Hybrid Treatment for Thoracoabdominal Aortic Aneurysms in Patients with Marfan Syndrome

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Marfan syndrome is a heritable disorder of connective tissue leading to aortic aneurysms and other cardiovascular complications associated with reduced life expectancy. Although contemporary management of ascending aortic disease requires open surgical reconstruction, the combined retrograde visceral revascularization and endovascular exclusion (hybrid procedure) of entire thoracoabdominal aorta has been introduced for the management of descending thoracic and abdominal aortic pathology. The present experience reports 2 cases of thoracoabdominal aortic aneurysms, in Marfan patients, previously submitted to major cardiovascular surgical procedures, through a hybrid approach.

Marfan syndrome (MS) is a connective tissue disorder characterized by ocular, cardiovascular, and skeletal manifestations, consisting of a genetic defect in the fibrillin gene located on chromosome 15q,[1] causing degenerative changes, both structural and functional to aorta, mitral, and tricuspid valve.[2]

The complications of aortic degeneration, as dissection, aneurysm and rupture, are the leading causes of death.[3]

When ascending aorta is involved by the disease, the initial elective procedure is usually repair of the aortic root and ascending aorta with a cardiovascular surgical procedure.[4]

Management of complications such as aortic rupture and aortic dissection represents a major challenge for surgeons as the gold standard treatment for patients with MS is total replacement of the aneurysmatic aorta through open approach. This technique, however, is burdened by high mortality and complication rates.

Hybrid approach through a retrograde visceral vessels revascularization, with subsequent endovascular exclusion of entire aorta, was introduced as an alternative treatment, to open surgery, in patients who have already undergone other major interventions, especially in prohibitive high-risk patients, as usual in Marfan patients.[5]

CASE REPORT

The present experience describes 2 hybrid procedures for the treatment of complex thoracoabdominal aortic aneurysm (TAAA) in Marfan patients.

Case 1

Woman, 46-year-old, hypertensive Marfan patient, affected by cerebral aneurysm without surgical indication according to the specialist because of small entity.

In 2000, she was submitted to replacement of aortic valve with Bentall operation, because of valvular insufficiency and ascending aorta dissection, with uneventful postoperative course (Table I).

In April 2007, she had a new dissection; a descending aorta entry tear and retrograde aortic dissection were detected, involving aortic arch and descending aorta. She was submitted to endovascular coverage of the descending aorta entry tear (GoreTag 20 × 150 and 31 × 150), with a regular postoperative course (Fig. 1A, B).

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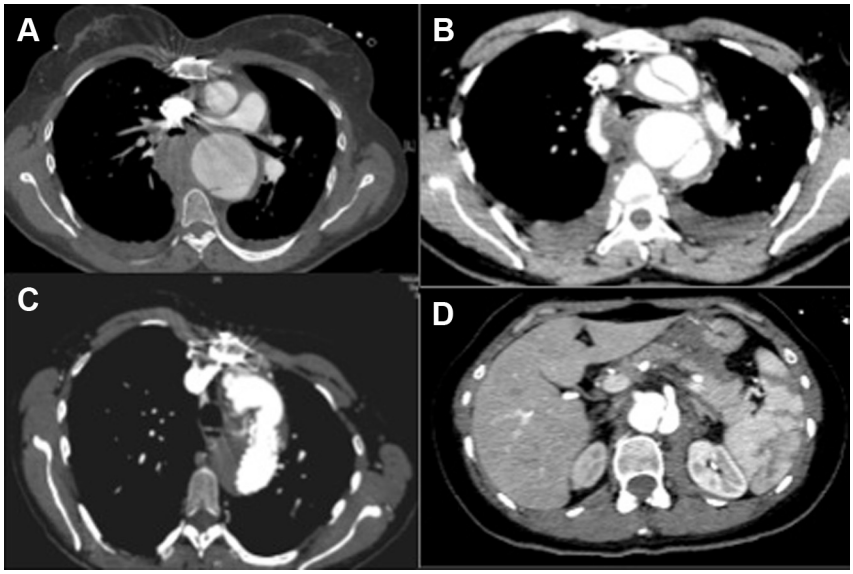

Fig. 1. Case 1: **(A)** Replacement of aortic valve and ascending aorta with Bentall operation and a new entry tear in the descending aorta (April 2007). **(B)** Descending aorta entry tear and retrograde aortic dissection until distal ascending aorta (April 2007). **(C)** Ascending aorta and arch replacement (May 2007). **(D)** Disease progression with false lumen enlargement (August 2007).

In May 2007, she had a progression of retrograde dissection involving supra-aortic vessels, treated by means of an open replacement of the distal portion of ascending aorta and arch and supra-aortic vessels reimplantation. (Fig. 1C)

In August 2007, a rapid false lumen expansion determined an intractable pain (Fig. 1D), which required the extension of the coverage of thoracoabdominal aorta until the subrenal aorta pre-carrefour (GoreTag, Flagstaff, AZ; 34 × 200) and stenting of isolated right common iliac artery dissection (Wallstent, Boston Scientific, Natick, Ma) and an open visceral debranching from left common iliac artery (K-Dacron 14 × 7mm, Intergard Heparin graft, Maquet Cardiovascular LLC, San Jose, CA; Fig. 2A, B).

The postoperative course was charged by the asymptomatic occlusion of celiac axis branch revealed by transient increase of hepatic enzymes (Fig. 2B).

The patient was submitted to angio computed tomography (CT) at 1 month and followed by clinical and echographic examinations at 1, 3, and 6 months and yearly thereafter.

In March 2012, a left renal stenosis was detected during an echographic examination, and therefore, the patient had an angiographic control and subsequent successful renal stenting.

Six months later, the patient had a left superoexternal quadrantectomy with lymphadenectomy.

In August 2013, for penetrating chest pain and anemia, thoracic and abdomen angio CT has been performed, showing the presence of thoracic type III endoleak (Fig. 2C), with an evidence of enlargement and a persistent abdominal patency of the false lumen in correspondence with the distal endoprosthesis landing.

Table I. Case 1 surgical history

Year	Procedures
2000	Replacement of aortic valve and ascending aorta with Bentall operation
2007 April	Coverage of the descending aorta entry tear
2007 May	Open replacement of ascending aorta and arch and supra-aortic vessels reimplantation
2007 August	Coverage of thoracoabdominal aorta, stenting of right common iliac artery, and open visceral debranching from left common iliac artery
2012 March	Renal stenting
2013 August	Emergency endovascular exclusion of type III endoleak and endovascular exclusion of the false lumen with glue and vascular plugs

Therefore, it was performed, an emergency endovascular exclusion of such endoleak (36 × 200 mm Valiant; Medtronic Vascular, Santa Rosa, CA). An additional endovascular exclusion of the distal false lumen with glue and vascular plugs was obtained through a right transfemoral access (Fig. 2D, E). The patient is still in good health (Fig. 2F).

Case 2

A 42-year-old woman, current smoker, suffering from hypertension, hypochromic microcytic anemia, nonspecific