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Case Report

Acute Aortic Dissection and Medullary Ischemia in a Patient with Marfan syndrome

Gustavo Henrique Belarmino Góes [*1], Eugenio Soares de Albuquerque [2], Diana Patricia Lamprea Sepulveda [3], José Maria Del Castillo [4], José Breno de Sousa Filho [5], Ricardo de Carvalho Lima [6], Dário Celestino Sobral Filho [7]

[1] Medical Student at Faculty of Medical Sciences, University of Pernambuco, Recife, Brazil. Cardiovascular Emergency Room of Pernambuco (PROCAPE / University of Pernambuco), Recife, Brazil; [gustavogoesmt@gmail.com]
[2] Echocardiographer and Preceptor of the Cardiology Medical Residency of the Cardiovascular Emergency Room of Pernambuco (PROCAPE / University of Pernambuco), Recife, Brazil; [albuquerqueeugenio@gmail.com]
[3] Chief of the Ward of Valvular Diseases, Preceptor and Vice-Coordinator of the Cardiology Medical Residency of the Cardiovascular Emergency Room of Pernambuco (PROCAPE / University of Pernambuco), Recife, Brazil. Master in Cardiology (University of Pernambuco); [lampreadiana@gmail.com]
[4] CEO of the School of Ultrasound of Pernambuco (Ecope); Voluntary Professor of Specialization in Echocardiography of the Cardiovascular Emergency Room of Pernambuco (PROCAPE / University of Pernambuco), Recife, Brazil. Vice-President of Echocardiography of the Department of Cardiovascular Image of the Brazilian Society of Cardiology; Doctorate in Medicine (National University de La Plata); [castillojmd@gmail.com]
[5] Founding Member of the Latin-American Society of Interventional Cardiology, Assistant Professor at the University of Pernambuco, and Head of the Hemodynamics Laboratory of the Cardiovascular Emergency Room of Pernambuco (PROCAPE / University of Pernambuco), Recife, Brazil; [jbreno50cardio@hotmail.com]
[6] Associate Professor of Cardiology at the University of Pernambuco; Executive Director of the Cardiovascular Emergency Room of Pernambuco (PROCAPE / University of Pernambuco). Head of Cardiovascular Surgery Team of the Cardiovascular Emergency Room of Pernambuco, Recife, Brazil. Doctorate in Medicine (Federal University of São Paulo); [rclima@elogica.com.br]
[7] Associate Professor of Cardiology at the University of Pernambuco. Research Coordinator of the Cardiovascular Emergency Room of Pernambuco (PROCAPE / University of Pernambuco), Recife, Brazil. Fellow of the American College of Cardiology and the European Society of Cardiology; [dsobral@uol.com.br]

[*]Corresponding Author: Gustavo Góes; Arnóbio Marques Street, 310, Santo Amaro, 50100-130 - Recife, PE - Brasil; Email: gustavogoesmt@gmail.com

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Abstract

Marfan syndrome (MS) is an autosomal dominant inherited connective tissue disease. The systemic complications of this disorder occur due to osteo-articular, cardiovascular, and ophthalmologic alterations. The cardiovascular involvement of MS is characterized mainly by aortic root and arch changes, resulting in increased susceptibility for the development of aneurysms. In the present study, we describe a case of type B abdominal aortic dissection in a patient with MS followed by spinal cord ischemia that rapidly evolved to paraplegia. Although rare, medullary ischemia is associated with a poor prognosis, as noted in the present case, wherein the patient died.

Keywords: ***Genetic syndrome, cardiovascular surgery, acute aortic syndrome.***

Introduction

Marfan syndrome (MS) is an autosomal dominant inherited connective tissue disease with an estimated prevalence of 2 to 3 cases per 10,000 individuals.[1] It is caused by a mutation in *FBN1* that encodes the fibrillin-1 protein, the main component of microfibrils extracellular.[2] The systemic complications of this disorder occur due to osteo-articular, cardiovascular, and ophthalmologic alterations, such as articular hyperflexibility, arachnodactyly, ectopia lentis (subluxation of the lens), among others.

The cardiovascular involvement of MS is characterized mainly by aortic root and arch changes, resulting in increased susceptibility for the development of aneurysms.[3] Progressive dilatation of the

aortic root, potentially leading to dissection and rupture, is the main cause of death in patients with MS.[4,5]

In the present study, we describe a rare case of medullary ischemia secondary to abdominal aortic dissection (Stanford type B) 3 years after dissection of the ascending aorta (type A) in a young patient with MS.

Case report

A 25-year-old male patient was admitted to the Cardiovascular Emergency Room of Pernambuco (PROCAPE / University of Pernambuco) with a type A aortic dissection and severe aortic insufficiency and underwent urgent Bentall surgery for correction. During this period, he was also diagnosed with MS, presenting with dolicocephaly, a high and arched palate, tall stature, hyperflexibility of joints, and arachnodactyly. The patient was hypertensive and had a family history of MS (mother, sister, aunt, and uncle).

Three years later, he was again admitted to the Emergency Room due to high intensity pain in the thoracic region, associated with pallor and malaise. Computed tomography (CT; figure 1) revealed a type B aorta dissection and aortic arch aneurysm.

He underwent correction surgery with 3 endoprosthesis implants and reimplantation of vessels of the base via a Dacron® tube graft. During hospitalization, he developed infective endocarditis (IE) in the aortic valve, confirmed by transthoracic echocardiogram (TTE) (figure 2) and blood cultures *(Enterococcus)*. The patient received antimicrobial treatment for IE by *Enterococcus*. However, due to worsening of his clinical condition and persistence of fever, the therapeutic regimen was changed. Furthermore, a transesophageal echocardiogram (TEE) was performed, which revealed an abscess in the aortic mitotic region and mitral valve involvement, leading to severe mitral regurgitation, presence of vegetation in the aortic prosthesis, and significant left ventricular systolic dysfunction (figure 3). A progressive worsening of the clinical status was noticed each day, and the patient developed sudden severe thoracoabdominal transient pain following dorsal irradiation of the back. A CT scan of the chest and abdomen (figure 1) showed: a) dissection of the abdominal aorta (superior mesenteric artery emerging from the true light), but not of the celiac trunk, and gastrointestinal ischemia was suspected; b) reentry in the false light after emergence of the superior mesenteric artery; c) endoprosthesis in the thoracic aorta and partial thrombosis of the false lumen; d) distal thoracic aorta with a circular flap and reentry in the false light. The echocardiogram of the abdominal aorta showed flow in the true light, which was central to the false light and the presence of a reentry point (figure 4). The case was discussed with the surgical team, and clinical treatment was selected due to the severity of the condition and the clinical impossibility of further surgical intervention. The patient developed left lower limb folding and paresis in the right lower limb. Serial neurological examination revealed progressive paraparesis (evolving to paraplegia) over 12 hours along with bladder retention, suggesting a spinal cord injury caused by dissection of the thoracic aorta or epidural abscess due to endocarditis.

Magnetic resonance imaging of the thoracic spine was requested, but the patient continued to show clinical worsening, developing severe pain and abdominal distension, as well as respiratory discomfort, prior to death.

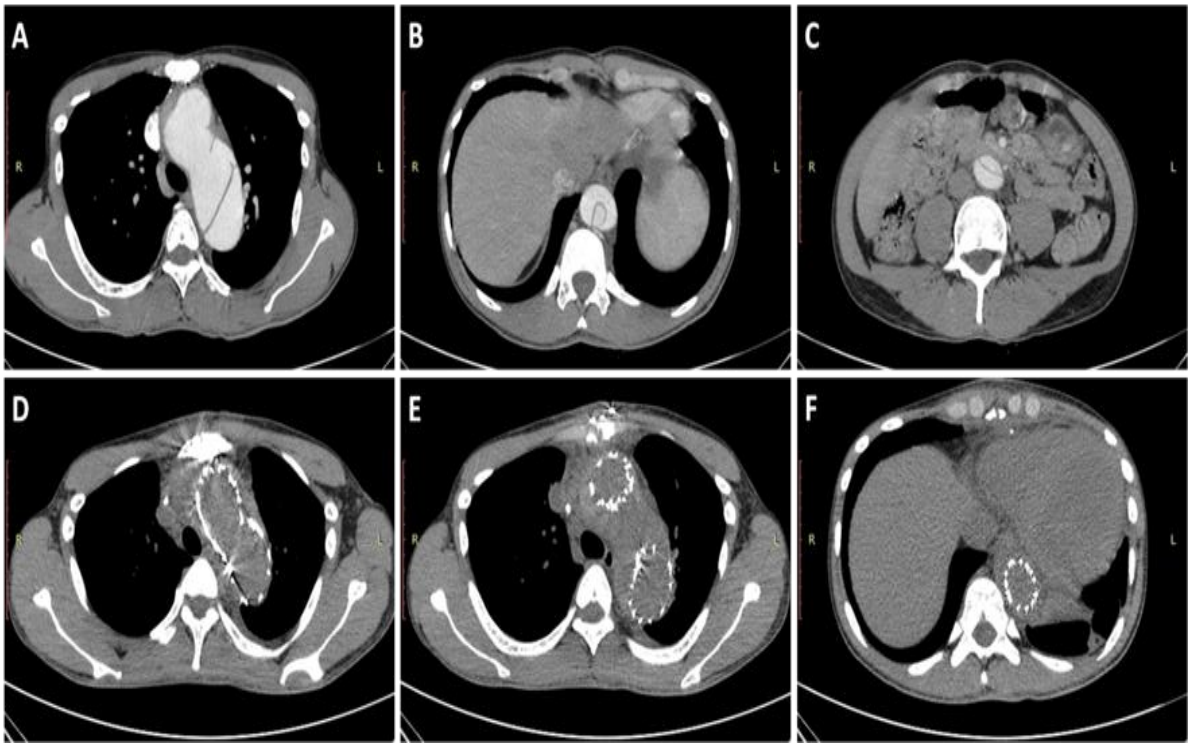

Figure 1: A, B and C. Dilated aorta diffusely presenting endoprosthesis in the ascending aorta, crossa and ascending thoracic aorta, with signs of aortic dissection in the thoracoabdominal transition. D, E and F. Presence of prosthesis in the aorta partially visualized in the thoracoabdominal transition, with signs of dissection that are observed until the emergence of the right renal artery.

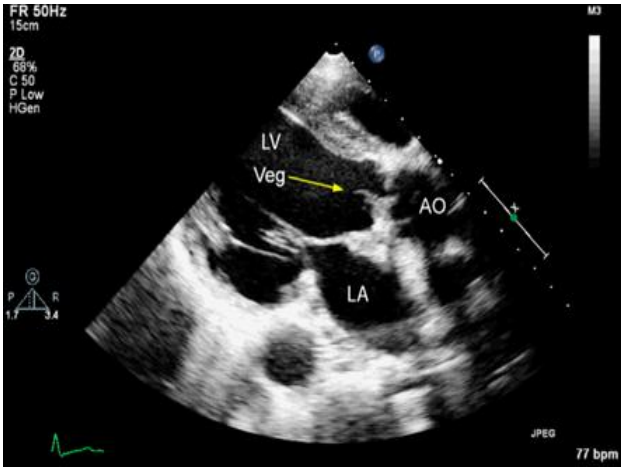

Figure 2: Transthoracic echocardiogram in longitudinal parasternal section showing vegetation (Veg) adhered to the aortic biological prosthesis projecting to the left ventricle (LV) exit pathway. AO: aorta; LA: left atrium.

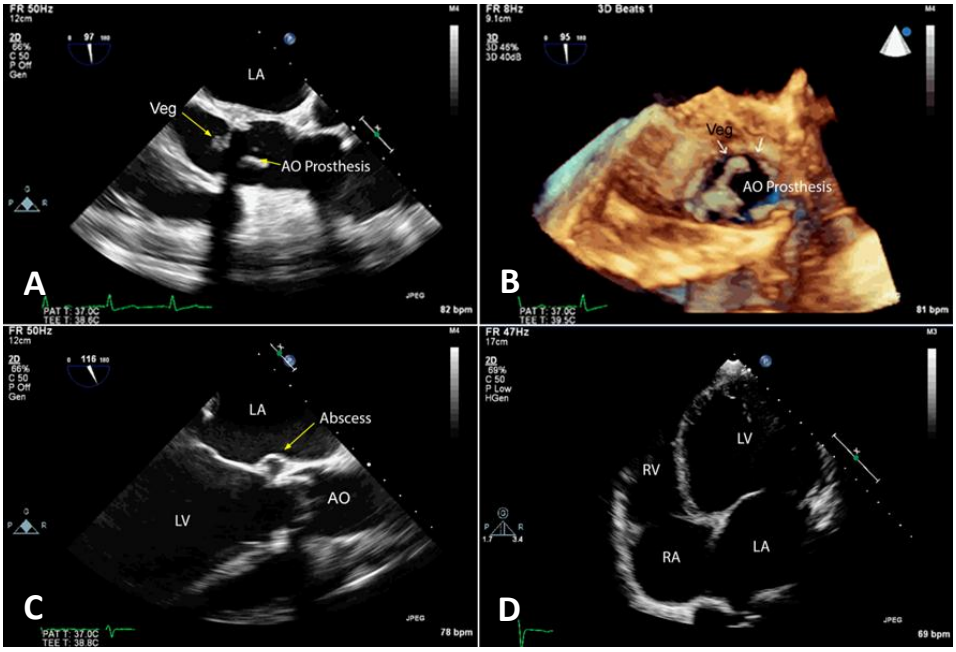

Figure 3: Transesophageal and transthoracic echocardiogram (TEE and TTE, respectively) in a patient with Marfan syndrome. A. Esophagus incised at 120° showing vegetation (Veg) adhered to the cusps of the aortic biological prosthesis (AO Prot); B. Three-dimensional base vessel transverse section TEE showing vegetation adhered to the leaflets of the aortic prosthesis; C. TEE of the middle esophagus at 120° showing the abscess of the mitro-aortic junction; D. TTE of the apical position of the 4 chambers showing left ventricular and atrial dilatation due to systolic function deficit. LV: left ventricle; RV: right ventricle; LA: left atrium; RA: right atrium.

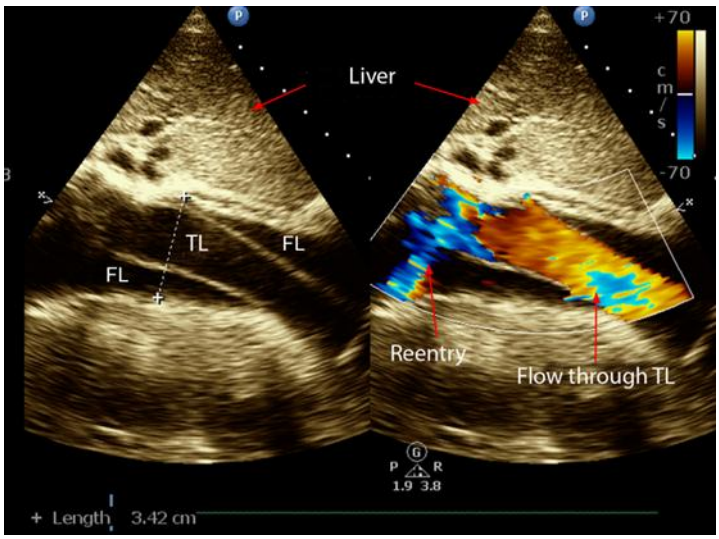

Figure 4: Echocardiogram of the upper abdominal aorta showing the longitudinal axis where the aortic artery is observed with the presence of a double dissection flap with color flow in the true light (TL) and reentry in the posterior region of the false light (FL).

Discussion

MS is a connective tissue disorder with pleiotropic manifestations that affect the lens, stimulate excessive growth of long bones, and dilation of the aortic artery, increasing the likelihood of dissection and rupture, resulting in a high mortality rate.[2] This dilation occurs in the ascending and descending aortic artery. It rarely affects the abdominal aorta, as reported in the present case.

Although MS presents with systemic involvement, the cardiovascular alterations are life-threatening, and the different aortic artery attacks are the main reason for the high mortality rates associated with the syndrome. Aortic dissections in patients with MS are usually type A, and the complications associated with this type of rupture can lead to symptoms, such as heart failure, syncope, signs of ischemia in other organs, focal neurological symptoms, cardiac tamponade, and even paraplegia.[6]

The examination of choice for diagnosis of aortic dissection is CT, which must be performed rapidly to provide essential information (extent and classification of dissection) to define the best therapeutic course. A meta-analysis of 1139 patients with aortic dissections showed that angiography using a multidetector had a sensitivity of 100% and specificity of 98%.[7]

The treatment of type A aortic dissections (more frequent in MS) is primarily surgical. The intervention should be immediate, with the objective being avoiding vessel rupture and consequent death by cardiac tamponade.[5] On the other hand, in type B dissections, the evolution is much more favorable with clinical treatment, with a mortality rate of 10% in 30 days, while in surgically treated patients mortality is 31%.[8]

The endovascular approach is usually not indicated for patients with MS due to poor aortic tissue quality and continuous dilatation, even after technically successful procedures.[9] Ágg et. al.[10] identified 3 extracardiac markers this indicate a high risk of aortic dissection: striae atrophy in the skin, transforming growth factor beta, and plasma activity of matrix metalloproteinases.

There is, therefore, the possibility of preventive surgery in the presence of an ascending aortic aneurysm in patients with the syndrome. The procedure consists of replacing the aorta with a low-porosity Dacron® conduit incorporated into a mechanical aortic valve, with the coronary arteries anastomosed to the graft.[6] The ischemic complications range from 16% to 34% and may involve any of the major arterial lateral branches, resulting in ischemia in the myocardium, brain, spinal cord, gastrointestinal tract, and/or limbs.[11,12]

Atypical presentations of aortic dissection may also occur, one of which is the development of acute paraplegia as the first symptom. Although rare, spinal cord injuries are associated with a considerably increased morbidity and mortality rates, as well as poor prognosis.[5]

Spinal cord ischemia may develop with paraplegia, resulting in loss of function and control of the bowel and bladder; spinal cord ischemia is one of the major complications of open or endovascular thoracic surgery. It is caused by an aortic repair or endovascular stent of relevant arteries that irrigate the spinal cord associated with other risk factors, such as hypotension and anemia. The severity of spinal cord ischemia symptoms depends on the area of the spinal cord affected. This can result in paraparesis or paraplegia, sensory deficits, ataxia, and bowel and bladder dysfunction. These symptoms may be transient or permanent.[13]

Medullary ischemia occurs due to obstruction of the intercostal and lumbar arteries, which supply the anterior spinal artery. The latter is also supplied by the root arteries, where blood flow is lower; therefore, this area is more susceptible to ischemia development. Since it occurs at a lower level, abdominal aortic dissection is required, as noted in the present case. Simultaneous mesenteric ischemia is responsible for severe abdominal pain and acute paraplegia.[5]

Placement of a graft or stent can lead to a reduction in spinal blood flow and can cause direct cytotoxic action within a short period of time. In addition, edema develops, accompanied by the development of the spinal cord compartment syndrome and increased pressure due to the action of these cytotoxic agents. Increased pressure, in turn, causes direct cell damage and reduced spinal perfusion.[14]

Conclusion

In the present study, we describe a case of type B abdominal aortic dissection in a patient with MS followed by spinal cord ischemia that rapidly evolved to paraplegia. Although rare, medullary ischemia is associated with a poor prognosis, as noted in the present case, wherein the patient died.

Conflicts of interest

The authors declare no conflicts of interest.

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Glossary of abbreviations

CT: Computed tomography
IE: Infective endocarditis
MS: Marfan syndrome
TEE: Transesophageal echocardiogram
TTE: Transthoracic echocardiogram