Work in progress report - Vascular thoracic

An alternative surgical approach for the combined treatment of pectus excavatum and acute aortic dissection type-A in Marfan syndrome

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Abstract

Acute aortic dissection type-A (AADA) is a life-threatening condition especially in patients with Marfan syndrome (MFS) simultaneously suffering from severe pectus excavatum (PE). We report on emergency surgery for combined treatment of PE and AADA in a patient with MFS using an alternative approach. It leads to excellent exposure of the dislocated heart and great vessels enabling Bentall procedure followed by funnel chest repair with modified technique of Adkins and Blades. We achieved favorable functional and cosmetic results. Therefore, we conclude the surgical approach presented is feasible for standard treatment of AADA and consecutive repair of PE.

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1. Introduction

Acute aortic dissection type-A (AADA) is a life-threatening condition, still associated with significant morbidity and mortality. This applies especially to patients with Marfan syndrome (MFS) [1], who often (approx. two third of all Marfan patients) also present with chest-wall deformities, such as pectus excavatum (PE) [2, 3]. In severe cases of PE, median sternotomy is an unsuitable approach for open-heart surgery, since the heart and great vessels are displaced into the left hemithorax. We report on an emergency operation for AADA and simultaneous correction of concomitant severe PE in a Marfan patient using an alternative approach.

2. Clinical summary

A 28-year-old female patient with MFS presented with retrosternal pain radiating to her spine. A computed tomography (CT)-scan revealed an ascending aortic aneurysm (7.9×7.1 cm), displaced into the left hemithorax, combined with an AADA (Fig. 1a,b), and a severe PE with minimal distance from sternum to column vertebrae of 7 mm (Fig. 2a). Initially, she refused surgical treatment. Echocardiography revealed aortic valve insufficiency grade III. With increasing dyspnea and discomfort, the patient agreed to an operation.

Extracorporeal circulation was established via femoral vessels. A longitudinal midline incision upon the sternum with mobilization of the left pectorals was undertaken. We then proceeded with a left-sided costotomy of ribs two to five and placed a normal chest retractor providing excellent exposure of the heart and the ascending aorta (Fig. 3a,b). The aorta was clamped directly proximal to the innominate artery where aorta had normal diameter. The dissection started at the left coronary artery and was limited to the ascending aorta (classified DeBakey type-II). Inspection of the aortic valve revealed severely enlarged cusps with fenestrations, not suitable for valve preserving reimplantation. After implantation of a composite graft for replacement of the aortic valve and the ascending aorta [St. Jude Medical (SJM) Conduit, 33 mm] and intraoperative control via transesophageal echocardiogram, the patient was decannulated and heparin was antagonized.

Thereafter, we started with funnel chest repair using an individualized strategy: as was done previously on the contra-lateral side we mobilized the right pectorals followed by right-sided costotomy of ribs two to five. The corpus sterni was separated from the manubrium sterni with transversal sternotomy, insertions of ribs were partially resected and the corpus was removed. After transversal insertion of a slightly curved stainless steel bar (Sulamaa-strut, 16 cm) and its fixaton to ribs with wires, corpus sterni was repositioned in an inverted fashion on top of the strut. Manubrium sterni and corpus sterni were conjoined with wires and conjunction to the ribs was established via 2.0-vicryl after insertion of two pleural drainages, one on each side. At last wound-closure was performed with single buttons sutures. Bypass-time was 166 min, aortic cross-clamp-time 98 min and the procedure
took 249 min. Two thousand one hundred milliliter packed red blood cells, 900 ml fresh frozen plasma and 400 ml thrombocytic concentrate were transfused. The patient’s recovery was uneventful. Physical examination and CT-scan (Figs. 2b and 4) revealed a favorable functional as well as cosmetic result. Meanwhile we have successfully operated on a second patient with the same method.

3. Discussion

Surgery for AADA in patients with severe PE remains a challenge. In view of the extreme lateral displacement of the heart, median sternotomy appeared unsuitable for this patient. Combined corrections of chest-wall deformities and intracardiac lesions have been reported previously [4, 5]. However, a review of the literature has yielded only two reports on combined repair of AADA and severe PE in Marfan patients. Molina et al. described extension of a midline-sternotomy with an anterior lateral thoracotomy into the left second and sixth intercostal space thereby enabling access through a ‘trap door’ [6]. Similarly, Nisamoglu et al. combined partial upper sternotomy with left anterior thoracotomy providing access to the heart [7]. Both cases were performed without simultaneous correction of PE. Javangula et al. described a one-stage operation for elective aortic root replacement and PE in MFS performing different skin-incision and PE correction [8]. For funnel chest repair minimally-invasive technique regarding to Nuss et al. is widely used [9]. In this case, minimally-invasive strategy was not amenable due to previous correction of AADA and because of the patient’s age. In addition, there are two more alternatives for funnel chest repair, the sternal turnover procedure [10] and the procedure with transverse skin incision [11].

The approach presented here, avoids longitudinal division of the sternum completely enabling correction of PE with a modified technique of Adkins and Blades [12], a formation...

Fig. 1. (a) Computed tomography scans show acute aortic dissection type-A and (b) which is limited to the aneurysmal ascending aorta.

Fig. 2. (a) Transversal computed tomography underlines severe pectus excavatum before correction and (b) successful stabilization of the sternum with a Sulamaa-strut.
Incision of the left margin of the funnel chest, costotomy of ribs two to five and placement of a normal chest retractor provided excellent exposure of the dissected ascending aortic aneurysm.

Preoperative and postoperative three dimensional-CT-reconstruction of a patient with Marfan syndrome suffering from severe pectus excavatum.

of Ravitch’s and Sulamaa’s method [13, 14]. Furthermore, it provides excellent exposure of the ascending aorta and the aortic arch. Our strategy permitting both procedures in one surgical session appears simple and effective: the primary costotomy along the left margin of the funnel provided excellent exposure to the heart and enabled repair of the dissected aortic root. Thereafter, the incision was used for subsequent funnel chest repair.

Our approach is limited by the patient’s clinical situation and it must be mentioned that this case represents a specific situation: a clinically stable patient with circumscriptive aortic dissection DeBakey type-II. In unstable situations the operative strategy must follow the urgency of the intervention; therefore, a time-consuming cosmetic approach may not be indicated to save the patient’s life.

The presented surgical approach for combined treatment of AADA and PE in patients with MFS, enables excellent exposure of the dislocated heart and great vessels, is feasible for standard treatment of AADA and consecutive repair of PE and leads to favourable functional and cosmetic results.

References