A Giant Pseudo-Aneurysm on the Anastomosis Site for a Redo Bentall Operation due to Behçet Disease Treated by Thoracic Endovascular Aortic Aneurysm Repair with a Custom-Made Stent Graft

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ARTICLE INFO
Received February 27, 2020
Revised June 11, 2020
Accepted June 19, 2020

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†This study was presented as a poster at the 49th Annual Meeting of the Korean Society of Thoracic and Cardiovascular Surgery in Jeju, Republic of Korea, November 5–8, 2017.

Case report

Cardiovascular involvement in patients with Behçet disease is rare, although prosthetic valve failure is a frequent and serious complication. Park et al. [1] reported that they performed a Bentall procedure for aortic rupture in a patient with Behçet disease who had undergone surgery for prosthetic valve detachment 3 times previously. We hereby report the successful treatment of a patient who had an aortic pseudo-aneurysm with 2 thoracic endovascular aortic repair (TEVAR) procedures using a custom-made stent graft. Because the patient’s pseudo-aneurysm was excessively large (about 7.4 cm) and had multiple foci, surgical treatment for impending rupture of the ascending aorta through a third sternotomy was deemed too risky. Since this patient was at high risk for recurrence of pseudo-aneurysm, we planned to perform TEVAR with a custom-made stent graft.

A 34-year-old man who had undergone aortic valve replacement 8 years ago underwent an additional Bentall operation due to mechanical valve dehiscence 2 years later. Subsequently, he was diagnosed with Behçet disease and Batter syndrome. A week after being hospitalized again due to chest pain and dyspnea, a large pseudo-aneurysm was detected on computed tomography. Because of the excessively large size of the pseudo-aneurysm, surgical treatment seemed very risky. Therefore, we planned to perform thoracic endovascular aortic repair (TEVAR) and treated him successfully. However, the patient experienced recurrence of the same symptoms 4 months later, and was found to have type IV endoleak. He received a TEVAR procedure again, and it was successful.

Keywords: Pseudoaneurysm, Thoracic endovascular aortic repair, Behcet disease, Type IV endoleakage

A 34-year-old man visited the emergency room (ER) complaining of chest pain that had started a week previously. He had a medical history of Behçet disease (recurrent oral and genital ulcerations with a negative pathergy test), Batter syndrome, and gout. He had undergone an aortic valve replacement (St. Jude Medical Regent 25-mm aortic valve; St. Jude Medical Inc., Little Canada, MN, USA) due to aortic valve regurgitation 8 years ago. Due to mechanical valve dehiscence and aortic root dilatation, he underwent a Bentall operation (St. Jude mechanical 27-mm valved graft; St. Jude Medical Inc.) via redo sternotomy 6 years ago (2 years after the first operation). A chest radiograph taken at the ER showed mild mediastinal widening with no active lung lesions. Electrocardiography showed that the patient’s ventricular bigeminy and cardiac enzyme levels were normal. Based on the patient’s medical history and symptoms upon presentation, it was unlikely that the cause of chest pain was acute coronary syndrome. There-
fore, chest computed tomography (CT) was performed and the results showed multiple outpouching aneurysms along a 7.4-cm area in the ascending aorta (Fig. 1A). The origin of the aneurysms was just distal to the graft anastomosis site from the past Bentall operation. The patient was immediately hospitalized in the intensive care unit to maintain a low blood pressure (<120 mm Hg systolic blood pressure) and to ensure thorough bed rest. Because surgical treatment for impending rupture of the ascending aorta with a third sternotomy was deemed to be too risky, we planned to perform TEVAR with a custom-made stent graft (34 mm×80 mm; S&G Biotech, Yongin, Korea). The TEVAR procedure was successfully performed on the ascending aorta. Follow-up CT showed no additional extravasation of contrast medium from the ascending aorta. Three months later, the pseudoaneurysm sac was smaller than on previous scan results (Fig. 1B). Unfortunately, the patient was hospitalized again 7 months later with dyspnea and chest tightness. CT showed a 4-cm area of type IV endoleak on the ascending aorta at the previous TEVAR site (Fig. 1C). We performed a second TEVAR procedure, placing a handmade graft (32 mm×40 mm; S&G Biotech) inside the previous TEVAR graft. The procedure was successful. One year later, follow-up CT results showed a smaller pseudoaneurysm and no sign of endoleak around the previous TEVAR site (Fig. 1D). Two years after the second TEVAR procedure, he was doing well based on outpatient follow-up, without any symptoms. The patient provided written informed consent for the publication of his clinical details and images.

Discussion

Behçet disease is a well-known disease that causes non-specific arterial and venous vasculitis [2]. In cardiac Behçet disease, dilatation of the proximal aorta, interatrial septal aneurysm, mitral valve prolapse, and mitral regurgitation are common findings [3]. Surgical treatment is generally required, but Unfortunately, proximal and distal anastomotic aneurysms form quite frequently after these surgical procedures [4]. Recurrent cardiac surgical interventions increase the risk of mortality and morbidity [5]. As an alternative to open surgery, TEVAR has become popular for treating thoracic aortic pathology since 2005 [6]. Herein, we describe a rare case of cardiac Behçet disease with a pseudoaneurysm at the artificial Dacron graft anastomosis site on the ascending aorta. It was treated successfully by TEVAR.

We performed TEVAR instead of a third sternotomy for the proximal anastomotic pseudoaneurysm, but conventional TEVAR grafts were not appropriate for this patient. He had undergone a Bentall operation previously, which required us to use a stent that did not cover the coronary buttons. We requested a domestic company to create a special custom-made stent graft, which they crafted after analyzing the CT scan results of this patient. The lower part of the stent was designed to cover the area just above the aortic valve with the bare stent, and it worked safely.

Since cardiac Behçet disease is a chronic vascular disease, we had to perform routine follow-up checks, including frequent CT scans during the early stage. Later in the stable stage, the frequency of CT scans could be reduced to once or twice a year. The patient complained of dyspnea...
after 7 months had passed since the initial TEVAR procedure and we found type IV endoleak as a rare complication of TEVAR. Although the first TEVAR procedure covered the entire pseudoaneurysm, we found the leakage point at the middle of the stent graft on the angiogram and CT scan. Therefore, we performed an additional TEVAR procedure. However, this is not a standard treatment strategy, and it was performed strictly as a bailout procedure. One year after the second procedure, CT showed a significantly smaller pseudoaneurysm. The patient did not show any particularly worrying problems in later visits over the course of 2 years since the second procedure.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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References