Thoracic Endovascular Aortic Repair in A Situs Inversus Totalis Patient with DeBakey Type 3 Dissection

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Abstract
Situs inversus totalis is a rare autosomal recessive congenital anomaly. There are very few reports in the published literature of abdominal aortic aneurysm in patient with situs inversus totalis, all of whom underwent open aneurysm repair. DeBakey type III acute aortic dissection is life threatening condition with organ malperfusion. A 74-year-old male, referred our department with back pain and dyspnea. Contrast-enhanced computed tomography (CT) suggested DeBakey type III aortic dissection and situs inversus totalis. Thoracic endovascular aneurysm repair (TEVAR) were successfully performed. Postoperative control CT showed successful treatment of the DeBakey type III dissection with TEVAR in a situs inversus totalis patient. His postoperative course was uneventful. With situs inversus totalis, which is the first case in the literature that DeBakey type 3 dissection is treated with TEVAR. TEVAR is applicable in Situs inversus totalis patients.

INTRODUCTION
The normal arrangement of internal organs is known as situs solitus while situs inversus is generally the mirror image of situs solitus. Situs inversus is a congenital defect defined by a mirror image anatomic variation of the thoracic and abdominal organs [1]. This is not a disease but this condition is infrequently seen. Most cases with situs solitus are associated with other cardiac or noncardiac malformations. However, patients with situs inversus totalis uncommonly have other associated malformations [2-4] and, therefore, it is not unusual for them to reach older ages in which degenerative aortic stenosis is common.

Inversion of cardiovascular structures is an added procedural difficulty for endovascular replacement treatment. DeBakey type III dissection is still a potentially life threatening condition. Standard surgery is associated with high mortality and morbidity rates. Endovascular stent graft repair of aortic disease is a treatment that offers an alternative to conventional surgery. Transluminal endovascular grafting for the treatment of DeBakey type III dissection has attracted considerable attention because it is less invasive than conventional operative procedures.

CASE
A 74-year-old male who was referred to our department because of progressive back pain and dyspnea on effort, on physical examination blood pressure was 170/100 mmHg, heart rate was 108 / bpm. The upper extremity pulses were palpable but lower extremity pulses were weak. Liver and kidney functions were correct and he had no malperfusion signs. He was diagnosed DeBakey Type 3 dissection and situs inversus totalis with CT Angiography (Figure 1). Abdominal ultrasonography determined situs inversus of abdominal organs. CT angiography examination showed that the entry of dissection was found 8 cm distal to the subclavian and it extends up to 3 cm above the celiac artery. Descending aorta was measured 30 mm below the subclavian artery and 29 mm above the celiac artery. Maximum measured of art was 42 mm at the dissection zone.

Under general anesthesia, both femoral arteries were prepped with incision and blunt dissection. 6F introducers were placed in both femoral arteries with Seldinger technique. Left femoral artery was used to place a pigtail catheter for angiography showing an aortic situs inversus with an entry tear of the aortic dissection (Figure 2). Right femoral artery was used to introduce the end graft device. Guide wire was advanced to the ascending aorta through the right femoral artery introducer and exchange catheter was placed on it. Guide wire was removed and carrier wire was sent to the ascending aorta via an exchange catheter. Two different length of thoracic stent graft were used. Proximal stent was 36x150 mm and distal stent was 36x10 mm. Proximal stent graft was placed after right subclavian artery to the descending aorta. Distal stent graft was overlapped from distal zone of the proximal stent graft to the celiac artery.
Stent graft implantation was successful and control angiography showed no end leak. TEVAR was successfully performed with transfemoral approach in cath-lab (Figure 3). After TEVAR procedure patient was followed 2 days in cardiac intensive care unit. Initial follow up of intensive care unit we used Na-Nitroprussid infusion for protection of hypertension. Additionally Metoprolol Tartrate 50 mg two times per day and Valsartan 80 mg per day was started. At 7 days follow up he has no malperfusion and he was discharged with good outcome. A postoperative CT scan performed one month later showed no end leaks or device related complications. (Figure 4).

**DISCUSSION**

The etiology of situs inversus totalis remains uncertain. Situs inversus totalis is a rare autosomal recessive developmental anomaly. Situs inversus totalis is very uncommon and usually diagnosed coincidentally as it does not affect the patient’s life. The recognition of concomitant anomalies, such as in the cardiac, venous, gastrointestinal, and urinary systems, is extremely important because these anomalies may disturb the surgical procedure for the concurrent disease in situs inversus totalis. The coexistence of DeBakey type III dissection has been seldom encountered. The presence of anatomical anomalies significantly increases operative risk. The surgical management of patients with abdominal aortic aneurysm in situs inversus totalis is discussed.

Imaging techniques such as transesophageal echocardiography, contrast-enhanced CT, and magnetic resonance imaging have equal sensitivities of 97% to 98% but variable specificities. Computed tomography (CT) angiography is widely available, has a rapid acquisition time, provides images with high spatial resolution and is the most commonly used imaging modality for anatomical evaluation of the thoracic aorta. The CT procedure is faster and more readily available than MRI and images of both the entire aorta and the aortic branch vessels can be acquired in a single scan. Although Transthoracic echocardiogram (TTE) has a lower overall sensitivity of 59.3% in diagnosing all types of aortic dissection, it may still be helpful in the emergency setting, particularly in hemodynamically unstable patients, where there are sometimes unavoidable delays in getting contrast enhanced CT or more invasive investigations such as transesophageal echocardiography to establish the
diagnosis. However, using TTE, the sensitivity in diagnosing Stanford Type A (or DeBakey Types I and II) dissection is 78.3% for acute and 87.5% for subacute dissections, respectively [6]. The sensitivity in diagnosing Stanford Type B (or DeBakey Type III) dissections by TTE is even lower at 40% and 29.4% for acute and subacute dissections, respectively [6]. Usually we use CT scan to diagnose and surgical decision.

There were seldom cases in the published literature of thoracic aortic dissection or aneurysm that was treated with conventional surgery in situs inversus totalis patients. The decision for either TEVAR or conventional open surgery depended on the patient’s clinical status on admission and on individually present complications with regard to the technical feasibility of the intended procedure [7]. Patients who presented with more proximal aneurysms of the descending aorta in which a sufficient proximal und distal landing zone allowed for endovascular stentgrafting were more likely to be treated by TEVAR while those with extensive aneurysms (often involving the entire thoracic and/or thoracoabdominal aorta and its branches) were treated by conventional open surgery [7]. According to our literature review, this is the first case about situs inversus totalis and DeBakey type III dissection patient whom was treated with TEVAR.

CONCLUSION

Although challenging, conventional aortic surgery can be safely accomplished in patients with situs inversus totalis. Situs inversus with mirror-image of the heart is a rare condition and we thought this is not a handicap for succesfull application of TEVAR.

REFERENCES