, a multi-disciplinary involvement along with close hemodynamic monitoring is the key of success in those cases.

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