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Aorta: Case Report

Intraoperative Type B Aortic Dissection With Malperfusion in a Patient With Marfan Syndrome

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We present the case of a 72-year-old man diagnosed with an aortic root aneurysm who was then diagnosed with Marfan syndrome. The patient suffered an intraoperative type B dissection with lower extremity malperfusion managed with an axillary-bifemoral extra-anatomic bypass.

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Intraoperative aortic dissection is a surgical emergency that carries a high rate of morbidity.[1] Risk factors include atherosclerosis, hypertension, and collagen disorders such as Marfan syndrome[1] caused by mutation of the gene encoding fibrillin 1 (*FBN1*). In general, these events involve the ascending aorta or transverse arch. Complicated dissection of the descending aorta with malperfusion to the lower extremities is now commonly treated with thoracic endovascular aortic repair or with traditional open repair when endovascular approaches are not feasible or appropriate, as in connective tissue conditions.[2] We present the rare case of an intraoperative descending aortic dissection with malperfusion in a patient with suspected Marfan syndrome.

A 72-year-old man presented to the aortic clinic with an incidental finding of an ascending aortic aneurysm. He denied symptoms of chest or back pain, and he was active. His family history was significant in that his daughter died of congestive heart failure secondary to an unknown mitral valve problem. Physical examination revealed a height of 188 cm and weight of 90 kg, with prominent facial features and scoliosis. He has a history of coronary artery disease and percutaneous coronary intervention.

Transthoracic echocardiography revealed enlarged aortic sinus segment at 4.8 cm, trileaflet aortic valve, and mild eccentric aortic regurgitation. Computed tomography angiography (CTA) revealed the following: sinus segment, 5.2 cm; ascending aorta, 3.4 cm; transverse arch, 3.3 cm; and descending thoracic aorta, 3.5 cm. A mild degree of aortic calcification was noted.

Given the physical examination findings and family history, genetic testing was performed. A 35-gene panel analysis (Invitae) identified a variant in the *FBN1* gene c.2644G>A (p.Ala882Thr) that was considered a variant of uncertain significance (VUS). The patient was also found to be a carrier of autosomal recessive homocystinuria.

Early surgical intervention was recommended over continued surveillance. Aortic root replacement was performed through a median sternotomy. The aorta was cannulated in the mid transverse aortic arch through purse-string sutures by Seldinger technique with a 21-mm cannula under transesophageal echocardiography (TEE) guidance. The ascending aorta was notably of normal size; therefore, an aortic root replacement with a porcine valve by modified Bentall technique was planned. A 27-mm aortic root (Freestyle; Medtronic) was implanted with coronary buttons created and reattached in standard fashion. We routinely use this graft in older patients; in our experience, it provides good durability and excellent hemodynamics. A distal anastomosis was performed directly to the ascending aorta with no need for circulatory arrest. Blood pressure readings in both arterial pressure monitors remained stable and near equivalent for the duration of the cross-clamp period.

At the conclusion of the procedure, the valve had normal function on inspection under TEE. At the completion of the study, the patient was noted to have a flap in the descending thoracic aorta (Figure 1A). Epiaortic ultrasound was used to assess the ascending aorta and transverse arch; no evidence of dissection or false lumen was noted. On imaging of the descending

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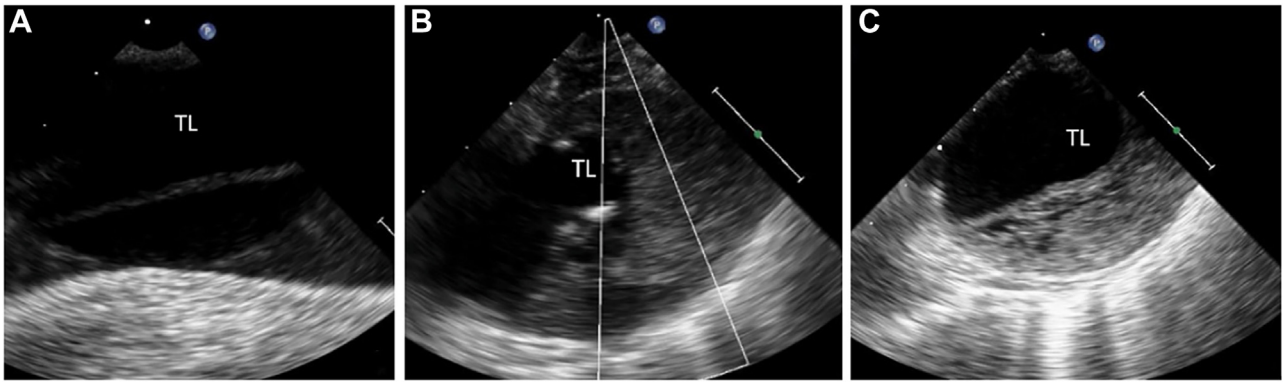

FIGURE 1 Transesophageal echocardiography depicting type B aortic dissection (A) after root replacement, (B) after administration of protamine with thrombus formation and true lumen (TL) compression, and (C) after axillary femoral bypass, identifying true lumen expansion.

aorta, there was notable color flow in the false lumen down to the level of the diaphragm. Conservative management was selected for this patient, given its confinement to the descending thoracic aorta. Protamine was administered, and the aorta was observed on TEE. Progressive thrombus formation was visualized in the false lumen of the descending thoracic aorta, with the true lumen becoming increasingly compressed (Figure 1B). The right femoral artery catheter became damped, reading a mean blood pressure of 20 mm Hg, while the right radial monitoring catheter remained stable at 70 mm Hg; these are routinely placed for monitoring in aorta cases.

Given the concern for lower extremity and visceral malperfusion, the common femoral arteries were exposed; there was no flow. Therefore, a right axillary bifemoral artery bypass was performed. At the conclusion of the procedure, palpable popliteal pulses had returned. The true lumen of the descending aorta was noted to have expanded with vigorous flow (Figure 1C; Video).

CTA performed on postoperative day 1 identified significant intramural hematoma of the descending aorta (Figures 2A, 2D) with compression of the true lumen. CTA was repeated on postoperative day 4; notably, the true lumen was expanded (Figures 2B, 2E) with recanalization of the celiac and superior mesenteric artery vessels. The extra-anatomic bypass was noted to have closed. The patient's postoperative course was otherwise unremarkable. CTA performed at 6 months postoperatively showed significant aortic remodeling (Figures 2C, 2F).

Genetic testing was offered to additional members of the family. Three new Marfan patients were identified, 2 of whom were children younger than 15 years, both with sinus of Valsalva aneurysms.

According to the genetic test report from Invitae, the c.2644G>A *FBN1* variant was not present in population databases, and this variant has been observed in individuals with clinical features of Marfan syndrome. Other variants affecting the same amino acid residue, including c.2645C>T (p.Ala882Val), have previously been determined to be pathogenic, suggesting that this amino acid residue is significant in the function of the *FBN1* gene and may have clinical effect when disrupted. This variant was later reclassified by Invitae as a pathogenic variant of *FBN1* gene.[3]

COMMENT

This case describes a new pathologic variant of the *FBN1* gene. It also highlights the value of close observation of clinical features for genetic disorders and demonstrates an alternative management strategy for surgeons facing intraoperative descending aortic dissection for which stent placement is less ideal.

This intraoperative aortic dissection flap could have been exacerbated by blood pressure fluctuations and anticoagulation. There is a possibility of an iatrogenic origin of the tear, given the use of Seldinger cannulation technique. In our case, the wire and cannula are threaded under TEE guidance to the distal aortic arch. There was no concern for dissection at the time of cannulation or afterward seen on TEE until the end of the case. It is reasonable to avoid any instrumentation inside the aorta in patients with known connective tissue disease. This patient had a VUS that was later reclassified to pathogenic. Many patients undergoing aortic surgery have a familial aortopathy. Surgeons should use caution with intraluminal access in these cases.

Our understanding of the genetics of aortic disease remains in relative infancy, and the role of genetic testing for patients with aneurysmal disease is undefined.[4] *FBN1* is typically associated with Marfan syndrome. In most patients, testing reveals a VUS,

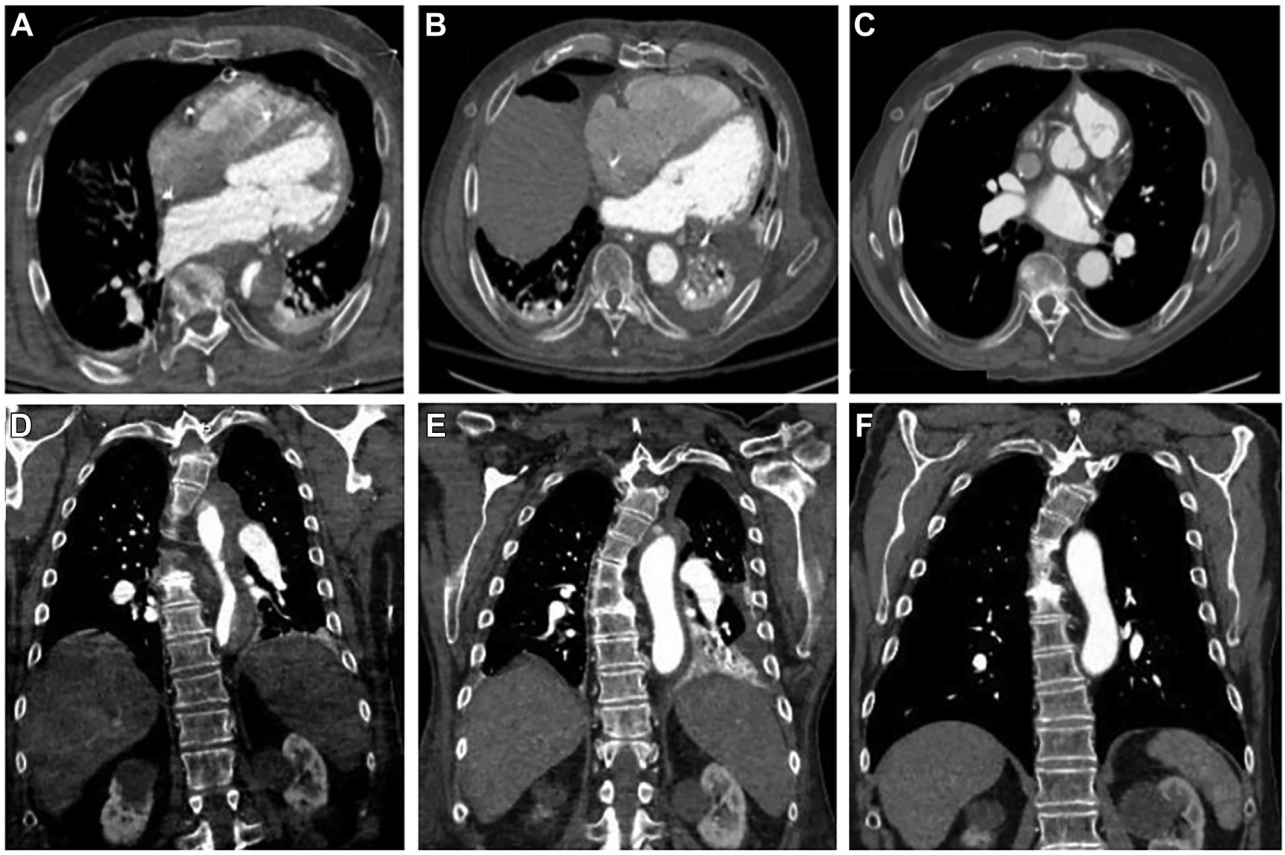

FIGURE 2 Computed tomography angiography depicting stages of remodeling: axial views (A) on postoperative day 1, (B) on day 4, and (C) at 6 months; and coronal views (D) on day 1, (E) on day 4, and (F) at 6 months.

placing great importance on clinical interpretation. In our practice, patients with significant family history, bicuspid aortic valves, or features of connective tissue disease are offered genetic testing. Patients with VUS in genes known to predispose to aneurysm and dissection are offered surgical intervention if an aneurysm has an absolute size of >5.0 cm or if indexed measures reach significance. Those with negative aortopathy test results who have strong family history are referred for more extensive genetic testing (typically the John Ritter Research Program).

An endovascular approach to this case would be met with obstacles such as minimal available imaging in a standard operating room. A frozen elephant trunk graft would be another option to perform under circulatory arrest. However, stent grafts are not generally recommended in Marfan patients according to expert consensus guidelines.[5] Traditional open approach would require repositioning of the patient and prolonged lower extremity ischemia. In this case, axillary to femoral artery bypass allowed reperfusion within 30 minutes and improved true lumen pressure and flow sufficient to allow aortic remodeling.

The Video can be viewed in the online version of this article [https://doi.org/10.1016/j.atssr.2023.12.006] on http://www.annalsthoracicsurgery.org.

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