Hybrid management of descending thoracic pseudoaneurysm in a patient with DiGeorge syndrome

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ABSTRACT

We present a case of a 42-year-old man with DiGeorge syndrome and congenital cardiac anomalies including a type B interrupted aortic arch who had previously undergone two bypasses between the ascending and descending thoracic aorta in childhood. He was found to have a 7.4-cm pseudoaneurysm of the descending thoracic aorta with the left subclavian artery arising from the aneurysm. The patient was treated with a single stage hybrid repair including left common carotid to subclavian bypass followed by thoracic endovascular aortic aneurysm repair. (J Vasc Surg Cases Innov Tech 2024;10:101535.)

Keywords: Aneurysm; Aorta; Cardiac; Thoracic; DiGeorge

DiGeorge syndrome (chromosome 22q11.2 microdeletion) is associated with congenital heart disease, with the most common cardiac defects being conotruncal anomalies, aortic arch anomalies, and ventricular septal defects (VSDs).[1,2] For young patients with aortic arch and proximal thoracic aortic pathology, open surgery is the gold standard.[3] However, endovascular and hybrid strategies with subclavian preservation are also accepted.[3] In this report, we describe a single-stage hybrid repair for a descending thoracic pseudoaneurysm in a patient with DiGeorge syndrome, aberrant anatomy, and significant surgical history. The patient provided informed consent for publication.

CASE REPORT

The patient is a 42-year-old man who presented with an asymptomatic 7.4 cm distal aortic arch pseudoaneurysm (Fig 1, *A-D*). He has a history of DiGeorge syndrome and congenital cardiac anomalies including type B interrupted aortic arch, double outlet right ventricle, and VSD requiring multiple operations in childhood. At 3 weeks, he underwent a left thoracotomy with placement of an 8-mm Gore-Tex graft from the proximal to distal thoracic aorta, ligation of a patent ductus arteriosus, and pulmonary artery banding. At the age of 1.5 years he underwent sternotomy, VSD and patent foramen ovale closure, and pulmonary artery debanding. At the age of 8 years, he underwent redo left thoracotomy with placement of a 19-mm Dacron graft between the ascending and descending thoracic aorta, approximately 2 cm from the distal anastomosis of his prior graft, with his original bypass graft left in situ. After the operation at age 8, the patient did well. The patient is active, does not have any cardiopulmonary symptoms, and has normal hepatic and renal function. Hospitalizations in childhood resulted in severe post-traumatic stress disorder and anxiety (not currently on psychiatric medications), which led to absent follow-up until the current presentation. Computed tomography angiogram (CTA) revealed the anastomotic descending thoracic pseudoaneurysm, patent supra-aortic trunks with a bovine arch, occluded ascending to descending thoracic bypass graft, and patent redo bypass graft (Fig 1, *A-D*). The left subclavian artery (LSCA) originated from the aneurysmal section of the aorta, and the pseudoaneurysm was exerting mass effect with compression of the left pulmonary vasculature (Fig 2, *A-D*). Abdominal and access vessels were unremarkable (Fig 2, *A-D*). CTA of the head and neck demonstrated a patent left common carotid artery and a left vertebral artery originating from the LSCA.

Preoperative evaluation included transthoracic echocardiogram which revealed a normal left ventricular ejection fraction (55%-60%), moderate pulmonary regurgitation, and left pulmonary artery stenosis. A ventilation-perfusion scan revealed decreased perfusion to the left lower lobe of the lung. Multidisciplinary discussions among vascular surgery, cardiac surgery, and cardiology was had, including all operative options, possible complications, and backup plans. Complete resection of the aneurysm to address the pulmonary artery compression was not deemed necessary given nonsevere disease and absence of symptoms. Owing to his post-traumatic stress disorder the patient expressed a strong preference to avoid open surgery. We proceeded with a hybrid open and endovascular approach with a single stage carotid to subclavian artery bypass followed by thoracic endovascular aortic aneurysm repair (TEVAR).

Technique. The procedure was performed under general endotracheal anesthesia with electroencephalographic

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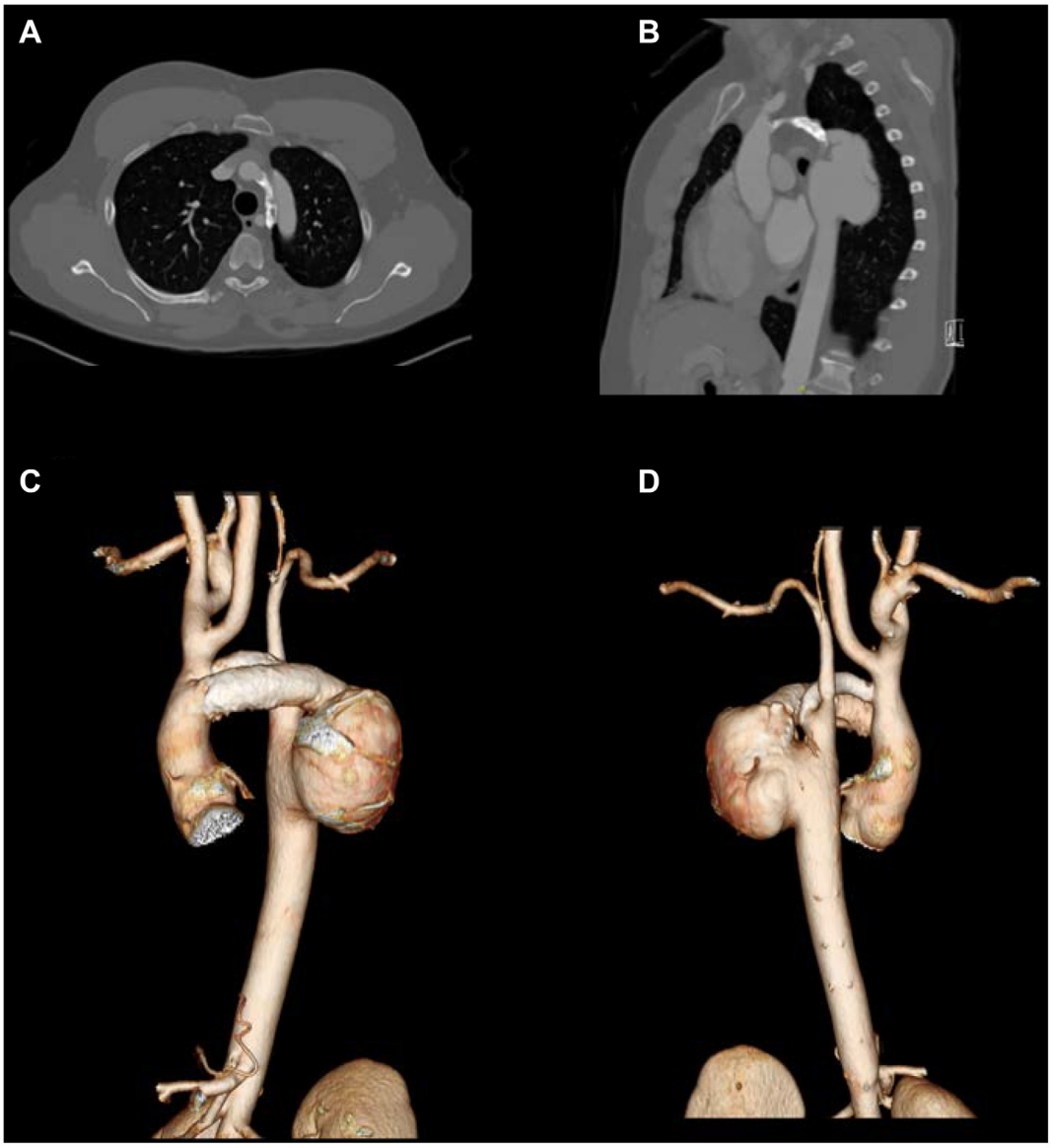

Fig 1. (**A**–**D**) Anatomy of the patient's prior ascending to descending thoracic aorta bypass grafts with pseudoaneurysm at the distal anastomosis.

neuromonitoring by the vascular surgery team. Through a supraclavicular incision, the left common carotid artery and LSCA were exposed and the patient was fully heparinized. Using 8-mm Dacron, a carotid-to-subclavian bypass was performed in the standard fashion. Unilateral percutaneous right common femoral artery access was obtained and upsized to 20F. The native LSCA was cannulated, and a 7F sheath advanced. An angiogram was obtained which identified the origin of the left vertebral artery from the LSCA and a widely patent carotid-to-subclavian bypass (Fig 3, *A*). A 12-mm Amplatzer plug (Abbott Cardiovascular, Plymouth, MN) was then advanced and deployed into the native LSCA proximal to the origin of the left vertebral artery. An angiogram confirmed successful embolization and a patent left vertebral artery (Fig 3, *B*). An aortogram for fusion onlay calibration was performed (Fig 3, *C*). Two Gore CTAG devices (W. L. Gore & Associates, Newark, DE) were sequentially advanced and deployed (26 × 10 cm followed by 28 × 15 cm), covering the native left LSCA (Fig 3, *D*). Completion cone beam CT scan and rotational digital subtraction angiogram demonstrated patent supra-aortic trunks and a main stent graft without endoleak (Fig 4, *A*, *B*). This case took a total of 3 hours and 50 minutes with 15.2 minutes of fluoroscopy time (472 mGy radiation, 245 mL contrast).

The patient was extubated in the operating room and was intact neurologically. He recovered in the intensive care unit on the night of surgery and was transferred to the vascular surgery floor on postoperative day 1. We had specialists from our psychology division meet with the patient to provide support, and he did well without any significant anxiety exacerbation. He was discharged home on postoperative day 2 with lifelong

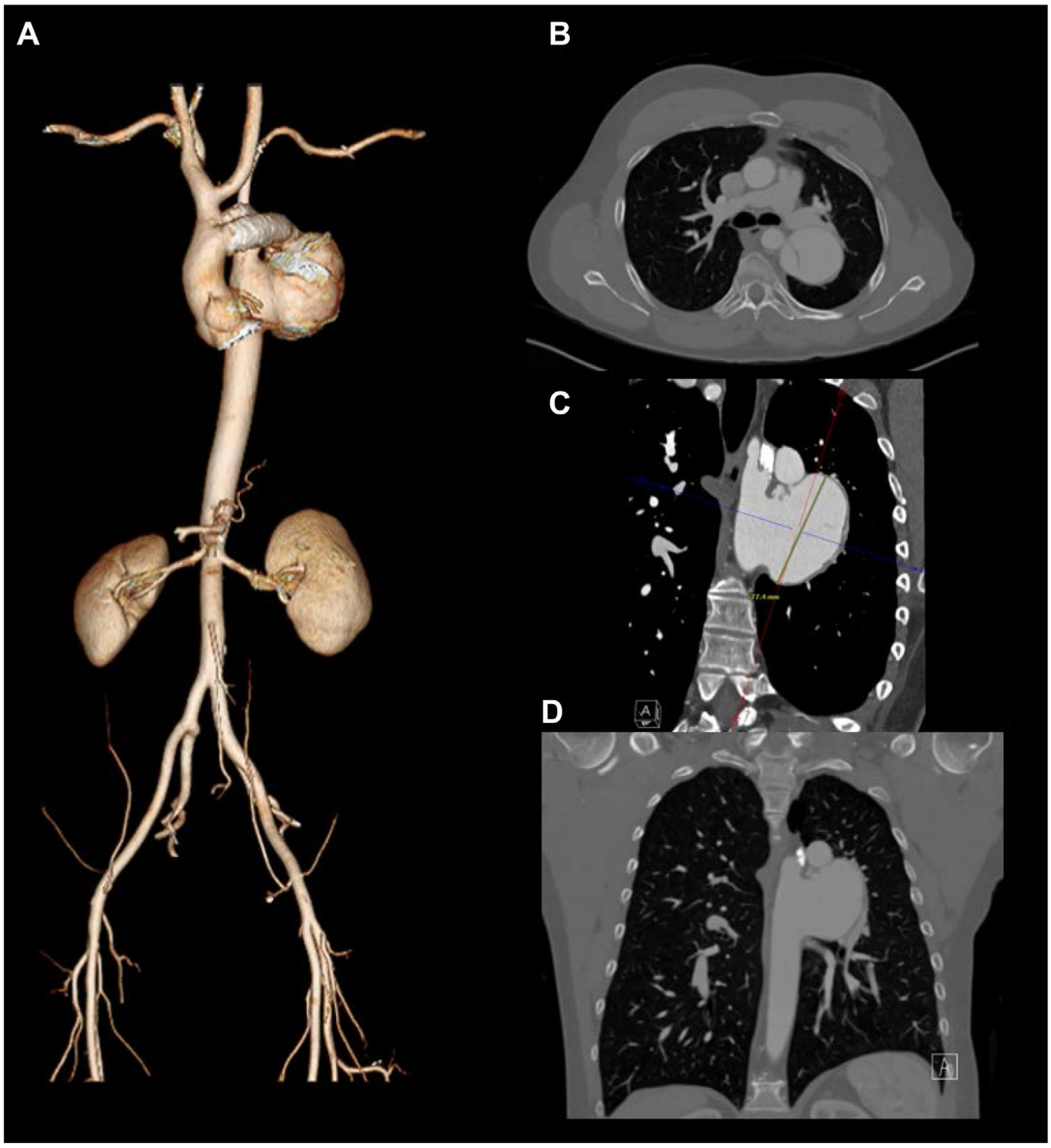

Fig 2. (**A**–**D**) Computed tomography angiogram (*CTA*) of descending thoracic pseudoaneurysm arising from the distal bypass of the patients ascending to descending thoracic aortic bypass graft.

aspirin and he quickly returned to baseline functional status. CTA at 6 months demonstrated patent stent graft and carotid subclavian bypass without evidence of endoleak and significant regression of the aneurysm (Fig 5, *A-D*).

DISCUSSION

The treatment of thoracic aortic aneurysms is performed predominantly with endovascular stent grafts. Young and active patients are considered for open repair typically; however, multiple previous operations can be a relative contraindication given the morbidity and mortality associated with redo open aortic arch surgery.[4,5] In this patient, an open operation would have been challenging in a fourth time redo field, requiring a multitime redo thoracotomy under circulatory arrest with cardiopulmonary bypass. The use of a hybrid approach facilitated endovascular repair of the thoracic aorta with LSCA preservation.

One aspect of this case that deserves mention is the possibility of a total endovascular repair, either covering the LSCA without revascularization or using a fenestrated-branched endovascular repair.[6,7] It is recommended that, in elective and select urgent cases, subclavian revascularization should be performed before coverage of the LSCA with TEVAR.[8,9] This approach has low morbidity and acceptable long-term patency rates.[10-13] To minimize the risk of Endoleak after carotid-subclavian bypass with TEVAR, the prevertebral segment of the native LSCA needs to be occluded.[7] Occlusion of the prevertebral native LSCA using an

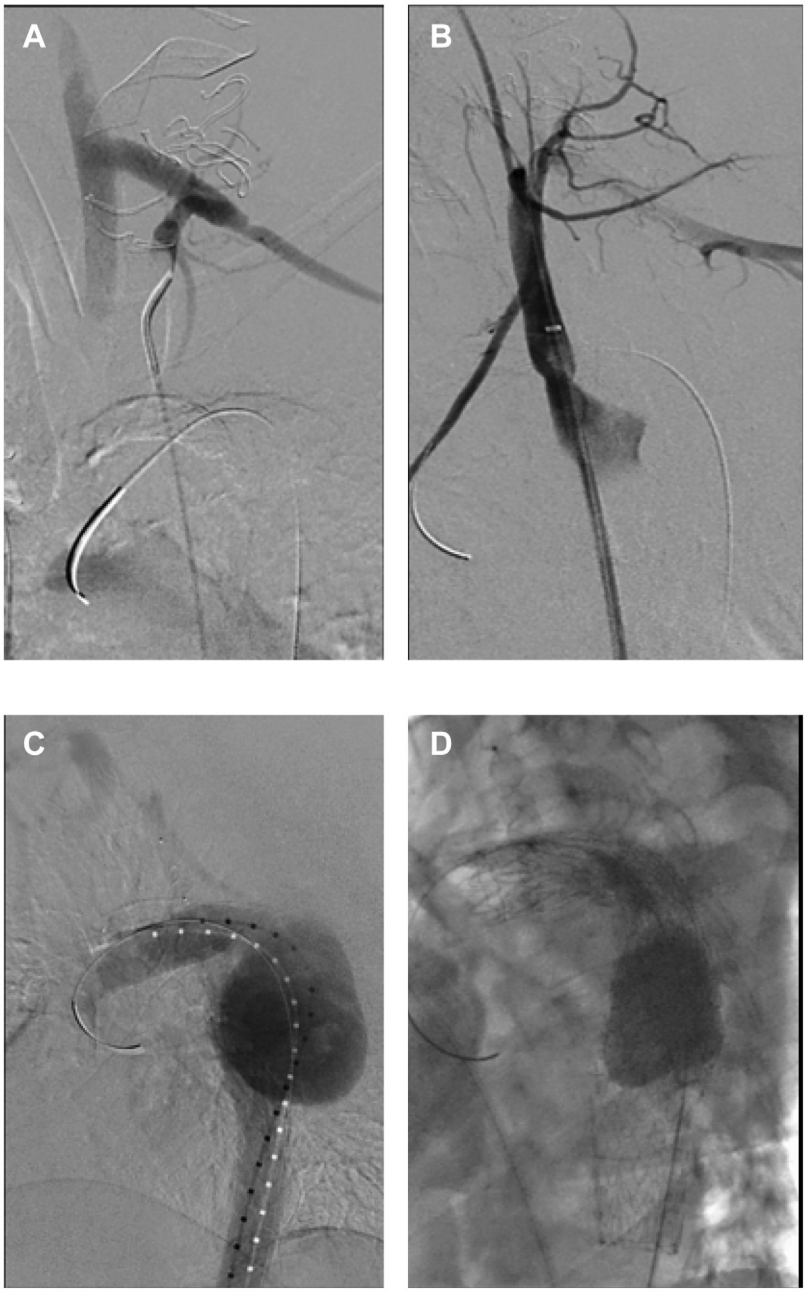

Fig 3. (**A**–**D**) Intraoperative fluoroscopy images.

Amplatzer plug after carotid-subclavian bypass, before TEVAR, has been described and has good technical success.[14] We opted for a bypass instead of a transposition owing to the very posterior location of the left subclavian. A total endovascular approach with LSCA preservation could have been performed with off-label use of the Gore thoracic branch endoprosthesis or with a physician-modified stent graft with a retrograde left subclavian branch.[6,7,15] Although these procedure would be feasible technically, a thoracic branch endoprosthesis would have been very off-label with this patient's anatomy, and the patency of a retrograde subclavian branch would have been difficult to match that of a carotid subclavian bypass, especially in such a young patient. Although a graft could have been designed with a fenestration, rather than a retrograde branch, for the LSCA, in the setting of a large aneurysm at that level, the gap between the fenestration and the origin of the LSCA would be such that the risk of type IIIc endoleak would be high.

CONCLUSIONS

In this case of a complex pseudoaneurysm of the descending thoracic aorta as a result of operations for congenital cardiac abnormalities, a hybrid approach

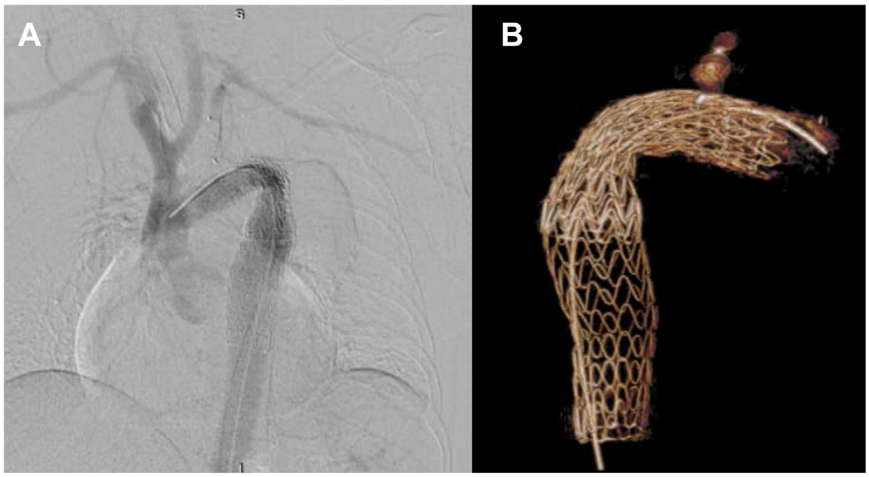

Fig 4. (**A** and **B**) Postoperative completion imaging.

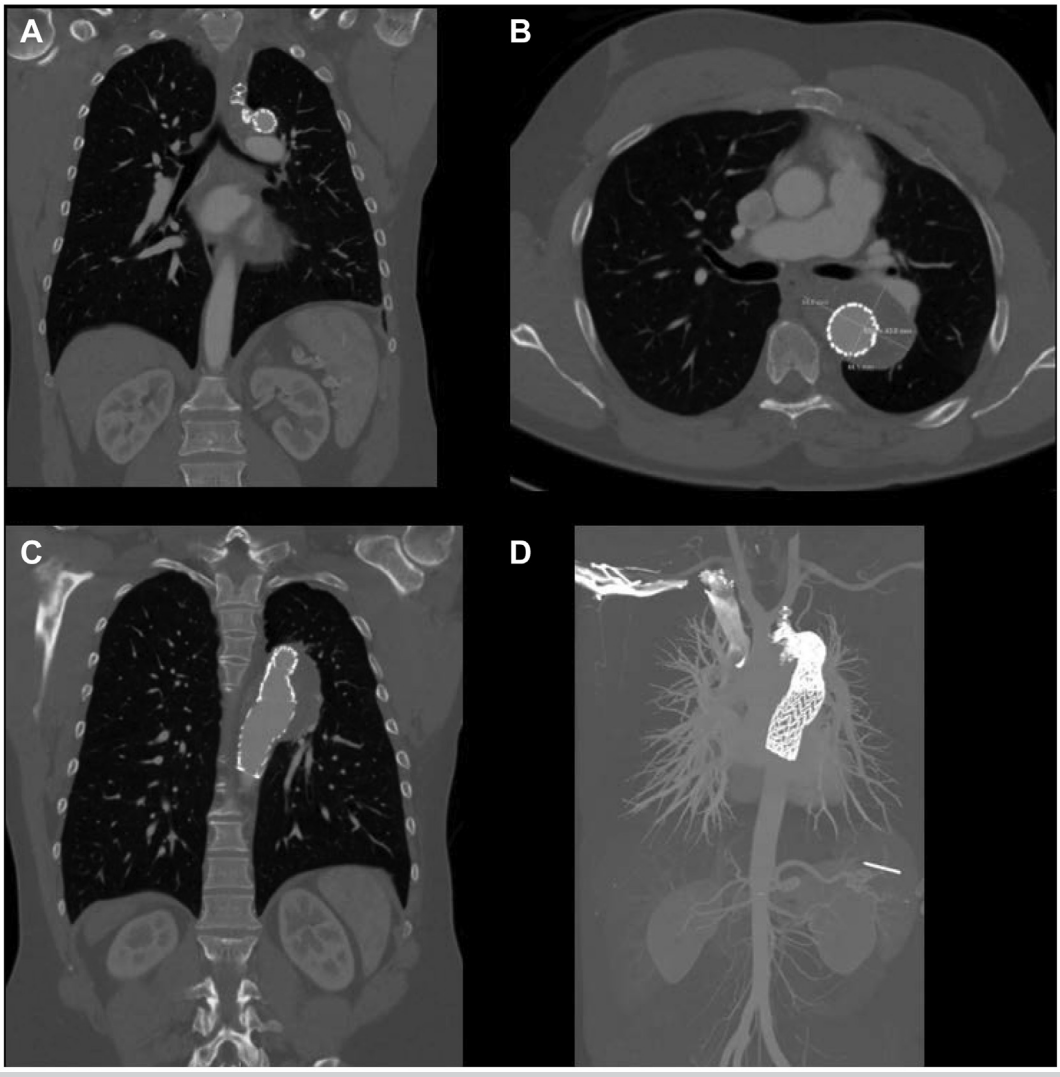

Fig 5. (**A**–**D**) Computed tomography angiogram (*CTA*) imaging performed 6 months postoperatively.

with TEVAR and left carotid-to-subclavian bypass obviated the need for open surgical intervention with a satisfactory outcome.

DISCLOSURES

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