Case report - Vascular thoracic

Open door approach by partial sternotomy and sterno-costo-chondroplasty for annuloaortic ectasia with pectus excavatum

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Abstract

A 32-year-old female patient with Marfan syndrome was admitted for repair of annuloaortic ectasia and severe pectus excavatum. Because the chest cage deformity was severe, concomitant reparative surgery of the chest wall was performed. Partial median sternotomy and left second–fifth rib division was made to obtain good surgical field. The patient underwent valve-sparing aortic root remodeling successfully. After complete neutralization of heparin, additional division of the right ribs was performed and each rib and the sternum was reshaped. Pectus excavatum was completely repaired by this method. We believe this approach is efficacious for intracardiac repair with severe pectus excavatum.

Keywords: Pectus excavatum; Sterno-costo-chondroplasty; Marfan syndrome; Aortic root sparing operation

1. Introduction

In Marfan syndrome, various types of thoracic cage deformities are accompanied by heart disease and diverse surgical approaches are proposed. Pectus excavatum obstructs the surgical field because of abnormal deviation and rotation of the mediastinum. We report a case of Marfan syndrome with severe pectus excavatum and annuloaortic ectasia (AAE) successfully managed with a unique approach.

2. Case presentation

The patient was a 32-year-old female who had AAE and aortic regurgitation. She was referred to our Cardiovascular Division for heart murmur and an abnormal chest roentgenogram during pregnancy. Her family history indicated multiple cardiovascular events suggesting acute aortic dissection in her relatives and siblings. She had arachnodactyly, scoliosis and pectus excavatum. Her height and weight were 172 cm and 50 kg, respectively. Echocardiogram indicated aortic root dilatation of 49 mm in diameter and moderate aortic regurgitation. Magnetic resonance imaging (MRI) of the chest revealed a flat chest cage, pectus excavatum and a deviation of mediastinal organs to the left hemithorax. The minimal distance between the sternum and the vertebral column was 12.5 mm. Computed tomography (CT) Haller-index was 15.0 [1] (Fig. 1). Repair of AAE was conducted 12 months after she gave birth by normal delivery. Spirometry demonstrated her vital capacity was decreased to 57% of predicted value. Aortography showed AAE, moderate aortic regurgitation, dilated left ventricle with diffuse hypokinesis. The left ventricular ejection fraction was 33.8%. Concomitant surgery for AAE and pectus excavatum was performed.

Left partial sternotomy was performed from second left intercostal space to the xiphoid process. The soft tissue of the left chest including the major pectoral muscle was dissected and retracted to the left side. Left ribs from third to sixth rib were divided at the mid-clavicular line, where the angulation of the rib was acute. The left internal thoracic artery and intercostal arteries were preserved. Then a retractor was applied to the sternum, so that a good exposure of heart and great vessels was obtained (Fig. 2). Cardiopulmonary bypass was instituted by aortic cannulation and bicaval venous drainage. The aortic root was severely dilated but aortic cusps had normal morphology. Aortic root reconstruction was performed using Yacoub's remodeling technique [2] with a 26 mm albumin coated Dacron graft. After weaning from the extracorporeal circulation, heparin was neutralized and hemostasis was obtained. Repair of pectus excavatum was then performed. The sternum was transected and the right fifth and sixth ribs were cut and reshaped at the midclavicular line. Divided ends of left ribs were trimmed in a wedge shaped manner and reattached by bioabsorbable cords (PDS-CORD®, Ethicon, Inc, Somerville, NJ, USA). The sternal edge was cut in a wedge shape and the sternum was closed with three bioabsorbable pins (Super FIXSOR®), Takiron Co., Ltd., Osaka, Japan) and PDS-CORDs to reinforce the sternum (Fig. 2c). By this procedure, a flat and stable chest cage was obtained.

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The postoperative course was stable and the endotracheal tube was extubated on postoperative day 1. The length of intensive care unit (ICU) stay was two days. The postoperative echocardiography revealed good coaptation of the aortic cusps with no aortic regurgitation. The postoperative CT-scan demonstrated corrected deviation of the mediastinum (Fig. 1b). Middle term follow-up at three years revealed normal function of the aortic valve and a flat but not deformed thoracic cage.

3. Discussion

Several problems should be solved in adult cardiac surgery associated with pectus excavatum. Mild degree pectus excavatum does not obstruct the surgical field and routine median sternotomy incision is possible. However, moderate or severe pectus excavatum makes the heart and great vessels deviate to the left hemithorax and it is difficult to obtain a good surgical field. Moreover, risk of cardiac compartment syndrome due to compression from the depressed sternum should be considered after open-heart surgery. Complicated cardiac repair requires longer bypass time with prolonged myocardial ischemia and edema of the myocardium which further exacerbates cardiac function, thereby creating a vicious cycle.

The classical approach for pectus excavatum described by Ravitch is accompanied by extensive dissection of the sternum and affected ribs [3]. Long-time internal fixation by ventilator or metallic bar support of the sternum is necessary. Javangula and colleagues reported a use of Goretex strips (W.L. Gore & Associates, Inc, Flagstaff, AZ, USA) as an alternative to metallic bar [4]. Pevni and colleagues reported concomitant coronary artery bypass grafting and sterno-chondro-plasty using a transverse wedge resection of the sternum and bilateral subperichondrial rib resection [5]. In their method, extensive dissection of the anterior musculoskeletal system is necessary so there is a concern for bleeding after complicated aortic root repair. The implantation of the pectus bar advocated by Nuss is the revolutionary minimally-invasive procedure for repair of pectus excavatum in children [6]. Concomitant surgery for the repair of congenital heart anomaly and pectus bar implantations is efficacious in children. However, short- and long-term outcome of the Nuss procedure for adult patients is unknown. Sternal turnover has been reported as a useful procedure for correction of pectus excavatum [7]. Concomitant Bentall operation and sternal turnover was reported in 1983 [8]. A good surgical field for open-heart surgery may be attained by sternal turnover. The most serious complication of sternal turnover is ischemic bone necrosis and resultant osteomyelitis [9]. Chest wall infection is a life threatening complication causing mediastinitis, graft infection and detachment of graft. Fatal massive bleeding may occur. Shiraiishi and colleagues reported simultaneous Bentall operation and modified Ravitch procedure with a Gore-Tex sheet [10]. In the present case, we opened the partially divided sternum and right ribs like a door to expose the deviated mediastinum. Blood flow from the left internal thoracic artery and intercostal arteries were preserved to avoid ischemic necrosis of a dissected piece of the thoracic cage. A wedge resection and refixation of ribs and the sternum enabled us to make a flat anterior chest. The use of bioabsorbable pins and cords helped to reinforce and stabilize the chest cage and sternum. This procedure enabled routine postoperative
respiratory care to be administered with short-time ventilator support. The cosmetic correction of the chest cage was satisfactory.

In conclusion, the open door approach is efficacious for concomitant operation for pectus excavatum and complex heart repair.

References