A 68 year old man was admitted to our hospital with dysphagia and back pain. Contrasted computed tomography showed “Shaggy aorta” forming a saccular descending aortic aneurysm with edematous esophagus. Low density area in the intramuscular layer of the esophagus suggested the possibility of connection between the esophagus and the aneurysm. The patient underwent endovascular treatment of the aneurysm. The postoperative course was uneventful, and the patient was discharged from the hospital with improvement in his symptoms. Although there are reports suggesting endovascular treatment as a contraindication for shaggy aorta due to risk of embolization, it may be considered as an option for a patient who is in need of surgical treatment.

Keywords: shaggy aorta, stent graft, blue-toe syndrome, statin

Introduction

Shaggy aorta syndrome is a rare disorder causing peripheral, renal, and visceral ischemia, caused by multiple cholesterol emboli from the aorta. The natural history of the process involves recurrent showers of emboli with progressive ischemic damage to the tissues involved. In such a case, prosthetic replacement of the diseased aorta provides effective treatment against recurrent embolization. Shaggy aorta may also complicate aneurysm that needs treatment. Manipulation of the aorta aggravates embolization and some reports suggest contraindication for surgery in patients with shaggy aorta. Here, we report a successful endovascular treatment of enlarging descending aortic aneurysm presenting dysphagia in a patient with shaggy aorta.

Case Report

A 68 year old man was referred to our hospital with dysphagia and back pain. He had past medical history of chronic kidney disease, diabetes mellitus, hypertension, and hypothyroidism. He also underwent surgical resection of right common iliac arterial aneurysm four years previously. Gastrointestinal endoscopy performed in annual medical checkup one year prior to admission did not show any sign of mass formation in the esophagus.

Computed tomography on admission, showed descending aortic aneurysm, which did not exist in the computed tomography performed 4 years previously. The esophagus was swollen, but there was no mass formation. Although there was no sign of extravasation, the esophagus revealed low density area in the intramuscular layer which suggested the possibility of connection between the esophagus and the aneurysm (Fig. 1B and 1C). The aorta also showed severe arteriosclerosis and friable mural thrombus presenting “shaggy aorta” (Fig. 2A–2C). Laboratory data showed chronic renal deficiency with creatinine of 3.25 g/dl. However, there was no sign of other systemic
Stentgraft Treatment in Shaggy Aorta

Because of the risk of aneurismal rupture, gastrointestinal endoscopy was not performed. Barium radiology was pursued for examination of the esophagus which showed compression of the esophagus from the aneurysm. However, the intraluminal channel of the esophagus was preserved (Fig. 1A).

Preoperative work up with myocardial scintigram showed redistribution in the posterior lateral wall of the left ventricle suggesting ischemic heart disease. Although there was a risk of embolization by catheter manipulation, endovascular treatment was scheduled due to risk of coronary complication and excessive surgical stress under prolonged anticoagulation environment in conventional surgical treatment. Medication with statin was introduced before surgery expecting plaque stabilization, and administration of atrial natriuretic peptide and hydration was started one day before surgery for prevention of contrast-induced nephropathy.3–7)

For minimal manipulation of the aorta, treatment with a single stent graft was considered. The treatment length was 134.5 mm. The diameter of the proximal neck just above the ulcer like protrusion was 28 to 31 mm. The aortic curvature above celiac artery was chosen for the distal landing zone, which was 38 mm in length, and 30 to 31 mm in diameter. (Fig. 2B and 2C) The aortic diameter at the celiac artery was enlarged to 32mm. Deployment of TAG thoracic endoprosthesis: TGT 3420 (W.L. Gores & Associates, Newark, Delaware, USA) from the ulcer like protrusion to aortic curvature was considered for the treatment. With a possibility of additional treatment with a larger stent graft, 24Fr introducer catheter (W. L. Gores & Associates, Newark, Delaware, USA) was chosen.

The patient was placed in a right hemilateral position. Under general anesthesia, femoral artery was exposed on both sides. Lunderquist guide wire (Cook Medical, Bloomington, Indiana, USA) was carefully inserted up to distal aortic arch through right femoral artery. Pigtail catheter (Cook Medical, Bloomington, Indiana, USA) was inserted from left femoral artery up to the distal aortic arch for angiogram. 24Fr sheath catheter was administered through the right femoral artery with care, taking 5 minutes to reach the celiac artery. TAG thoracic endoprosthesis: TGT 3420 was inserted through the 24Fr sheath catheter gently, taking 10 minutes to reach the deployment position. With the stent graft deployment, blood was flushed out from the introducer sheath to prevent peripheral embolization. Angiogram after deployment showed excision of the aneurysm without an endoleak. Preservation of visceral arteries was also confirmed, and a 24Fr sheath was carefully taken out.

The patient was extubated in the operation room. There was no sign of systemic embolization including cerebral infarction and blue toe syndrome. Laboratory data also showed no elevation in lactate, creatinine kinase, lactate dehydrogenase after the operation.

Post-operative computed tomography, performed 10
days after the operation, showed total excision of the saccular aneurysm and improvement in the esophagus swelling (Fig. 3A and 3B). Dysphagia and back pain has diminished, and the patient was discharged from the hospital without any major complication.

**Discussion**

Shaggy aorta causes disseminated cholesterol microembolization resulting in ischemic damage to the organs involved. Invasive arteriography and anticoagulant therapy are known to be the predisposing and aggravating factors. Friable mural thrombus is easily released into the blood stream by manipulation of the aorta. Furthermore, anticoagulant therapy interferes with the formation of fibrin layer over an eroded or ulcerated atherosclerotic plaque thus inhibiting cementing of cholesterol and atheromatous debris by fibrin.

Hayashida et al. reported a case in which anticoagulant therapy and intra-aortic catheterization exacerbated the patient’s renal function and triggered another massive microembolization of the visceral arteries resulting in a fatal outcome. For those with visceral involvement, extra-anatomical bypass with ligation of the distal external iliac arteries appears to be associated with lower morbidity and mortality, as well as prevention of recurrent visceral embolization. However, contraindication to surgical treatment could the only way to prevent this complication.

On the other hand, Sanada et al. reported successfully treated case of thoracic aortic aneurysm with shaggy aorta by using homemade intra-aortic filter device to prevent embolization during stent-graft repair.

Although there is a risk of shower emboli, treatment should be considered for those with recurrent emboli or aneurysm formation. Conventional surgical treatment is also an aggravating factor for embolization due to manipulation of the aorta and prolonged anticoagulant environment during cardiopulmonary bypass. Although manipulation of the aortic lumen with catheter may be devastating, lesser surgical stress including operation time and shorter anticoagulated environment is a beneficial factor for endovascular treatment.

Expecting atherosclerotic plaque regression and fibrous cap formation, statin therapy was introduced before surgery. Takarada et al. reported that inhibition of matrix metalloproteinases induced by statin therapy resulted in accumulation of collagen and several extracellular matrix components resulting in thickening of the fibrous cap. Furthermore, reduction of LDL cholesterol itself increases fibrous-cap thickness due to reduction of smooth cell muscle apoptosis which is promoted by oxidized LDL. Although there are no reports connecting statin therapy with shaggy aorta, there is a growing evidence showing atherosclerotic plaque regression by statin therapy.

The patient was positioned in a hemilateral position to shift the angulation of the visceral arteries from gravity vector, thus expecting lesser risk for embolization. The catheter procedure was performed with care, and a 24Fr
sheath catheter was introduced, proximal to celiac artery, to minimize direct contact of the device with the atherosclerotic plaque of the shaggy aorta. At last, the device was introduced into the aorta gently before deployment.

**Conclusion**

We experienced a successful endovascular treatment of descending aortic aneurysm with shaggy aorta. Although catheter manipulation is reported to be a contraindication for shaggy aorta, it could be considered as an option for patients who need surgical treatment.

**Disclosure Statement**

I acknowledge that 1) we have no financial or other interest in the manufacture or distribution of the device and that 2) we do not have a financial interest in the manufacturer of the device, or receive financial incentives from the manufacturer.

**References**