Case Report

Combined Bentall and modified Ravitch procedures in a patient with Marfan syndrome

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Abstract

Marfan syndrome is an inherited, connective-tissue disorder transmitted as an autosomal dominant trait. Cardinal features of the disorder include tall stature, ectopia lentis, mitral valve prolapse, aortic root dilatation, and aortic dissection. Pectus excavatum may exist as an isolated lesion or in association with a genetic syndrome such as Marfan syndrome. We report the successful management of a simultaneous correction of pectus excavatum and the underlying cardiovascular diseases.

Key words: Aortic aneurysm, Marfan syndrome, pectus excavatum

Introduction

Marfan syndrome is an inherited, connective-tissue disorder transmitted as an autosomal dominant trait. It has an estimated prevalence of one in 3000 to 5000 individuals.[1] Cardinal features of the disorder include tall stature, ectopia lentis, mitral valve prolapse, aortic root dilatation, and aortic dissection.

Pectus excavatum may exist as an isolated lesion or in association with a genetic syndrome such as Marfan syndrome. Pectus excavatum is seen in two-thirds of patients with Marfan syndrome or related connective tissue diseases.[2] Surgical correction of pectus excavatum is usually performed for cosmetic reasons and sometimes for physiological reasons. Cardiopulmonary complications resulting from mechanical compression by the depressed chest wall may be an indication for surgical correction, especially in patients with underlying cardiac disease. Cardiac compression can contribute to postoperative hemodynamic instability if the pectus deformity is left uncorrected. Anesthetic challenges for patients with Marfan syndrome and pectus excavatum include aneurysm of ascending aorta with risk of aortic dissection, severe aortic regurgitation, kyphoscoliosis, and reduced respiratory reserve resulting from pectus deformity.

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Access this article online	
Quick Response Code:	Website: www.joacp.org
	DOI: 10.4103/0970-9185.105812

Case Report

A 14-year-old boy, known case of Marfan syndrome, presented with chief complaints of dyspnea, while playing, which had progressed in last two months to dyspnea on routine activities. Patient had a typical Marfan habitus with arm span > height (194 > 179 cm), chest deformity (pectus excavatum; Figure 1), and kyphoscoliosis. He had a normal airway (Mallampatti class I). Past history included retinal surgery and lens replacement for retinal detachment and lens dislocation. Diagnosis of Marfan syndrome was made

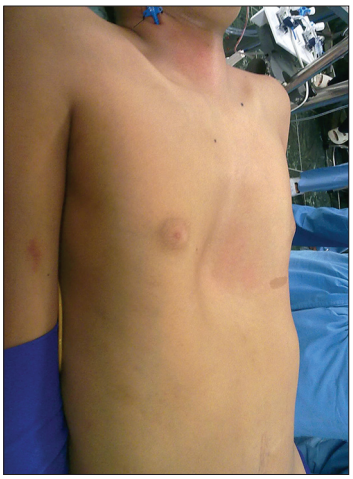

Figure 1: Chest wall deformity (pectus excavatum)

in accordance with the Ghent criteria.[1,3] Cardiovascular examination revealed diastolic murmur at apex. Routine laboratory values were unremarkable. Chest X-ray showed scoliosis with convexity to left, crowding of ribs on right side, kyphosis in lower thoracic region, and marked displacement of the heart to the left. Pulmonary function tests revealed moderate restrictive ventilation defect. Transthoracic echocardiography showed tricuspid aortic valve with severe aortic regurgitation, no aortic stenosis, aortic annulus 26 mm, severely dilated aortic root (68 mm), aneurysmal ascending aorta extending up to origin of right brachiocephalic artery, no aortic dissection and normal ventricular function. Computed Tomography of chest revealed a dilated aortic root, a fusiform aneurysmal proximal ascending aorta, severe pectus excavatum (5.2 cm central depression of the sternum), displacement of the heart into left side of chest with compression of left lung and kyphoscoliosis [Figure 2]. Bentall procedure with concomitant chest wall repair was planned, and informed consent was taken for the same.

General anaesthesia was induced with intravenous (IV) midazolam (0.05 mg/kg), fentanyl (3 mg/kg), thiopentone (4 mg/kg) and pancuronium (0.1 mg/kg). Adequate care was taken to support the limbs during anesthesia and surgery. Intraoperatively standard cardiac monitoring was done including invasive arterial pressure, central venous pressure and transoesophageal echocardiography. Anesthesia was maintained with isoflurane in oxygen and intermittent doses of midazolam, fentanyl and pancuronium. Tidal volume of 7-8 ml/kg was used to maintain normocarbia. Surgery was initiated with midline incision and median sternotomy. Bone wax was used to control bleeding from sternal medulla. After systemic heparinization (400 IU/kg), aorto-bicaval cannulation was done. Aorta was cross clamped, heart was arrested with cold blood cardioplegia and patient cooled to 28°C. A 25 mm St Jude mechanical valved conduit (St Jude Medical, Inc, St Paul, MN.) was placed and coronary buttons were implanted to the conduit. Aortic cross clamp time was 99 minutes and cardiopulmonary bypass time was 144 minutes. After a smooth weaning from cardiopulmonary bypass, hemostasis was achieved.

After achieving hemostasis, chest wall repair was started. The pectoral and rectus muscle flaps were elevated off the sternum and costal cartilage. Total subperichondrial resection of the deformed costal cartilages was achieved by incising the perichondrium anteriorly, and the entire third to eighth cartilages were removed to the costochondral junctions bilaterally. The sternum was dissected free from the intercostal neurovascular bundles bilaterally, medial to the internal mammary arteries. The deformed portion of the sternum was transacted in two points, and the divided sternum was fixed and corrected in elevated position by using a dynamic compression plate (3.5 mm/15 holes, Synthes Holding AG, Solothurn, Switzerland) passed horizontally posterior to the ribs to support the lower xiphisternum [Figure 3]. Following the correction of the deformity, all intercostal bundles were attached to the sternum. The plate was fixed to the xiphisternum and to the ribs bilaterally with steel wires. The pectoral and rectus muscles were reattached to the sternum after positioning of mediastinal and pleural drains. Routine closure of sternum was done with steel wires. Total duration of the surgery was six hours.

Postoperative analgesia was achieved with IV morphine infusion (10-30 mg/kg/hr) till the patient was on mechanical ventilation, followed by IV paracetamol (15 mg/kg, six hourly). Patient was hemodynamically stable, and trachea was extubated on day one. Patient had a good cosmetic repair [Figure 4] and was discharged from the hospital on tenth day.

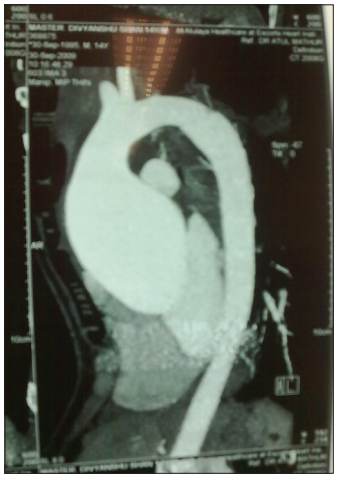

Figure 2: Computed tomography of chest revealing dilated aortic root and aneurysmal ascending aorta

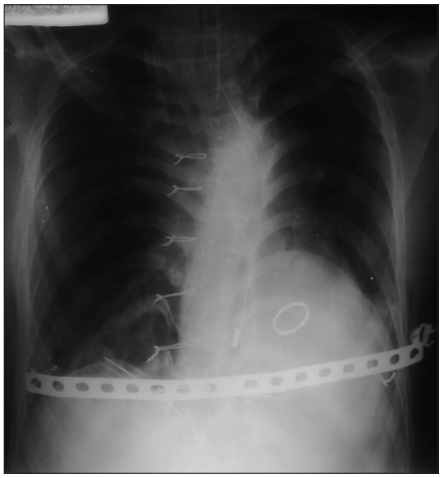

Figure 3: Chest X-ray with dynamic compression plate supporting lower xiphisternum

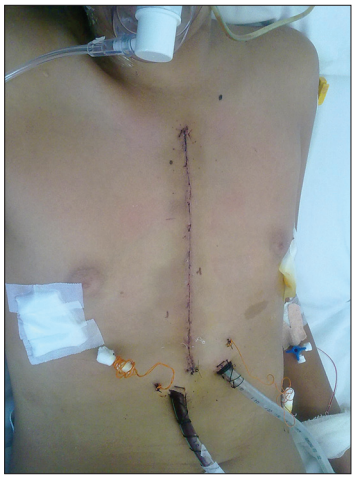

Figure 4: Postsurgical good cosmetic repair

Discussion

Marfan syndrome is marked by the triad of musculoskeletal, aortic, and ocular pathology.[4] It is an autosomal dominant connective tissue disorder that results from mutations in the fibrillin-1 (FBN1) gene on chromosome 15, which encodes for the glycoprotein fibrillin. FBN1 is a major building block of microfibrils. A mutation in the gene can reduce the amount and/or quality of FBN1 that is available to form microfibrils. Growth factors are released inappropriately, causing the characteristic features of Marfan syndrome. Abnormalities involving microfibrils result in weakening of the aortic wall. Progressive aortic dilatation and eventual aortic dissection occur because of tension caused by left ventricular ejection impulses. Likewise, deficient FBN1 deposition leads to reduced structural integrity of the lens zonules, ligaments, lung airways, and spinal dura. The primary cause of morbidity and mortality associated with Marfan syndrome is aortic dissection. Bentall and de Bono[5] in 1968, described the technique of replacing the ascending aorta and aortic valve in patients with Marfan syndrome.

Pectus excavatum and pectus carinatum are chest wall deformities in children, the former being more common with a reported incidence of one in 400-1000.[6] There is a definite association of cardiovascular abnormalities with thoracic skeletal deformities in patients with Marfan syndrome. The main issue in such cases is how to approach and repair both the pectus excavatum and cardiac lesion in a single stage. The problem associated with a pectus deformity is the difficulty in performing a midline sternotomy with symmetrical retraction while providing adequate exposure. Preservation of both internal mammary arteries is also imperative for appropriate sternal wound healing.

There are concerns regarding combined correction which include an increased risk due to complications such as bleeding, infection, very extended operating times, and limited exposure of the heart.[7] Successful single-stage correction of both lesions without any complications has been reported.[8,9] Staged operation for the two different disorders is associated with following disadvantages (i) Uncorrected pectus excavatum may severely impair cardiovascular function in the immediate postoperative period by precipitating cardiac displacement/compression. (ii) Leaving either the pectus excavatum or the cardiac defect untreated may make second intervention necessary. (iii) A second intervention may be fraught with technical difficulties because of adhesions, the risks of further anesthesia, and costs. (iv) Moreover, if the patient requires anticoagulation or antithrombotic therapy after the first operation, the risks associated with a second operation may increase greatly.

Preoxygenation is helpful in patients with poor respiratory reserve. Anesthetic agents are chosen with the goals to provide afterload reduction and prevent bradycardia (hazardous in patients with aortic regurgitation). To reduce the risk of aortic dissection, sympathetic stimulation caused by laryngoscopy, intubation, incision and sternotomy, should be minimized by adequate supplementation of sedatives and narcotics. Patients with Marfan syndrome are at increased risk of spontaneous pneumothorax and emphysema, hence positive pressure ventilation with large tidal volumes, high airway pressure and high positive end-expiratory pressure should be avoided.[10] In patients with Marfan syndrome, the ligaments, tendons and joints are usually loose, resulting in subluxation and dislocations. Excessive traction on the jaw during laryngoscopy should be avoided to prevent temporomandibular joint dislocation. Due to hypotonia and hyperextensibility, the joints should be handled very gently. Proper positioning and limb support should be given to avoid joint trauma during anesthesia and surgery.[10] Neuraxial block may be difficult because of spinal deformity.

Good sternal healing has been reported with the temporary or permanent retrosternal bars, resulting in an essentially normal, cosmetically appealing restoration of the chest wall with no pectus recurrence.[11] Although recent clinical reports have shown favourable results with minimally invasive repair of pectus excavatum (Nuss procedure), the Ravitch procedure is considered superior, especially when concomitant open heart surgery is performed via mid-sternotomy. With Nuss procedure, there is risk of sternal dehiscence from the pushing forces of the elastic bar in the anterior chest wall.[12]

Ravitch procedure, however, is associated with potential difficulties in approach for reoperation. Secondly, migration of the bars postoperatively can occur spontaneously or accidently during external cardiac resuscitation. The migrated bar may

injure the heart, lungs, or great vessels. Thus, if required, open rather external cardiac resuscitation should be performed.[12]

In conclusion, a simultaneous correction of pectus excavatum and the underlying cardiovascular diseases can be performed successfully. The technique described provides excellent access to the underlying cardiac lesion, stable chest wall and good cosmetic results.

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How to cite this article: Kansara B, Singh A, Girotra S, Iyer KS. Combined Bentall and modified Ravitch procedures in a patient with Marfan syndrome. J Anaesthesiol Clin Pharmacol 2013;29:95-8.
Source of Support: Nil, **Conflict of Interest:** None declared.