

Case Report

Hybrid Repair for Mega-Aortic Syndrome due to Giant Cell Aortitis in a Heart Failure Patient

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The present report describes a case of mega-aortic syndrome accompanied with severe aortic regurgitation in a 75-year-old man who underwent a two-stage hybrid repair. Intraoperative pathologic findings at the first repair, consisting of Bentall operation and total arch replacement with a Lupiae graft, aided the identification of the giant cell aortitis. Despite complicating hemorrhagic stroke, steroid therapy was initiated and endovascular repair was subsequently completed. Over more than 2 years of follow-up, the patient continued steroid therapy and is doing well without any reintervention.

Keywords: giant cell aortitis, mega-aortic syndrome, hybrid repair

Introduction

Giant cell aortitis (GCA) is an uncommon cause of thoracic aortic aneurysms, which often involve the aorta from the aortic root through the arch, extending to the thoracoabdominal aorta, the so-called mega-aortic syndrome (MAS).^{1–3} We performed a two-stage hybrid repair, combining open and endovascular techniques, to treat MAS due to GCA in a heart failure patient with severe aortic

regurgitation.

Case Report

A 75-year-old man was admitted with a 2-month history of chronic heart failure due to severe aortic regurgitation. Color Doppler echocardiography demonstrated a large aortic root aneurysm with severe aortic regurgitation. Aortic regurgitation was presumably caused by central insufficiency secondary to annular dilatation. The left ventricular ejection fraction was 50%, the left ventricular diastolic dimension was 60 mm, and the left ventricular systolic dimension was 46 mm. Pulmonary hypertension was also identified, and estimated systolic pulmonary artery pressure was 45 mmHg.

Chest computed tomography (CT) revealed the development of MAS, with diameters of 60 mm in the ascending aorta, 58 mm in the aortic arch, and 62 mm in the descending thoracic aorta (Fig. 1). He did not present the typical symptoms of temporal arteritis and polymyalgia rheumatica. The laboratory examinations showed a white blood cell count of 5400/mm³, hemoglobin concentration of 9.7 g/dl, hematocrit of 28.9%, and C-reactive protein (CRP) of 0.58 mg/dl. The erythrocyte sedimentation rate level was not determined. Although he was given optimal pharmacotherapy, his heart failure did not improve with 854 pg/ml of brain natriuretic peptide before the operation. To aid the management of the heart failure and mega aorta, a two-stage hybrid repair, combining first-stage open aortic replacement and second-stage endovascular stent grafting, was planned.

At the first stage, the patient underwent Bentall operation and total arch replacement. Intraoperatively, the ascending aorta presented a glittering cobblestone appearance compatible with aortitis (Fig. 2A). Under cardiopulmonary bypass, the aneurysmal root wall with the intimal wrinkling (Fig. 2B) was removed, and histological evaluation was performed, which confirmed GCA (Fig. 2C). The inflammatory infiltrate of the mononuclear cells within the media of the aortic wall showed multinucleated giant cells (Fig. 2D). However, the aortic leaflets failed to reveal any evidence of vasculitis. The aortic leaflets showed dystro-

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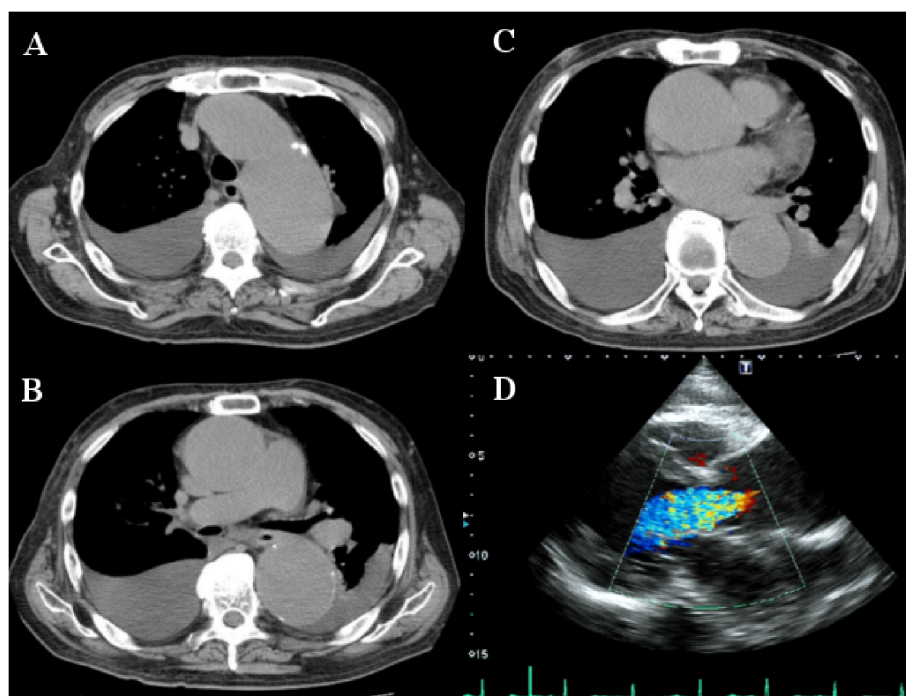


Fig. 1 Preoperative computed tomography showing a mega thoracic aorta with bilateral pleural effusion. (A) 58mm aortic arch aneurysm. (B) 60mm ascending and 62mm descending aortic aneurysm. (C) 40mm descending thoracic aorta just above the diaphragm. (D) Preoperative echocardiography showing severe aortic regurgitation with a dilated ascending aorta.

phic changes with a mild degenerative process. Given the patient's advanced age and dystrophic changes in the aortic valve, we decided to perform aortic root replacement. Bentall procedure was performed using a 25-mm Crown Phospholipid Reduction Treatment aortic valve prosthesis and a 30mm Mitroflow Valsalva Conduit (LivaNova, London, UK). The autologous pericardial reinforcement of the proximal anastomosis was performed as previously described by us.⁴⁾ The coronary buttons were reimplemented as a Carrel patch on the skirted section of the composite Valsalva graft. Thereafter, graft-to-graft anastomosis was performed between the 30mm Lupiae (Vascutek Termo Inc., Scotland, UK) graft and the Valsalva composite valve graft. The Lupiae graft has a unique branch, the so-called "bovine trunk," which further divides into three branches (first, second, and third branches).²⁾ After the establishment of a moderate hypothermic circulatory arrest (bladder temperature: 24°C), a bilateral selective antegrade cerebral perfusion was commenced. The aortic arch was transected between the left carotid and left subclavian arteries. The left subclavian artery was detached from the aortic arch, and a Dacron tube graft was sutured end to end to the left subclavian artery in advance. After the reinforcement of the distal anastomotic site, open distal anastomosis was performed. The diameter at the distal

anastomotic site of the aorta was 47mm. Because we had to anastomose the graft with the aneurysmal distal aorta, the outer side of the aorta was reinforced with a Teflon felt strip, and the inner side was also reinforced with the short segment of the prosthetic graft.

Systemic reperfusion was resumed via the rim of the Lupiae graft. The left carotid artery, brachiocephalic trunk, and left subclavian artery were then reconstructed in turn. The cardiopulmonary bypass time was 353 min, the aortic clamp time was 235 min, and the circulatory arrest time with selective antegrade cerebral perfusion was 65 min. The patient received 12 units of packed red blood cells, 20 units of fresh frozen plasma, and 15 units of platelets.

The patient tolerated the procedure and then was extubated on postoperative day 2. He received aspirin and warfarin with an international normalized ratio of 1.5 to 2.0. He was neurologically intact 3 weeks after the surgery. On postoperative day 22, the patient suffered right hemiparesis with mild aphasia. CT revealed a hemorrhage stroke in the left frontal lobe. In addition, given the intraoperative histologic diagnosis of GCA, steroid therapy was initiated. After the initiation of prednisolone at a dose of 20 mg/day, the CRP level gradually decreased from 9.7 mg/dl to 0.65 mg/dl within two weeks. Thereafter, the prednisolone dose was tapered in accordance with the

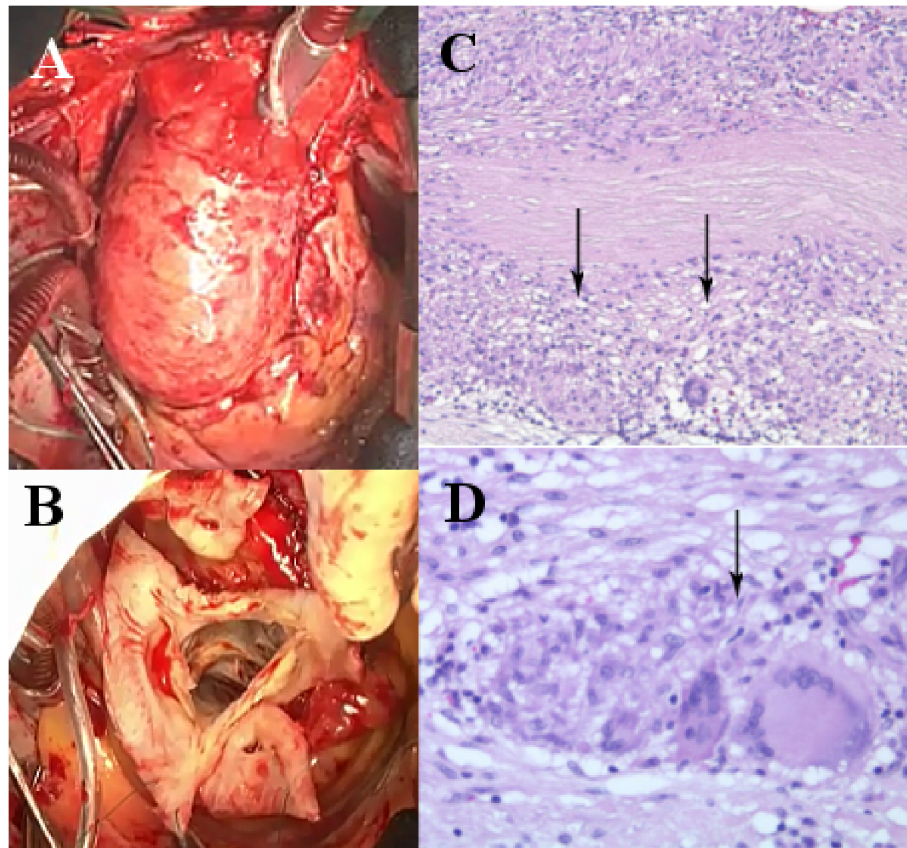


Fig. 2 (A) Intraoperative view of the ascending aortic aneurysm with a glittering cobblestone appearance. (B) Interior view of the coronary button and residual part of the involved aortic root with intimal wrinkling. (C) Histologic examination of the aortic wall showing an inflammatory infiltrate of mononuclear cells within the media of the aortic wall (arrow). (D) Multinucleated giant cells (arrow).

CRP level, with the final dosage at 8 mg/day. Additionally, the patient's condition remained clinically stable.

After a 4-month recovery period with the stabilization of the cerebral damage, the patient was brought to the operating room for the second-stage surgery. CT before the stent grafting demonstrated the slightly expansion of diameter of the prosthetic arch graft to 32 mm. The endovascular repair of the residual descending thoracic aortic aneurysm consisting of two stent grafts (Relay, 38 mm×38 mm×25 cm proximal device and 44 mm×44 mm×25 cm distal device; Bolton Medical, Barcelona, Spain) was performed. The Lupiae graft just distal to the origin of the bovine trunk created an optimal straight Dacron landing zone on the ascending aorta.

The distal landing zone was the descending aorta just above the celiac trunk. Postoperative three-dimensional CT demonstrated an intact repair with aneurysm exclusion and without endoleaks (Figs. 3A and 3B). Over more than 2 years of follow-up, the patient continued steroid therapy with a maintenance dose of 8 mg/day of prednisolone and is doing well without any reintervention.

Discussion

Extensive aortic disease in GCA is relatively rare in Japan as compared with the United States and European countries. From previous epidemiological studies, the prevalence of giant cell arteritis in Japan in 1997 (revealed to be 1.47 per 100,000 population older than 50 years of age, according to the 1997 Japan census data) is extremely low compared with that in the United States (reported as approximately 200 per 100,000 population older than 50 years of age).⁵⁾ Kobayashi et al. have also reported that the average age at onset was 71.5 years old, and the male:female ratio was 1:1.7.⁵⁾ Thoracic aortic aneurysm due to GCA, in which the ascending aorta and aortic arch are the most commonly affected, is usually a late complication of the disease. Evans et al. reported that 11.5% of patients had developed a thoracic aortic aneurysm during a median time of 5.8 years after the diagnosis of giant cell arteritis.⁶⁾ Moreover, patients with giant cell arteritis are 17.3 times more likely to develop a thoracic aortic aneurysm compared with the general population.⁶⁾ Oc-

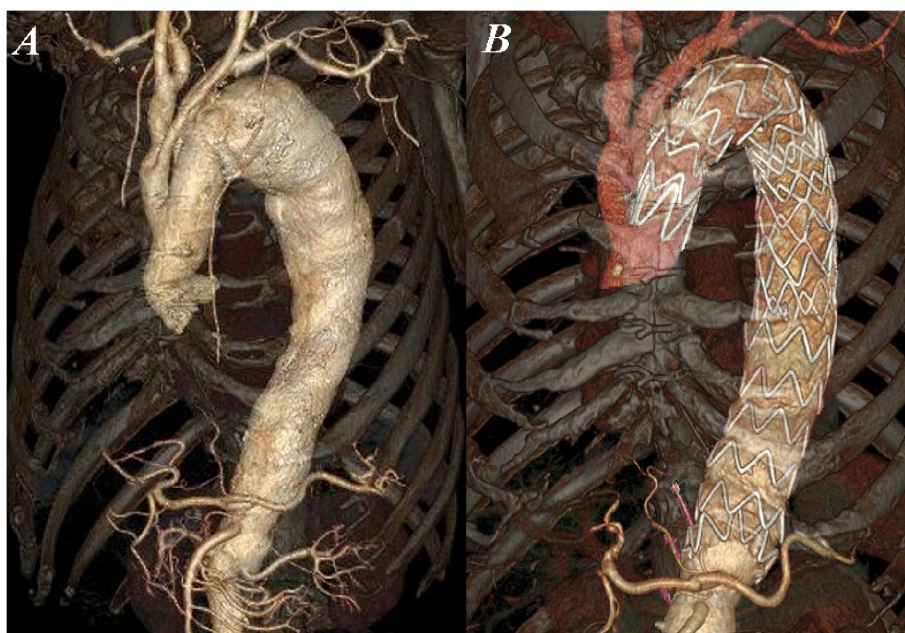


Fig. 3 (A) Preoperative three-dimensional computed tomography (CT) demonstrating an intact first-stage repair and the mega aorta from the distal arch to the descending aorta. (B) Three-dimensional CT after the second endovascular repair demonstrating an intact repair with aneurysm exclusion.

asionally, the development of MAS with varying degrees of the descending aorta with GCA is observed, potentially leading to catastrophic events, such as aneurysmal rupture and aortic dissection.^{2,7)}

Aortic regurgitation with GCA is often central insufficiency due to the dilatation of the sinotubular junction. With the normal aortic valve cusps viewed macroscopically, valve-sparing root replacement could be indicated.^{1,7,8)} Gagné-Loranger et al. reported that valve sparing was performed in 16 patients (50% of aortic valve procedure). One patient required an aortic valve replacement redo owing to a recurrent aortic regurgitation 4 years after the valve-sparing procedure.¹⁾ However, in the presence of native aortic valve disease, the aortic valve procedure should be tailored in accordance with the patient's advanced age and comorbidities, resulting in low morbidity and mortality.

Our patient developed hemorrhagic stroke as a complication. The underlying pathologic process in GCA may be an autoimmune reaction involving arterial elastic tissue. However, the intracranial arteries have much less elastic fibers in the media.⁹⁾ To date, reports documenting intracranial vasculitis as a direct cause of stroke in patients with GCA are scarce. Giant cell arteritis mainly affect medium and large extracranial arteries. Embolization from thrombosed vessels damaged due to arteritis into the cerebral supply artery may be causes of cerebrovascular accidents.⁹⁾ Similarly, in our patient, intracranial magnetic resonance angiography after the hemorrhagic stroke

demonstrated no significant findings, such as stenosis and occlusion of the intracranial vasculature, which is compatible with a diagnosis of vasculitis. In the present case, the postoperative antiplatelet and anticoagulation therapy had been initiated. However, given these considerations, the possibility remains that the anastomotic site of the branch of the prosthetic graft or the prosthetic valve might be related to stroke.

Hybrid aortic repair using an endovascular technique has emerged as an alternative to the conventional two-stage elephant trunk technique.^{3,10)} Recently, Roselli et al. reported that the high rate of endoleaks for the endovascular completion in patients who have already undergone conventional elephant trunk technique.¹⁰⁾ This reason is possibly due to the free-floating design of the original version of the elephant trunk. Esposito et al. developed a novel hybrid treatment for MAS called the "Lupiae technique."³⁾ They advocated that Lupiae technique offers a fixed elephant trunk that seems to outperform the free-floating one. In the present case, the first repair using the Lupiae graft ensured a more reliable proximal landing zone on the ascending aorta. However, the adoption of endovascular repair in GCA remains questionable. Gagné-Loranger et al. cautioned that patients with GCA may be at increased risk for endoleaks, especially when the landing zone is diseased.¹⁾ Reports of the anastomotic pseudoaneurysm in patients with GCA were scarce compared with Takayasu arteritis. However, patients with aneurysms by GCA carry an incidence of aortic dissection. In

addition, several reports on later aneurysmal involvement of the descending or thoracoabdominal aorta have been made.^{2,7)} Therefore, patients with MAS due to GCA are at risk of late vascular reintervention. Owing to the modest abdominal aortic dilatation involving the celiac artery in the present case, a routine follow-up consisting of CT scan and inflammatory marker surveillance is of paramount importance.

Conclusion

In conclusion, a two-stage hybrid repair, combining open and endovascular techniques, is effective in the treatment of MAS due to GCA. Given the risk of late reintervention, routine imaging surveillance is critical to assess further aortic development with GCA.

Disclosure Statement

All authors have no conflict of interest.

Author Contributions

Study conception: TI, KF

Data collection: TI, KF

Analysis and Interpretation: TI, SK

Writing: TI

Critical review and revision: all authors

Final approval of the article: all authors

Accountability for all aspects of the work: all authors

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