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Case Report

Acute Aortic Dissection in a 35-year-old Woman With Turner Syndrome: A Case Report

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ABSTRACT

Turner Syndrome occurs when one of the X chromosomes is missing, either partially or completely. Heart defects associated with some cases of Turner syndrome can increase the risk of severe, life-threatening complications, including pulmonary hypertension and aortic dissection.

A 35-year-old woman, a known case of Turner syndrome, presented with acute dissection in the ascending aorta (Type A Stanford). The patient had a successful aortic valve repair surgery with a Dacron tube graft interposition. The risk of aortic dissection in Turner syndrome is 100 times greater than that in the general population.

Diagnosis was made by transthoracic echocardiography and computed tomography angiography. Due to the small size of the femoral artery, cannulation was done in the innominate artery. Cardiopulmonary bypass was established, and systemic cooling was initiated to a temperature of 25 ℃. The patient was discharged from the hospital in good condition. ***(Iranian Heart Journal 2023; 24(2): 114-117)***

KEYWORDS: Aortic dissection, Adult congenital heart disease, Bicuspid aortic valve, Turner syndrome, Type A Stanford

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A young woman, a known case of Turner syndrome, presented to our hospital with signs and symptoms of Stanford Type A acute aortic dissection. The patient had no history of aortic valve disease or aortic aneurysm in her previous echocardiographic examination, done a year earlier. She had a sudden onset of anterior chest wall pain and interscapular and shoulder pain starting 8 hours before admission. In her transthoracic echocardiography examination, an ascending aorta intimal flap was seen starting above the sinotubular junction. In the evaluation of the aortic valve, moderate functional aortic regurgitation was noticed, and the aortic diameter was measured to be 4.5 centimeters. In addition, the extension of the aortic false lumen could be seen in the aortic arch and the proximal descending aorta.

After a median sternotomy and pericardium opening, a 150 mL blood clot was found, and acute aortic dissection in the ascending aorta was confirmed. The femoral artery was the first to be tried for arterial cannulation; nonetheless, due to its small size, it could not be done. Subsequently, after the

complete dissection of the descending aorta and a branch of the aortic arch, a safe site in the innominate artery was prepared for cannula insertion. Venous drainage was done with a 2-stage venous cannula in the right atrial appendage. Cardiopulmonary bypass was initiated. Deep hypothermia up to 25 ℃ was induced. Following the cross-clamping of the aorta, a true lumen, a false lumen, and the site of the aortic rupture 3 centimeters above the ostium of the left main were detected. In the evaluation of the aortic valve, the aortic leaflet had a normal appearance with intact function; nevertheless, the diameter of the aortic valve annulus was mildly increased, which was corrected by suture commissuroplasty. Next, the entire length of the ascending aorta was replaced with a Dacron tube graft (No. 25) between the sinotubular junction and the proximal aortic arch. The postoperative course of the patient was uneventful. Her follow-up echocardiographic examination showed a mild residual aortic regurgitation and the normal function of the Dacron tube graft.

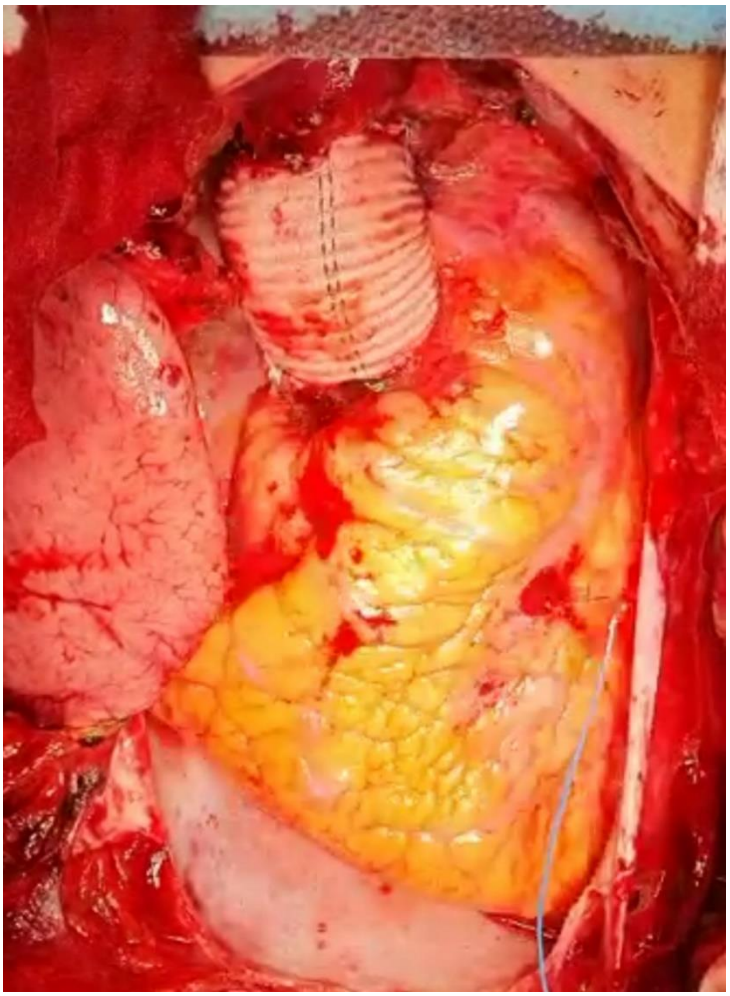

Figure 1: The image shows the interposition of the tube graft in the treatment of the dissected aorta.

DISCUSSION

Acute type A aortic dissection is the most lethal presentation of the cardiovascular manifestation of Turner syndrome. [1-3]
Turner syndrome is a common genetic disease that involves young women. The risk of acute aortic dissection in patients with Turner syndrome is 100 times greater than that in the general population. [4, 5]
Turner syndrome is caused by complete or partial monosomy of the X chromosome during fetal growth. [6-9]
The most common presentations of Turner syndrome are short stature and premature ovarian failure. In the evaluation of the intelligence of patients with this genetic disorder, full-scale intelligence is usually normal, with verbal scores typically higher than performance IQ scores. [10-12] Cardiac disease in Turner syndrome consists of bicuspid aortic valve (30%) and aortic aneurysm or dissection (15%). In some studies, Turner syndrome is the most common etiology of acute aortic dissection in young women. Other cardiac disorders with lower frequencies are aortic coarctations, persistent left superior venae cavae, and anomalous connections in the pulmonary vasculature. [13, 14]

Conflict of Interest: We declare no conflicts of interest.

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