Successd Hybrid Treatment for a Ruptured Thoracoabdominal Aortic Aneurysm in a Patient with Systemic Lupus Erythematosus

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A 49-year-old woman with a 27-year history of systemic lupus erythematosus (SLE) was admitted to our hospital with sudden-onset severe back pain. An emergency multidetector computed tomography (MDCT) revealed a ruptured thoracoabdominal aortic aneurysm (TAAA) 80 mm in diameter. Considering her condition and comorbidities, we performed an emergency hybrid treatment: visceral reconstruction followed by endoluminal aneurysm exclusion. She recovered uneventfully, except for the need for temporary hemodialysis. TAAA complicated with SLE is extremely rare. To our knowledge, this is the first successful report in the English literature of a ruptured TAAA in a patient with SLE who underwent hybrid treatment.

Keywords: ruptured TAAA, hybrid treatment, SLE

INTRODUCTION

Systemic lupus erythematosus (SLE) is frequently associated with cardiovascular manifestations, but it is rarely complicated with aortic disease.1 In this paper, we report an extremely rare case of ruptured thoracoabdominal aortic aneurysm (TAAA) in a patient with SLE, who was successfully treated with a hybrid procedure: combination of endovascular exclusion and visceral revascularization.

CASE REPORT

A 49-year-old woman was brought to our emergency department by ambulance for sudden onset, severe back pain. She was diagnosed with SLE at the age of 21 years, based on established criteria. Thereafter, she was given varying doses of prednisolone with or without azathioprine. For the past 10 years, she was free of SLE symptoms with prednisolone of 10 mg/day. She was also diagnosed with hypertension and hyperlipidemia. She developed chronic renal failure (serum creatinine concentration ranged from 1.72 to 2.37 mg/dL) as a result of lupus nephritis secondary to SLE. She had no history of smoking.

On arrival, she appeared pale with a cold sweat. Physiologic examination revealed a height of 136 cm, body weight of 31 kg, body temperature of 35.9°C, heart rate of 120 beats/min and blood pressure of 78/50 mmHg. Abdominal examination revealed a pulsatile epigastric mass with abdominal guarding tenderness. Laboratory investigations showed a leukocyte count of 15300/mm3, hemoglobin of 8.2 g/dL, hematocrit of 26.6%, platelet count of 9.9 × 104/mm3, blood urea nitrogen of 41.0 mg/dL, and serum creatinine of 2.16 mg/dL. An emergency multidetector computed tomography (MDCT) scan demonstrated a ruptured type III TAAA 80 mm in diameter and retroperitoneal hematoma around it. It also showed severe calcification of the entire aorta (Fig. 1). The celiac artery (CA) was occluded at its origin and was perfused...
with collaterals from the superior mesenteric artery (SMA) (Fig. 2).

For this patient, we decided to perform an emergency hybrid procedure with a combined open and endovascular procedure as a less invasive treatment for the following reasons: 1) vital instability intolerable for thoracotomy and single-sided ventilation, 2) existence of severe calcification of the entire aorta, making it difficult to apply cross clamping as well as graft anastomosis.

**Surgical Management**

Under general anesthesia, a midline celiotomy was made. Cerebrospinal fluid drainage was not used. A large aortic aneurysm with a hematoma was observed in the retroperitoneal space just below the diaphragm. The SMA, bilateral renal arteries (RAs), and bilateral common iliac arteries (CIAs) were dissected out in preparation for the bypass and insertion of the stent graft. The left CIA had severe calcification on palpation and was judged unfit for graft anastomosis. Because the patient’s body size was very small (a body surface area of 1.09 m²), RAs were also small, 3.5 mm in diameter. Therefore, we decided to employ saphenous veins as bypass grafts instead of the commonly used synthetic grafts of 6 to 10 mm in size. Then, great saphenous vein grafts (SVGs) were harvested from her bilateral lower extremities simultaneously. After systemic heparinization, one SVG bypass was placed from the right CIA end-to-side to the left RA. Then, the other SVG was placed from the side of the first SVG end-to-side to the SMA, just inferior to the pancreas. Although we tried to bypass the right RA, it was strongly compressed with the aortic aneurysm, which did not have adequate length and quality for grafting. At the same time, her vital signs progressively deteriorated into shock due to continuous bleeding from the retroperitoneal space; thus we decided not to bypass the right RA and ligated it at its origin to be sacrificed. The CA was not bypassed because its origin was occluded and had an acceptable visceral collateral pathway from the SMA. The origins of the host vessel were ligated to prevent type II endoleaks.

After visceral and renal bypass, a guide wire was advanced into the thoracic aorta via the left CIA under fluoroscopic guidance. A 24F introducer sheath was then inserted over a super stiff 0.035-inch Lunderquist wire (Cook Medical Inc., Bloomington, InUSA), followed by Gore® RTAG (W.L. Gore & Associates, Flagstaff, Arizona, USA) stent-graft insertion. After the position had been confirmed angiographically, the stent graft was deployed in the standard fashion. Three TAG graft components were used, each one overlapping the other by 3 cm. The first component (28 mm × 15 cm) was deployed at
5 cm above the terminal aorta. The second component (31 mm × 15 cm) was deployed proximal to and overlapping the first component. The third component (34 mm × 20 cm) was overlaid proximal to the second component, up to the mid portion of the descending aorta. The devices were then additionally expanded with a specially designed GORE®Tri-Lobe Balloon Catheter (W.L. Gore & Associates, USA) that allows the flow to continue during inflation.

The completion angiogram showed no endoleak, and there was good perfusion of the left RA and SMA via the right CIA bypass graft. Adequate opacification of the CA was observed by the collateral flow from the SMA.

**Postoperative Course**

The patient was admitted to the intensive care unit for 6 days. No neurological complication was observed. The postoperative course was delayed by renal failure, which was caused by preoperative lupus nephritis and intraoperative right kidney sacrifice. Serum creatinine concentration and blood urea nitrogen increased up to 3.78 mg/dl and 146.5 mg/dL respectively. She developed oliguria and required hemodialysis for 25 days. After discontinuation of hemodialysis, her serum creatinine level varied around 2.5 mg/dl. MDCT after 1 month showed good positioning of stent-grafts, no sign of endoleak and good visualization of the visceral bypass graft. The CA was opacified via collaterals from the SMA (Fig. 3). She is now in good condition, 2 months after the operation.

**Discussion**

SLE is a chronic autoimmune disease characterized by widespread inflammation of blood vessels and connective tissues, resulting in a broad range of clinical manifestations. The cardiovascular system is often damaged, mainly as panniculitis involving the pericardium, myocardium, endocardium and coronary arteries, but the occurrence of an aortic aneurysm is extremely rare. Among aortic aneurysms, TAAA has been reported in only few cases, and a ruptured one has not yet been reported.

However, in association with prolonged survival in SLE patients caused by the improved control of the disease with corticosteroid treatment, antibiotics, and renal replacement therapy, aortic manifestations have become more apparent and a significant clinical problem. In 2002, Kunihira, et al. reviewed worldwide reports of aortic aneurysms associated with SLE and found 22 cases of non-dissected aneurysms. Common features in such patients were young age, female dominance, presence of systemic disease and longstanding corticosteroid therapy. The present case was of a 49-year-old female having comorbidities of hypertension, hyperlipidemia and chronic renal failure, and a 20-year steroid user; thus presenting similar features.

The etiology of aortic aneurysm formation in patients with SLE is reported to be affected by steroid therapy. Prolonged corticosteroid administration plays a major role in accelerating atherosclerosis, which can result in aortic aneurysmal enlargement, possibly together with primary aortic wall involvement and/or vasculitic damage, particularly severe medial destruction. In the present case, severe aortic atherosclerosis and development of TAAA seemed to be closely related to 20 years of steroid therapy, as well as pathophysiology of SLE.

Open surgical repair of non-ruptured TAAAs is still challenging. Repair of such complex aneurysms requires extensive exposure, cardiopulmonary bypass, and supraceliac

![Fig. 3](A) front view, (B) lateral view White arrow: graft to the left RA, Black arrow: graft to the SMA Postoperative three-dimensional CT angiography scan shows good positioning of the stent-grafts, no sign of endoleak and well visualization of visceral bypass graft. There observed adequate opacification of the celiac artery by the collateral flow from the SMA. RA: renal artery; SMA: superior mesenteric artery; CT: computed tomography
Hybrid Treatment of Ruptured TAAA with SLE

In conclusion, we have described an extremely rare case of ruptured TAAA in a patient with SLE. We performed a hybrid treatment: visceral reconstruction followed by endoluminal aneurysm exclusion for this patient successfully. To our knowledge, this is the first successful report of a ruptured TAA associated with SLE treated with a hybrid treatment in English literature.

**Disclosure Statement**

The authors have no conflict of interest to declare.

**References**