Aortic dissection in a patient with Marfan syndrome managed with multilayer flow modulator

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Marfan syndrome is a well-defined autosomal dominant disorder of connective tissue and the defective gene has been mapped to the fibrillin – 1 (FBN1) gene on chromosome 15. Mutations of these genes lead to defects in multiple organ systems among these systemic complications, those of cardiovascular system are the most critical because most deaths are due to aortic or mitral valve incompetence, aortic dissection, aneurysm rupture or mural complications of an ascending aortic aneurysm.

We present the case of a patient with Marfan syndrome in whom we success fully performed an aortic endovascular repair for a subacute complicated type B dissection, using multilayer technology.

A 27-years-old man with Marfan syndrome and a history of mitral valve replacement, presented with the chief complaint of substernal and back pain. The pain was moderately severe, intermittent, and associated with altered general condition. Patient history revealed that the was diagnosed with marfan syndrome at the age of 9. He had undergone replacement of the mitral valve by full sternotomy, 4 years ago.

Bidimensional transthoracic echocardiography showed a well-functioning prosthetic valve in the mitral position with out paravalvular leak or aortic regurgitation. Thorough examination of the arch and of the thoracic aorta revealed an image of intimal flap suggesting aortic dissection.

Computer tomography demonstrated the presence of an acute type B thoracic aortic dissection (TBAD), originating from the left subclavian artery and ending at the level of inferior mesenteric artery. Computerized fluid dynamics (CFD) techniques were employed to model the blood hemodynamics for this case. The patient under went a novel endovascular approach consisting of a total aortic repair using multilayer technology. The patient had 1, 6 and 12 months clinical and imagistic follow-up. The diameter of true and false lumen were calculated using a vascular analysis software package – CFD (MIMICS – Materialize) by NEXTCardio Research Center. Computerized fluid dynamics (CFD) analysis showed an expanded true lumen with the false lumen index decreasing at each follow-up. Velocity curves calculated with ANSYS (Canonsburg) showed decreasing pressure in false lumen with the appearance of a regular laminar flow in the aorta and all branches immediately after the procedure.

Conclusion: This case is the first reported of a TEVAR for subacute complicated TBAD in a patient with Marfan syndrome using a novel endovascular approach. These case was success fully treated. We were not faced with any complications post intervention. We believe that our case highlights the fact that multilayer aortic stents are safe options in the endovascular treatment of complex aortic dissection, with low mortality and good procedural success.
Abstract:
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We present the case of a patient with Marfan syndrome in whom we successfully performed an aortic endovascular repair for a subacute complicated type B dissection, using multilayer technology.
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Bidimensional transthoracic echocardiography showed a well-functioning prosthetic valve in the mitral position with out paravalvular leak or aortic regurgitation. Thorough examination of the arch and of the thoracic aorta revealed an image of intimal flap suggesting aortic dissection.
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Conclusion: This case is the first reported of a TEVAR for subacute complicated TBAD in a patient with Marfan syndrome using a novel endovascular approach. This case was successfully treated. We were not faced with any complications post intervention. We believe that our case highlights the fact that multilayer aortic stents are safe options in the endovascular treatment of complex aortic dissection, with low mortality and good procedural success.