



Hybrid Approach of Ruptured Type B Aortic Dissection with an Aberrant Subclavian Artery in a Single Patient with Turner Syndrome: A Case Report

Shin-Ah Son¹, Kyoung Hoon Lim², and Gun-Jik Kim¹

¹Department of Thoracic and Cardiovascular Surgery, School of Medicine, Kyungpook National University, Kyungpook National University Hospital, Daegu, ²Department of Surgery, Trauma Center, School of Medicine, Kyungpook National University, Kyungpook National University Hospital, Daegu, Korea

Turner syndrome, also described as 45, X, may present with most serious cardiovascular anomalies including risk of aortic dissection and rupture. In emergency situation, management for aortic dissection with complicated anatomy accompanying vascular anomaly is challenging. Here, we report a rare case of ruptured type B aortic dissection with aberrant subclavian artery and partial anomalous pulmonary venous connection in a Turner syndrome. Through right carotid-subclavian artery bypass and thoracic endovascular aortic repair, successful hybrid endovascular management correlated with a favorable result in this emergency situation.

Key Words: Aorta, Endovascular procedure, Turner syndrome

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Corresponding author: Gun-Jik Kim

Department of Thoracic and Cardiovascular Surgery, School of Medicine, Kyungpook National University, Kyungpook National University Hospital, 130 Dongdeok-ro, Jung-gu, Daegu 41944, Korea
Tel: 82-53-200-5665
Fax: 82-53-426-4765
E-mail: straightroot@knu.ac.kr
<https://orcid.org/0000-0002-8051-2131>
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INTRODUCTION

Turner syndrome with gonadal dysgenesis and sex chromosome abnormalities is a disorder in which cardiovascular malformations are common [1,2]. Aortic dissection also occurs at a substantially higher frequency in patients with Turner syndrome than in the general population. The occurrences of cardiovascular anomalies and aortic deterioration have been subjects of debate, as have the American Academy of Pediatrics recommendations for the cardiac screening of patients with Turner syndrome [1]. The management of anatomically complicated aortic dissection is rather difficult and not yet widely reported.

Here, we report on the successful management of ruptured type B aortic dissection with an aberrant subclavian artery by using hybrid endovascular procedures in a patient

with Turner syndrome.

CASE

A 32-year-old woman with Turner syndrome (143 cm/49 kg, body surface area [BSA] 1.37 m²) was admitted to Kyungpook National University Hospital center with complaint of sudden-onset back pain. She had been diagnosed with Turner syndrome twenty years prior following examination for her short stature. She was regularly treated with hormone replacement therapy as an outpatient of the obstetrics and gynecology. She felt well, worked as a kindergarten teacher, and was unmarried. On admission, her initial systolic blood pressure and heart rate were 131 mmHg and 68 beats per minute, respectively. Her blood pressures were almost equal in the upper and lower extremities. Her

initial hemoglobin level was 11.8 g/dL, and electrocardiography revealed a normal sinus rhythm.

An initial computed tomography (CT) scan revealed intimal dissection on aortic isthmus level and a dissected flap extending from the left subclavian artery to the descending aorta. An elongated transverse aortic arch (ETA) and an aberrant right subclavian artery (ARSA) branching into the aortic isthmus as unusual anatomic location were seen. Periaortic hematoma formation was suspected of possible extravasation. In addition, a partial anomalous pulmonary venous connection (PAPVC) showed that the vertical vein connected to the left upper lobe drained into the left brachiocephalic vein (Fig. 1A, B).

We planned an arch and descending aorta replacement surgery via an anterolateral thoracotomy under cardiopulmonary bypass. However, the patient was hesitant with

regard to invasive surgery owing to her cosmetic concerns as a single woman. Her back pain gradually improved with analgesics, however, 3 days later, she again complained of acute sharp back pain. Subsequent CT scan revealed active aggravation of ruptured aortic dissection and aneurysmal dilatation on dissected aorta. The false lumen had increased in size to 10% of the aortic maximum diameter, measuring 3.9 cm, and left pleural fluid collection suspicious to hemothorax and periaortic hematoma had increased (Fig. 2A, B).

We decided to perform an emergency operation for the recurrent back pain, enlarging periaortic hematoma and hemothorax.

After consultation with the patient, we initially planned an anterolateral thoracotomy with left heart bypass. However, we determined that it would be too dangerous to clamp on the aortic isthmus near the ARSA owing to the

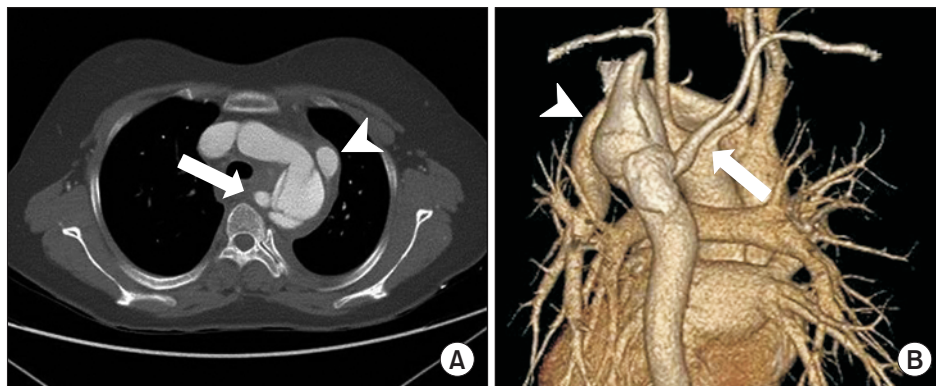


Fig. 1. (A) Initial chest CT scan shows intimal dissection from left subclavian artery level extending to the descending aorta and an aberrant right subclavian artery (white arrow) is derived near aortic isthmus dissecting flap. Partial anomalous pulmonary venous connection (white arrowhead) is running anterior aortic isthmus. (B) Chest CT angiography scan (posterior view) shows vertical vein (white arrowhead) draining from the left upper lobe to the left brachiocephalic vein. Aberrant right subclavian artery (white arrow). CT, computed tomography.

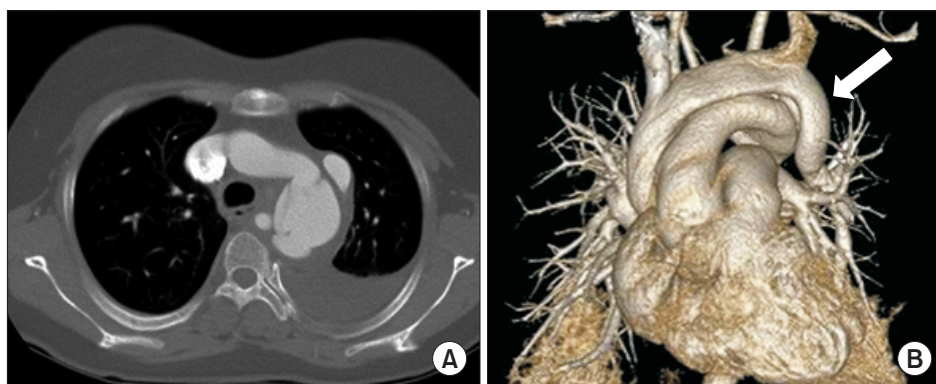


Fig. 2. (A) Follow-up chest computed tomography scan demonstrates active aggravated extravasation of ruptured descending aortic dissection and increasing size of the aneurysmal dilatation. Increased left pleural fluid collection noted, leading to suspicion of hemothorax and periaortic hematoma. (B) Vertical vein drain to left brachiocephalic vein (white arrow) runs directly above the dissecting flap.

frailty of the aneurysmal dilatation on the dissected aorta. In addition, the vertical vein running directly above the dissecting flap might have interfered with the operative field.

Instead of open surgery, we decided to perform emergency thoracic endovascular aortic repair (TEVAR). However, because the patients had the ARSA with a dominant vertebral artery, we decided to perform a carotid to subclavian artery bypass with the dominant vertebral artery before TEVAR.

Under general anesthesia, a right carotid and subclavian artery bypass was performed via supraclavicular transverse incision. A 7 mm Gore-Tex polytetrafluoroethylene Stretch graft was placed between the right carotid and subclavian arteries. After bypass, we exposed the patient's right common femoral artery (CFA) through a longitudinal incision. Through the left CFA, a sizing catheter was inserted with angiography guidance for checking the length. Pre-deploying aortogram revealed ARCA branching from the

aortic isthmus and aneurysmal dilatation on the dissected aorta including the left subclavian artery (Fig. 3A). An S&G SEAL thoracic stent graft (24×24×110 mm; S&G Biotech, Seongnam, Korea) was advanced through the right CFA. During stent graft insertion, a slightly strong resistance was encountered; however, post-procedural aortogram revealed no peripheral arterial injury. After stent graft deployment, aortography showed laminar blood flow through the devices and the thoracic aorta with complete exclusion of the dissecting aneurysm (Fig. 3B). There were no complications during the procedure, and the oversizing of stent grafts relative to the aorta did not exceed 10% to provide adequate radial force to keep the deployed stent grafts in place. The total operative time was 245 minutes, and the total contrast agent volume was 90 mL.

Postoperatively, the blood pressure in both arms was nearly same and there was no evidence of ischemia. After postoperative day (POD) 5, CT scan revealed a well-func-

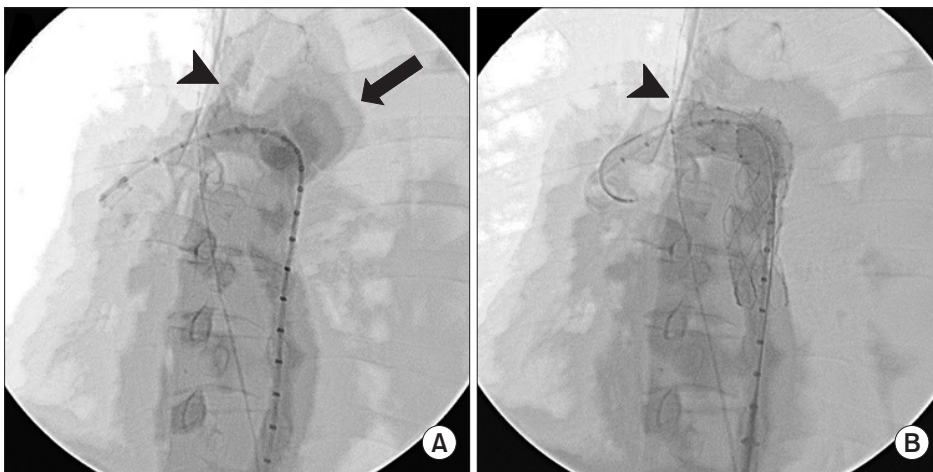


Fig. 3. (A) Pre-deploying aortogram shows the left common carotid artery (black arrowhead) and aneurysmal dilatation on the dissected aorta including the left subclavian artery (black arrow). (B) Post-deploying aortogram reveals the stent graft is well positioned just after common carotid artery (black arrowhead) without flow impairment.

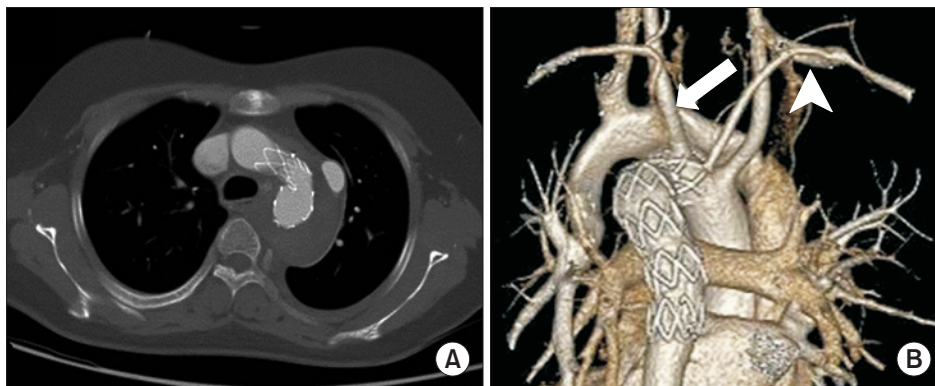


Fig. 4. (A) Postoperative chest CT scan demonstrates that endovascular stent graft in descending aorta is well functioned and complete exclusion of the dissecting aneurysm. (B) Chest CT angiography scan (posterior view) shows complete occlusion of both subclavian artery orifice with distal run-off reserved from collateral flow and the left common carotid artery (white arrow) has no flow impairment. White arrowhead shows well-functioning right carotid and subclavian artery bypass. CT, computed tomography.

tioning aortic isthmus stent graft without endoleaks, and complete occlusion of both subclavian artery orifices with distal run-off reserved from collateral flow (Fig. 4A, B). The patient was discharged on POD 6 without complications and is planning follow-up examination of the PAPVC at the cardiac and cardiovascular clinics.

DISCUSSION

Turner syndrome, or monosomy X, results from the complete or partial monosomy of the X chromosome. This is a relatively common, affecting >1 in 2,500 live female births. Most common features include short stature and gonadal dysgenesis, however, the most serious clinical aspects are due to congenital cardiovascular anomalies [1,3-6]. The most common congenital cardiac anomalies in surviving patients with Turner syndrome include bicuspid aortic valve and aortic coarctation. Multiple other structural alterations have also been reported [1-3]. Approximately 50% of asymptomatic women with Turner syndrome also have evidence of abnormal cardiovascular development [4], however, the cause of this abnormal development remains unknown [6].

Turner syndrome also leads to anatomic vascular anomalies, such as PAPVC, persistent left superior vena cava (SVC) or ETA [2,4,6]. Ho et al. [4] reported vascular anomalies in Turner syndrome detected on magnet resonance angiography, including PAPVC and persistent left SVC, in 13% of cases. In addition, 10% had ARSA, and 1%-2% had septal defects or mitral valve prolapse. In previous reports, Turner syndrome was also associated with other arterial and venous anomalies, notably PAPVC [1,4,7,8]. PAPVC was not found in normal female controls. Ho et al. [4] reported that none of the patients presented with clinical features referable to a significant left-to-right shunting. Moreover, PAPVC was most commonly observed from the left upper lobe to the left brachiocephalic vein and without clinical features, which also differs from non-Turner syndrome patients, in whom it is typically right-sided [4,9].

Of the aberrant arch vessel anomalies, ARSA is found in 8%-10% of women with Turner syndrome, but not in control subjects. This anomaly has been reported to occur in 0.4%-2% of the normal population, with the majority remaining asymptomatic [4,10]. Treatment of an aberrant subclavian artery with symptoms is indicated for symptomatic relief and for prevention of complications. In most cases, endovascular stent insertion has become a treatment option that, might reduce mortality and morbidity.

Gravholt et al. [3] estimated the incidence of aortic dissection in Turner syndrome to be 36/100,000, compared to 6/100,000 in the general population. The median age at

onset of aortic dissection was 35 years (range, 18-61 years) according to the Danish Registry. Most patients with Turner syndrome who develop aortic dissection have aortic valve disease and/or coarctation. In these patients, about two-third of dissections begin in the ascending aorta (Stanford type A) and one-third of dissections originate in the descending aorta [3,11].

Turner syndrome is frequently associated with a high frequency of aortic dilation that is a specific kind of aortopathy [11]. Of these, ascending aortic dilation is the most common, occurring in 15%-30% of cases [2].

An influential early study suggested that aortic disease in Turner syndrome was similar to that in Marfan syndrome, characterized by 'cystic medial degeneration', which is the common pathophysiology for aortic root dilation [11-13]. However, there is no evidence to suggest that patients with Turner syndrome experience progressive dilatation of the aorta over time, in the absence of predisposing risk factors, as seen in Marfan syndrome.

It remains unknown whether the dilatation of the ascending aorta predicts aortic dissection in Turner syndrome as it does in Marfan syndrome, and there are no current guidelines on what specific aortic diameter measurement should provoke concern in Turner syndrome [14]. Because body size is a major determinant of normal aortic dimensions, it may not be appropriate to apply standards derived from adult men to small women with Turner syndrome, many of whom have an adult height of >140 cm and BSA of nearly 1 m². Two major strategies have been proposed to circumvent the size problem [15].

Little is known of the prodrome of aortic dissection in Turner's syndrome. To gain a better understanding of the natural history of Turner's syndrome, the International Turner Syndrome Aortic Dissection Registry has been established. It remains unclear whether girls with Turner syndrome should undergo surveillance, with routine electrocardiogram and blood pressure testing [13]. Some reports recommended that Turner syndrome with known congenital heart disease should undergo comprehensive cardiac magnetic resonance imaging evaluation, close cardiologic follow-up, control of blood pressure, and a trial of angiotensin antagonist versus beta-blocker [6,14].

Cooper et al. [15] successfully performed open and endovascular techniques for the treatment of thoracic aortic dissection with ARSA in a patient with Marfan syndrome. They reported that type B dissections may be rarely associated with an underlying ARSA and the aberrant vessel may undergo aneurysmal dilatation (Kommerell's diverticulum) or the dissected aorta may become aneurysmal.

Ruptured type B aortic dissection in a Turner syndrome patient with ARSA is very rare. In particular, endovascular

approach of aortic rupture in a patient with Turner syndrome remains controversial because this condition seems to have aortopathy similar to that in Marfan syndrome. So, in emergency situation, the decision for treatment of aortic rupture is a large problem.

Generally, the more invasive approach via a median sternotomy or thoracotomy is better tolerated in younger patients. In particular, we determined that the anterolateral thoracotomy with a left heart bypass could not be performed, as we considered it too dangerous to place a clamp on the aortic arch proximal to the left subclavian artery owing to the frailty of the aorta. Moreover, the operative view for the proximal aortic clamp was not secured because the vertical vein was running directly above the dissecting aneurysmal aorta. Further, there was the possibility of damaging the vertical vein, which could result in pulmonary congestion and catastrophic hemorrhage. Instead of left thoracotomy, we chose a hybrid approach that combines the conventional surgical procedure with an interventional stent graft. Although endovascular approach is generally recognized as an alternative to open surgery, it is associated with common postoperative adverse events, such as endoleak, stent graft migration, and graft infection, which necessitate regular follow-up that could be troublesome for patients, especially young patients.

This hybrid approach might be advantageous because despite the need for regular follow-up, the patient could return to society quickly and the procedure is less invasive.

Considering the emergency operation and because

PAPVC was otherwise not evident clinically, we decided to keep her on follow-ups for her lesions and to examine her left-to-right shunt measurements.

In emergency situations, a hybrid approach for a patient with Turner syndrome provides a less invasive, safe alternative for ruptured acute aortic type B dissection. We believe that anatomically complicated aortic type B dissection may be treated successfully with a new procedure that could prevent aortic deterioration in Turner syndrome. Thus, this report underscores the importance of regular cardiologic follow-up of asymptomatic Turner syndrome, and illustrates the need for lifelong follow-up and patient and family education about the symptoms of aortic dissection.

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ORCID

Shin-Ah Son

<https://orcid.org/0000-0003-3317-6857>

Kyoung Hoon Lim

<https://orcid.org/0000-0002-6842-7129>

Gun-Jik Kim

<https://orcid.org/0000-0002-8051-2131>

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