CASE REPORT

Hybrid endovascular treatment of subclavian artery aneurysm associated with Marfan's syndrome: case report

André de Araújo Sacchi[I]; Alzumar Zacarias de Medeiros[II]; Roberto Ribeiro Filho[III]
[I]MSc. Assistant professor, Department of Surgery, Universidade Federal de Campina Grande (UFCG), Campina Grande, PB, Brazil. Specialist in Vascular and Endovascular Surgery, SBACV. Head, Endovascular Surgery Service, Hospital Antonio Targino, Campina Grande, PB, Brazil.
[II]Assistant professor, Department of Surgery, Universidade Federal de Campina Grande (UFCG), Campina Grande, PB, Brazil. Assistant physician, Endovascular Surgery Service, Hospital Antonio Targino, Campina Grande, PB, Brazil. Specialist in Vascular Surgery, SBACV.
[III]Assistant physician, Endovascular Surgery Service, Hospital Antonio Targino, Campina Grande, PB, Brazil. Specialist in Vascular Surgery, SBACV.

Correspondence

J Vasc Bras. 2008;7(2):144-149.

ABSTRACT

We report on a 46-year-old female patient with Marfan's syndrome and a right subclavian artery aneurysm following open repair of type A aortic dissection. The patient was treated with a hybrid approach, combining innominate to right common carotid stent grafting and carotid to right axillary artery bypass. The postoperative course was uneventful and a duplex ultrasound confirmed successful aneurysm exclusion.

Keywords: Subclavian artery, Marfan's syndrome, endovascular procedures, stent graft.

RESUMO

Descreve-se o caso de uma paciente feminina de 46 anos com síndrome de Marfan que foi diagnosticada com aneurisma da artéria subclávia direita após cirurgia aberta para reparo de dissecção aórtica tipo A. A paciente foi tratada por abordagem híbrida, que combinou o implante de uma endoprótese recoberta da artéria inominada para a carótida comum direita com uma ponte carótida para a artéria axilar direita. O pós-operatório transcorreu sem complicações, com a confirmação, por ultra-som, do sucesso da exclusão do aneurisma.

Palavras-chave: Artéria subclávia, síndrome de Marfan, procedimento endovascular, endoprótese.

Introduction

Subclavian artery aneurysms (SAA) are rare (< 1%) in relation to all peripheral aneurysms.[1] They can be asymptomatic, presenting only pulsatile mass in the supraclavicular region, or complicated, progressing to distal embolization, thrombosis or compression of neighboring structures. If left untreated, SAA may lead to rupture or ischemia of the affected limb.[2]

Conventional therapy involves aneurysm exclusion. However, in patients previously submitted to a median sternotomy, the procedure becomes a challenge. The endovascular treatment has been indicated in subclavian artery diseases, such as in cases of traumatic lesion aneurysms, with the aim of performing a minimally invasive procedure.[3,4]

The case reported herein is of a patient with Marfan's syndrome diagnosed with an asymptomatic aneurysm of the right subclavian artery after open surgery of type A aortic dissection. This article discusses the hybrid technique in the treatment of right subclavian artery aneurysm.

Clinical case

A 46-year-old female patient with Marfan's syndrome was diagnosed with asymptomatic aneurysm of the right subclavian artery measuring 3 cm in diameter. The patient has previous history of Stanford type A acute aortic dissection (DeBakey Type I) treated three years ago with open surgical repair, including aortic valve replacement with biological graft and ascending aortic interposition with Dacron graft (28 mm).

A transthoracic echography was performed during follow-up, revealing biological aortic valve with no signs of reflux, presence of chronic dissection from the valve tube in the ascending aorta until the proximal abdominal aorta with patent communication between the real and the false lumen (Figures 1 and 2), persistence of intimal *flap* at the subclavian artery and left common carotid artery origin, and absence of brachiocephalic trunk identification. Doppler color-flow ultrasound showed dilatation of the right subclavian artery and reduction in flow velocity in the left external and internal carotid arteries with systolic velocity of 40 and 35 cm/s and diastolic velocity of 15 and 5 cm/s, respectively. Computed angiographic tomography using multiple detectors with volume acquisition and 2.5 mm-thickness multiple-plane reconstruction confirmed a 5 cm-long and 3 cm-diameter fusiform dilatation of the right subclavian artery (Figures 3 and 4). Digital subtraction angiography of the supraaortic and right subclavian trunk was performed, revealing two sequential dilatations of the right subclavian artery, with the first starting close to the right common carotid artery emergence (Figure 5). The right and left vertebral arteries were identified in their origin (Figure 6). Physical examination of pulses was normal, with no signs of motor or sensitivity deficit. There was an impulsive mass in the right supraclavicular region.

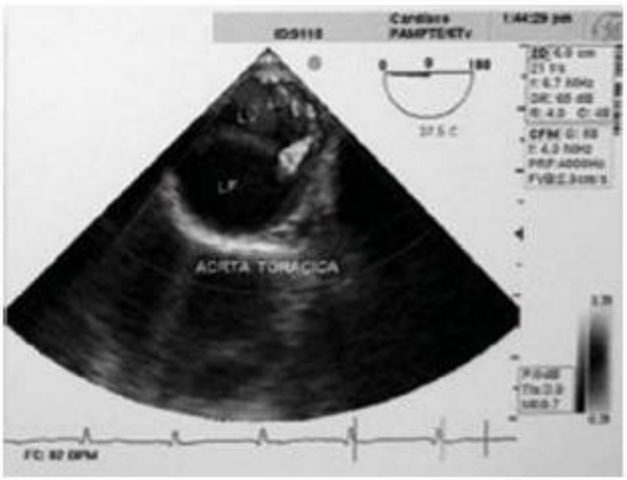

Figure 1 - Echocardiogram: aortic dissection (double lumen)

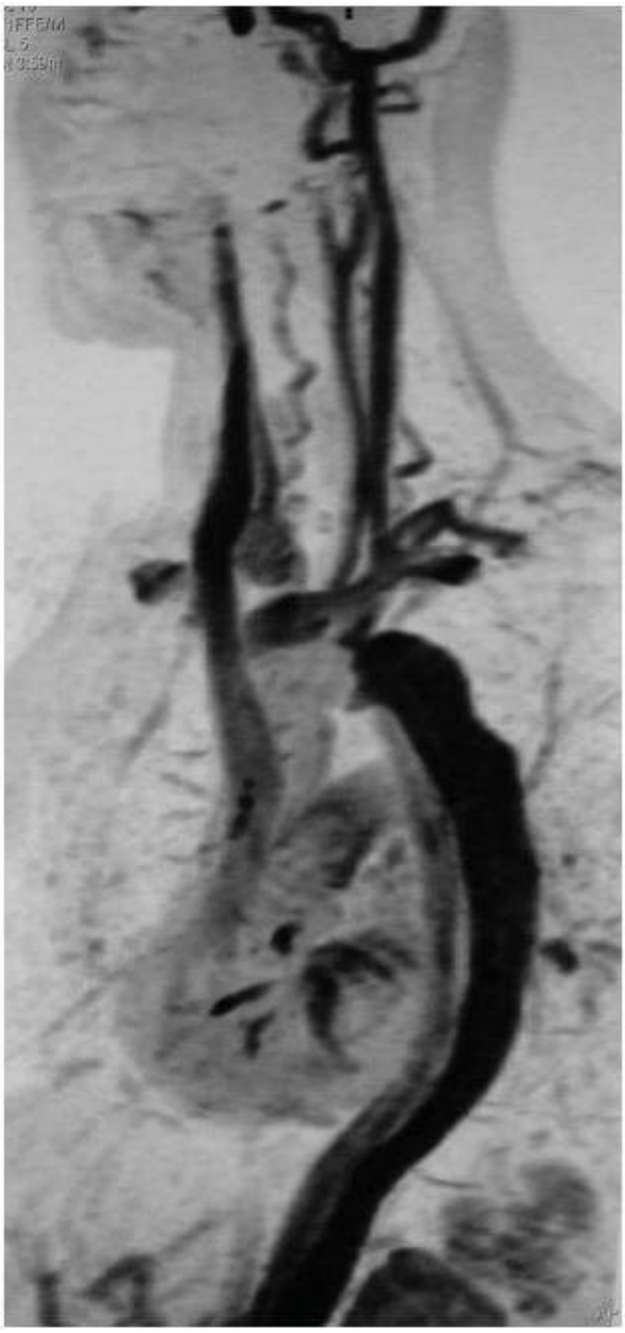

Figure 2 - Magnetic nuclear angiographic resonance: aortic dissection (double lumen)

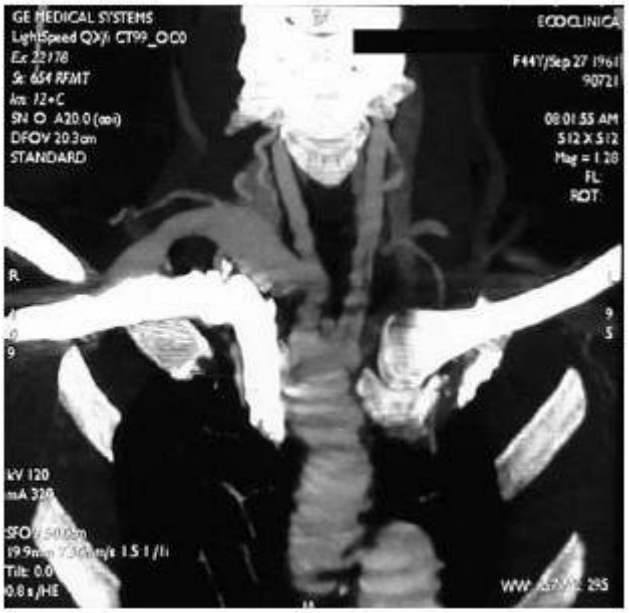

Figure 3 - Computed angiographic tomography: right subclavian artery aneurysm

Figure 4 - Computed angiographic tomography: right subclavian artery aneurysm

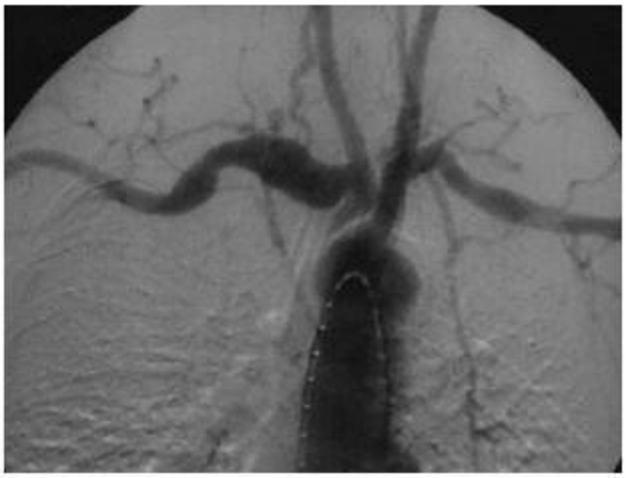

Figure 5 - Digital angiography (preoperative)

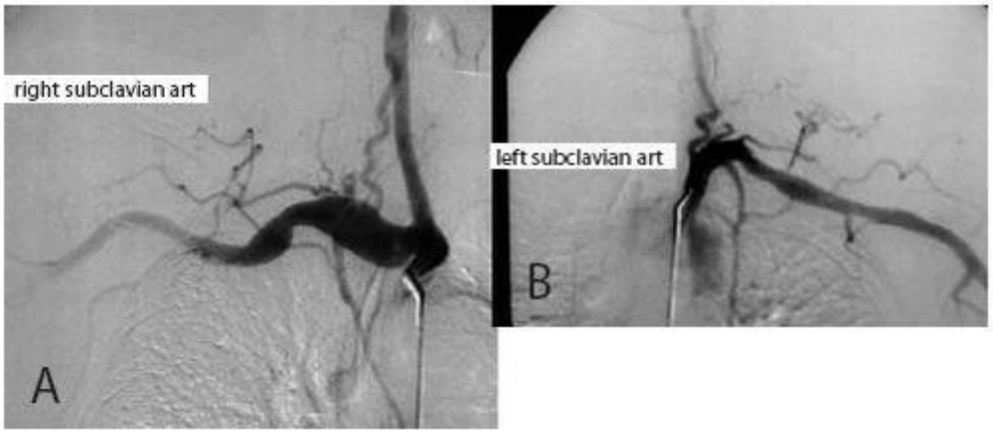

Figure 6 - A) Subclavian artery and right vertebral artery; B) subclavian artery and left vertebral artery.

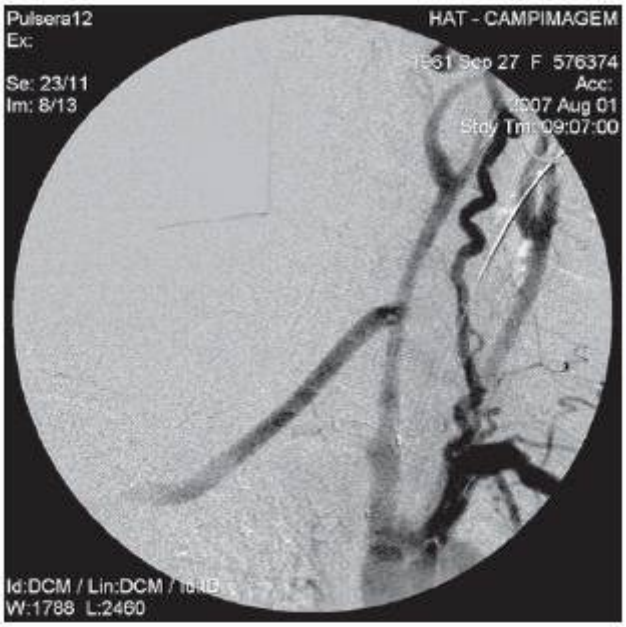

Figure 7 - Postoperative period (right axillary carotid bypass and common carotid BCT endograft).

A hybrid surgical treatment was indicated, with exclusion of the subclavian artery origin by endovascular treatment. A supraclavicular cross-sectional incision was performed, exposing and isolating the subclavian and right common carotid arteries. The costocervical and thyrocervical trunks and the internal thoracic artery were isolated and ligated. After identification of patent flow in both vertebral arteries using color-flow Doppler ultrasound and digital angiography, choice of treatment was ligation of the right vertebral artery. Systemic heparinization (70 IU/kg) was introduced. Right femoral percutaneous access was obtained for aortic arch angiography. An 8-mm ringed PTFE straight tubular graft was interposed, with terminolateral proximal anastomosis in the right common carotid artery. After the anastomosis was performed, a 30 cm-long 12F sheath was inserted (W.L. Gore & Associates, Flagstaff, Arizona, USA) through the tubular graft, using it to introduce an Amplatz guide wire (COOK) until the aortic arch. The rigid guide wire was used to insert a 16-mm x 10-mm, 7 cm-long GORE Excluder endoprosthesis - iliac extension (W.L. Gore & Associates, Flagstaff, Arizona, USA), extending from the brachiocephalic trunk origin until the common carotid artery, excluding the subclavian artery origin. The aneurysm distal segment was isolated, cross sectioning and ligating the subclavian artery next to the clavicle. A second infraclavicular dissection was performed to access the right axillary artery. The PFTE graft was inserted below the clavicle for terminoterminal right axillary artery anastomosis (Figure 7).

In the immediate postoperative period, the patient had palpable pulse for the whole right upper limb and mild complaints of parethesia in this limb and on the surgical site. She was discharged uneventfully and with no abnormalities, taking platelet antiaggregating agents (clopidogrel 75 mg/day + acetylsalicylic acid 100 mg/day).

Control color-flow Doppler ultrasound performed 30 days after the surgery did not show any flow in the aneurysmal sac and confirmed patent flow with no change in systolic and diastolic velocity in the common, internal and external carotid segments, and in the PTFE graft. The patient reports improvement in symptoms present in the preoperative period.

Discussion

The current incidence of subclavian artery aneurysm is unknown. Dent et al. found only two (0.13%) cases of subclavian artery aneurysm in a literature review of 1,488 patients with atherosclerosis.[5]

Etiology is variable, and the most common causes of atherosclerosis are trauma and poststenotic dilatation in compressions present in the thoracic outlet syndrome.[6-8] However, these lesions are also associated with degenerative disorders of the connective tissue, such as cystic medial necrosis, Marfan's syndrome, Ehlers-Danlos syndrome, Turner's syndrome, giant-cell syndrome, and Takayasu's arteritis. In addition, genetic causes were identified, and there are three cases described in the literature.[9,10]

The classical treatment consists of prosthetic graft interposition with terminoterminal anastomosis using a simple or combined transthoracic supraclavicular access.[11,12] Surgical treatment of subclavian artery aneurysms in patients with connective tissue disease may result in serious complications, such as artery deterioration, especially in those submitted to previous procedure.[13] In a series of 13 cases using open surgery for subclavian artery aneurysm repair, postoperative complication rate was 46%.[6]

The endovascular treatment offers a minimally invasive alternative for this subgroup of patients. MacSweeney et al. seem to have been the first to use a covered stent to repair a true subclavian artery aneurysm.[14] This home-made stent consisted of a Dacron-covered stainless steel self-expanding "Z" stent. Hilfiker et al. described nine patients treated by the endovascular technique due to traumatic lesions of the subclavian artery through use of different types of materials.[15] Technical success was 100%, and patency was 100% in a 29-month follow-up.[16] Other authors described patency rates between 80 and 100%.[4,5,9-13,15-17]

Another application of the endovascular treatment documented in the literature was to exclude an iatrogenic pseudoaneurysm of the subclavian artery.[18,19] Schoder et al. described a series of 12 patients submitted to endovascular treatment with implantation of a covered stent for aneurysmal disease and subclavian artery trauma. Technical success was 100%.[4]

Our patient had a right subclavian artery aneurysm, comprehending the common carotid artery close to the brachiocephalic trunk bifurcation. Any open surgical procedure to treat an aneurysm in that location would require trans-sternal access. Considering previous sternotomy for the patient and her comorbidities, this option would not be favorable. In this case, choice was for the hybrid technique – endovascular treatment associated with open surgery. Using the endovascular technique, a covered endoprosthesis was implanted from the brachiocephalic trunk until the common carotid artery, covering the right subclavian artery ostium. The surgical technique used was a right carotid-axillary bypass. This approach has already been successfully performed by other authors in isolated or even bilateral aneurysms of the subclavian artery.[20,21]

Another treatment option is the insertion of an endoprosthesis in the subclavian artery, excluding the aneurysm from its proximal neck until the distal neck. This option brings execution difficulties, since in our case the proximal case was not long enough to anchor an endoprosthesis without occluding the right common carotid artery ostium.

Another issue to be considered is the difference in diameter between the brachiocephalic trunk and

the distal subclavian artery. There is no available device in the market having required measures that could be used. Even so, the right carotid artery would have to be revascularized, since it would be occluded by the endoprosthesis.

González et al. described a case of subclavian artery bilateral aneurysm in which they reimplanted the left vertebral artery before implanting the endoprosthesis, and on the right side, after a frustrated attempt, the vertebral artery was ligated with no signs of vertebrobasilar ischemia.[20] Our patient has her right vertebral artery ligated. A previous angiographic study can assess patency of the vertebral system, both right and left, in its origin and its junction to form the basilar artery. In less than 5% of angiographies, there is lack of communication between both vertebral arteries, with the vertebral artery ending in the posteroinferior cerebellar artery in 1% of cases, more frequently to the right. Documentation of intracranial circulation, with attention to the vertebrobasilar system, is crucial in procedures that involve the possibility of right or left vertebral artery ligation.[22,23] Color-flow Doppler ultrasound confirmed flow and non-variation of its direction. Digital angiography identified flow in both vertebral arteries and, according to color-flow Doppler ultrasound, there was no change in flow direction or in systolic and diastolic velocity peaks. The patient progressed with no neurological signs or symptoms after the procedure.

Kasirajan et al. described two cases of endoprosthesis implantation in patients with Marfan's syndrome and cystic medial necrosis.[13] Two different types of endoprosthesis were used. The Wallgraft was used in the first case (Boston Scientific, Natick, MA, USA), and Viabahn in the second case (W.L Gore & Associates, Flagstaff, AZ, USA). After a 3-month follow-up, the patient with the Wallgraft thrombosed, whereas the patient who used Viabahn was patent. The Wallgraft rigidity and material – Elgiloy covered with polyethylene terephthalate – are not adequate for intraluminal use, thrombosing the treated segment, whereas the Viabahn, made of PTFE-covered nitinol, is more adequate for this purpose.

The relationship between Marfan's syndrome and idiopathic cystic medial necrosis has not been well defined. Vascular lesions in both disorders are microscopically similar.[24] Flexibility, adaptability for tortuous vessels and a more practical release system are important requirements when choosing the material for the endovascular treatment of the true subclavian artery aneurysm.

Conclusion

The combination of endoprosthesis covered from the brachiocephalic trunk to the right common carotid artery with right carotid-axillary bypass, even with ligation of the right vertebral artery ligation, made exclusion of the right subclavian artery aneurysm possible, preserving cerebral artery flow with minimal cerebral ischemia time.

The endoprosthesis used in our patient is commercially available in the market. The endovascular technique can be associated with the open surgery technique, becoming an alternative procedure for the treatment of subclavian artery aneurysm.

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Correspondence:
André de Araújo Sacchi
Rua Cônego Pequeno 615, Bairro Prata
CEP 58101-343 – Campina Grande, PB, Brazil
Tel.: (83) 3321.2442, (83) 3321.1976, (83) 9913.0509
Email: aasacchi@uol.com.br

No conflicts of interest declared concerning the publication of this case report.

Manuscript received November 5, 2007, accepted April 16, 2008.