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Case Report / Olgu Sunumu

Successful surgical repair of critical ascending and descending aortic aneurysms at nine-year intervals in Wiskott-Aldrich syndrome: A case report

Wiskott-Aldrich sendromunda dokuz yıllık aralıklarla kritik çıkan ve inen aort anevrizmalarının başarılı cerrahi onarımı: Olgu sunumu

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ABSTRACT

Wiskott-Aldrich syndrome is an uncommon X-linked inherited disorder related to primary immunodeficiency, infections, eczema, and thrombocytopenia. A 21-year-old male patient with this syndrome underwent descending aortic aneurysm repair at the age of 12. The patient had ascending aortic aneurysm with aortic valve regurgitation and surgical aortic root replacement was performed. To the best of our knowledge, this is the first case with Wiskott-Aldrich syndrome operated due to aneurysms development in different segments of the thoracic aorta in both childhood and young adult periods.

Keywords: Aortic aneurysm, aortic valve, thoracic aorta, Wiskott-Aldrich syndrome.

ÖZ

Wiskott-Aldrich sendromu, primer immün yetmezlik, enfeksiyonlar, egzama ve trombositopeni ile ilişkili, nadir görülen X'e bağlı kalıtsal bir hastalıktır. Bu sendromlu 21 yaşında erkek hasta 12 yaşında iken inen aort anevrizması onarımı geçirmişti. Hastanın çıkan aort anevrizması ve aort kapak yetersizliği vardı ve cerrahi olarak aort kökü replasmanı yapıldı. Bilgimiz dahilinde, Wiskott-Aldrich sendromlu bu olgu, hem çocukluk hem de genç erişkinlik döneminde torasik aortun farklı segmentlerinde anevrizma gelişmesi nedeniyle ameliyat edilen ilk hastadır.

Anahtar sözcükler: Aort anevrizması, aort kapağı, torasik aort. Wiskott-Aldrich sendromu.

Wiskott-Aldrich syndrome (WAS) is a rare X-linked recessive inherited disorder that is a primary immunodeficiency characterized by infections, eczema, and thrombocytopenia.[1] A relationship with vasculitis or aneurysmal formation has occasionally been reported in case reports.[2] In the literature, only a few patients treated surgically for aortic aneurysms have been described.[3-7]

In this article, we present a young male case who underwent a descending aortic aneurysm repair nine years ago and required a second operation due to ascending aortic aneurysm and aortic valve insufficiency. To the best of our knowledge, this is the first WAS case operated due to aneurysm development in different segments of the aorta at different ages.

CASE REPORT

A 21-year-old male patient was referred to our institution from an external center with a preliminary diagnosis of the ascending aortic aneurysm with aortic valve insufficiency. The patient had complaints of fatigue with physical activity and intermittent chest pain for the last six months. He was followed by the pediatric hematologist with the diagnosis of WAS since the age of 10. In his medical history, he underwent thoracic aortic aneurysm repair surgery nine years ago, and histopathological diagnosis of chronic aortitis after surgery. The bone marrow transplantation could only be performed seven years ago due to the absence of a match-related donor.

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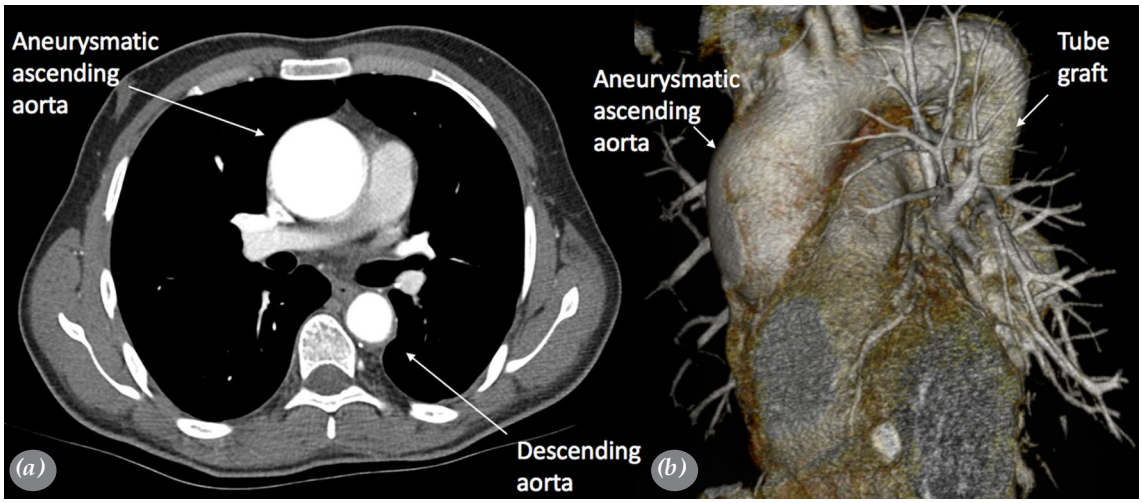

Figure 1. (a) Aneurysm of the ascending aorta is observed on preoperative CTA and **(b)** its three-dimensional reconstruction.
CTA: Çomputed tomographic angiography

The ascending aortic aneurysm was confirmed by a computed tomographic angiography (CTA) scan and the maximum diameter was measured 50 mm in the ascending aorta and 35 mm in the aortic arch (Figure 1). A tube graft with a diameter of 22 mm from the previous operation was observed in the descending thoracic aorta (Figure 2). The preoperative transthoracic echocardiogram showed severe aortic valve insufficiency with ascending aorta dilatation. Left ventricular volume was increased (left ventricular end-systolic dimension was 43.5 mm and left ventricular end-diastolic dimension was 64.5 mm), but ventricular function was preserved, with an ejection fraction of 60%. In the preoperative evaluation of the patient by the hematology department, there was no obstacle in terms of the operation.

An elective operation was performed by median sternotomy. The aortic cannula was inserted into the truncus brachiocephalicus to provide space for distal anastomosis. After the right atrial two-stage venous cannula and left heart venting cannula was inserted, cardiopulmonary bypass (CPB) was initiated. The ascending aorta was cross-clamped and incised vertically. The aortic wall was widely dilated, thickened and diffusely infiltrated by internal inspection. The del Nido cardioplegia was given for the myocardial protection via the coronary osteal cannulas. The aortic valve was tree leaflet, the leaflets were very thin, elongated, and the inflammation of the aortic wall extended to the aortic root (Figure 3a).

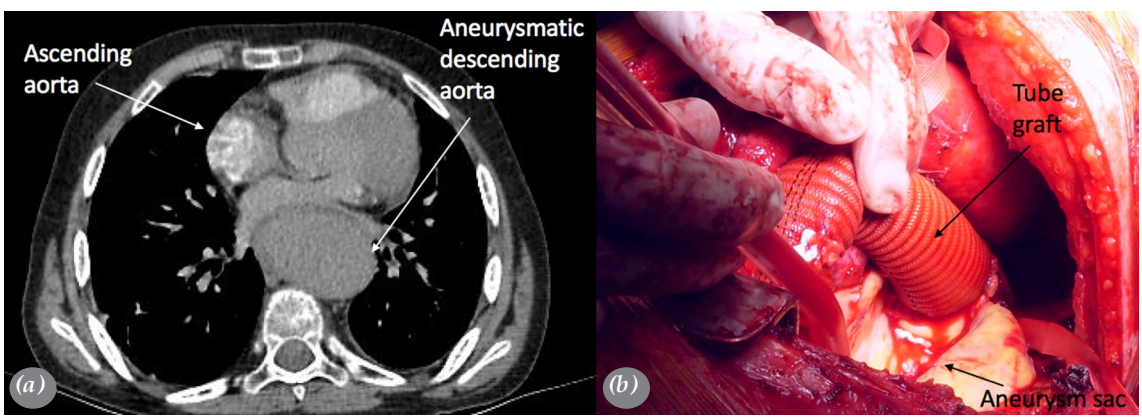

Figure 2. (a) The descending aortic aneurysm is observed on CTA performed before the first operation. **(b)** The image of the repairing the previous descending aortic aneurysm with the tube graft interposition.
CTA: Computed tomographic angiography.

After the surgical inspection of the aortic valve and root, we decided not to consider aortic valve-sparing operations as an option. The ascending aorta was transected from the level of sinotubular junction, the coronary buttons and the aortic leaflets were excised. The root was replaced with a 25-mm mechanical valved conduit under moderate hypothermia at 28°C, and the coronary buttons were sutured to the graft with the fenestrated Teflon® patch support (Figure 3b). The distal anastomosis of the graft was done under total circulatory arrest at the level of the truncus brachiocephalicus ostium proximal to the aortic arch (Figure 3c). The period of total circulatory arrest was 17 min, aortic cross-clamping was 60 min, and CPB was 161 min.

The postoperative recovery of the patient was uneventful, and he was extubated within 6 h after surgery. He was transferred to ward on postoperative Day 1, and discharged from hospital on postoperative Day 6. The postoperative echocardiogram showed a competent prosthetic valve and a normal left ventricle size. Histopathological evaluation revealed chronic aortitis by patchy inflammatory lymphocytes, plasma cells, and eosinophils. After a follow-up of four months, he is currently in a good clinical condition (Figure 3d).

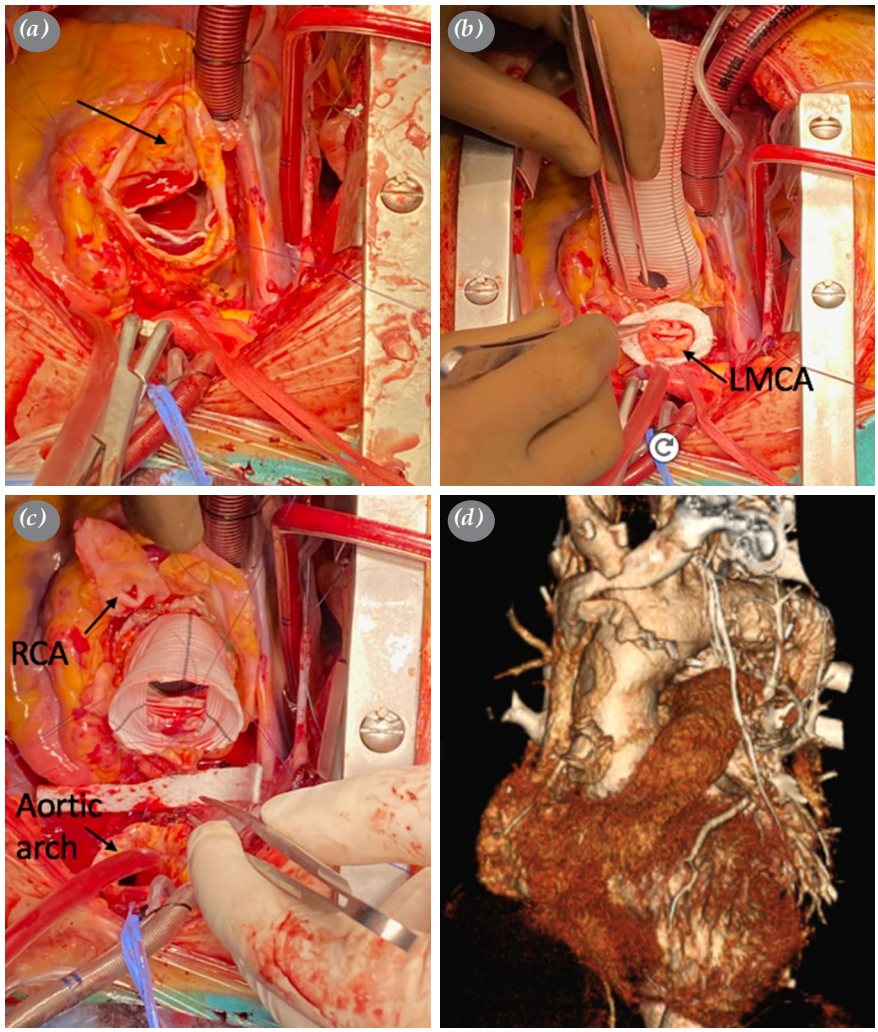

Figure 3. (a) The arrow shows the dilated, thickened and diffusely infiltrated aortic wall. **(b)** The image of suturing the left main coronary button to the graft with a fenestrated Teflon® patch support. **(c)** The image of suturing the distal anastomosis of the graft proximal to the aortic arch. **(d)** CTA scan of the patient with three-dimensional reconstruction at the postoperative third month.

LMCA: Left main coronary artery; RCA: Right coronary artery; CTA: Computed tomographic angiography.

DISCUSSION

Wiskott-Aldrich syndrome is a primary immunodeficiency disease due to the short arm of the X chromosome. The clinical presentation is characterized by bleeding complications due to thrombocytopenia, recurrent infections, eczema, and autoimmune vasculitis.[1]

Bone marrow transplant and/or splenectomy can improve the life expectancy of patients with WAS.[8] Previously, life expectancy for these patients was 6.5 years, while the average survival since the initiation of splenectomy and bone marrow transplantation has increased to 25 years.[3,8] As a result of their longer survival, these patients are at a higher risk for lymphoproliferative diseases, vasculitis, and aneurysm formation.[2] Our patient also a bone marrow transplant at the age of 14 and no problem was observed in his subsequent hematological follow-up.

Aneurysmal formation of the aorta is rare, but it is a life-threatening complication. Death risk of the aneurysmal rupture is increased in WAS due to thrombocytopenia and impaired platelet function. Moreover, the surgical course is more complicated in these patients due to the high probability of postoperative infectious and bleeding complications.[9]

Pellier et al.[10] conducted a study on 38 children with WAS and five of them had aortic aneurysm. Only one had symptom as an acute chest pain and he underwent aortic root replacement by the Bentall method due to ascending aorta aneurysm with aortic valvular insufficiency. Two patients were alive without symptoms and the remaining two patients died due to unrelated complications. They reported that aortic aneurysm progression was not clear in WAS whether it was caused by an infectious and /or inflammatory process, and suggested that large vessels should be systematically evaluated for aneurysm formation in the patients with WAS. We believe that WAS patients should be investigated in terms of aortic aneurysm, as in our case and this study, and that all patients with symptomatic aneurysm should be operated on.

To the best of our knowledge, surgical management of thoracic aortic aneurysm has been reported only in five young adults in the literature.[3-7] A descending aortic replacement was performed in our patient at the age of 12 due to a descending aortic aneurysm.[11] Our patient is the first patient in the literature operated due to aneurysm development in different segments of the thoracic aorta in both childhood and young adult periods. And, our case is the first case who developed an aortic aneurysm after bone marrow transplantation and was operated.

As for the surgery, we were unable to use valve-sparing operations due to excessive inflammation of the aortic wall.[9] Therefore, we performed aortic root replacement with Bentall method and replaced the all of the ascending aorta. We used a fenestrated Teflon® patch in the coronary anastomosis and the Teflon® felt in the distal suture line of the graft to prevent bleeding. There was no postoperative bleeding or any other complication in our patient and he was discharged on postoperative Day 6 without any problem. We believe that having a bone marrow transplant contributed to the uneventful postoperative period and rapid recovery of our patient.

In conclusion, the occurrence of aneurysm in Wiskott-Aldrich syndrome patients with improved survival due to large vessel vasculitis is not infrequent. This may be the result of a latent infection or an autoimmune process. Since surgical intervention may be required before life-threatening aortic dilatation begins, we suggest that all patients with Wiskott-Aldrich syndrome need regular clinical or radiological examination of the aortic root / valve and all segments of the thoracic aorta. We believe that if advanced aneurysm develops, surgery can also be successfully performed in this patient population.

Patient Consent for Publication: A written informed consent was obtained from patient.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

Author Contributions: Idea/concept, data analysis: M.A.Ö., A.Ş.; Study design: A.Ş.; Control/supervision: M.Ç., O.Y.; Data collection: M.A.Ö., H.F.A.; Literature review: M.Ç., O.Y., H.F.A.; Writing the article: M.A.Ö., A.Ş.; Critical review: A.Ş.; References and materials: M.A.Ö.

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