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Aneurysm of ascending and descending aorta in a 10-year-old-boy with Wiskott-Aldrich syndrome

Wiskott-Aldrich sendromlu 10 yaşındaki erkek çocukta çıkan ve inen aortada anevrizmatik dilatasyon

Introduction

Wiskott- Aldrich Syndrome (WAS) is a recessive genetic disorder linked to the X-chromosome characterized by immune deficiency, eczema and thrombocytopenia. To the best of our knowledge, a few cases of vasculitis or aneurysmal formation have been reported in this syndrome, but the association has not been well established (1-6). We report a patient with WAS and extensive aortitis causing severe aneurysmal dilatation in the everywhere of the aorta who underwent successful first stage operation involving replacement of ascending aorta.

Case Report

A 10-year-old boy had been followed up with the diagnosis of chronic idiopathic thrombocytopenic purpura (ITP) for 2 years. There was no history of eczema and repeating infections suggesting immunodeficiency. Because of persistent caught in the last two months, chest X-ray was performed and it suggested an ascending aortic aneurysm (Fig. 1). Echocardiography showed mild aortic regurgitation without aortic stenosis and aneurysmatic dilatation of ascending and descending aorta. 3D computed tomography revealed dilatation of the ascending, descending and abdominal aorta with extreme calcification and plaque (Fig. 1). He had two maternal cousins with the diagnosis of WAS. One of them had died with septicemia after splenectomy. There was no history of aneurysm in his cousins. Flow cytometry showed absence of WAS protein. We planned two-stage operation beginning with replacement of the ascending aorta due to high risk of rupture. Pulse methylprednisolone therapy (30 mg/kg/day for 3 days) was tried to correct the thrombocytopenia prior to cardiac surgery. The platelet count increased from 45.000 to 103.000/mm^3. 0.5 gr/kg IVIG was administered monthly. Apheresis thrombocytes were also given before the operation. He underwent valve sparing surgery. Vascutek® graft (24 mm) was replaced to aortic root and ascending aorta (Fig. 2). The luminal surface of the aorta was found to be covered by ulcerated and calcified necrotic plaques. Postoperative recovery was uneventful and no excessive

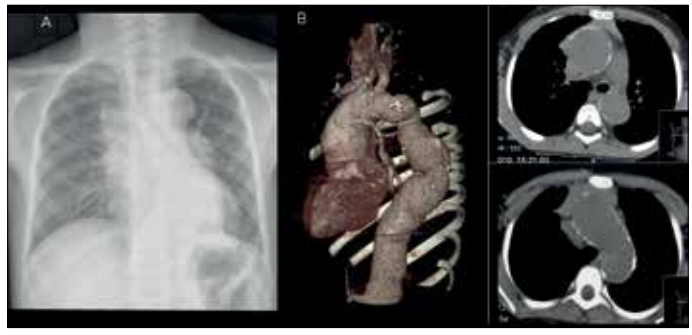

Figure 1. A) Preoperative roentgenogram shows the dilated thoracic aorta; B) Reconstructed three-dimensional computed tomography shows significant dilatation of the ascending, descending and abdominal aorta with calcification and plaque and transverse computed tomographic image demonstrates the dilated ascending and descending aorta with medial calcification

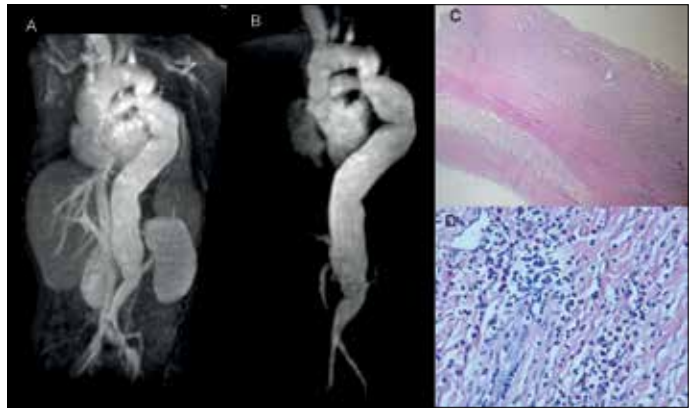

Figure 2. A) Preoperative MRI showing dilatation throughout the whole aorta; from the aortic root to the iliac bifurcation (the diameter at the ascending thoracic aorta was 47.5 mm, aortic arch 31 mm, descending thoracic aorta 38 mm and abdominal aorta 36 mm); B) Postoperative MRI revealed a satisfactory repair the aneurysm of ascending aorta; C) Degenerating changes inflammation and hyalinization in the full thickness of the aorta (HEX40); D) Mixed inflammatory inflammation with eosinophil leucocytes (HEX200)
MRI - magnetic resonance imaging

bleeding was observed. He was discharged on the postoperative 8th day. Six months after surgery, he remains stable. Graft replacement of the distal aortic arch and the descending aorta will be performed later.

Histological examination of the aorta revealed a markedly inflammation, fibrosis and hyalinization in all thickened layers of the aorta. Inflammation was dominantly mononuclear but also contained eosinophils and polymorpholeucocytes (Fig. 2).

Discussion

Systemic arthritis and aneurismal dilatation of the aorta has rarely been reported in the WAS syndrome. To the best of our knowledge, a few adult patients (1-4) and children (5, 6) have been previously reported in the literature. The pathogenesis of aortic aneurysms remains unclear. Inflammatory aortitis is considered a possible etiology of the aortic lesions (3). Aneurysms could be found in everywhere of the aorta such as only in the ascending aorta, or both in the ascending and descending aorta. In our case, aneurysm was severe and widespread through all parts of the aorta till to the iliac bifurcation. In a few patients, two -stage surgery was performed (2, 4) whereas Bernabeu et al. (1) presented a 33-year- old man with WAS who underwent ascending aorta, aortic arch and descending aorta aneurysm repair in a single stage operation. We also planned two-stage operation. The first stage involved replacement of the ascending aorta which was completed successfully. Second operation for distal aortic arch and descending aorta will be performed in the future. The risk of death from aneurismal rupture was seemed to be higher in our patient because of thrombocytopenia and impaired platelet function. Although surgical management of these patients is considered more complex than the general population, surgical intervention should not be delayed.

We think that aneurysm formation and vasculitis may be more common in WAS than reported. Recently, Pellier et al. (6) reported that they have identified aortic aneurysms in 5 of 38 patients with WAS (13%) detected during childhood at the age of 10 to 16 years during childhood.

Conclusion

We suggest that children with WAS should be examined with echocardiography and MRI periodically to evaluate aneurysms of the aorta and surgical intervention shouldn't be delayed when it is indicated.

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Vacuum-assisted closure for skin infection in a patient with Berlin Heart Excor biventricular assist device

Berlin Heart Excor sol ventrikül destek cihazı takılmış bir hastada oluşan deri enfeksiyonunun vakum destekli kapama sistemi ile tedavisi

Introduction

Skin infection on cannulation and driveline sites is a serious and difficult complication of ventricular assist device (VAD) implantation procedures. Management of this complication is important to improve the morbidity and reduce the mortality rates (1). Vacuum assisted closure (VAC) is an effective tool for treatment of chronic wounds with application of continuous suction, which accelerates the healing process (2). We have used VAC for a patient with Berlin Heart Excor biventricular assist device, which had persistent skin infections during 14 months of postimplantation period until transplantation.

Case Report

Fifty two year old male patient was diagnosed as congestive heart failure 8 years ago. He had an implantable cardioverter-defibrillator implant in 2009. He was included to our transplantation program in 2010. In April 2010, he was hospitalized in the intensive care unit due to decompensated heart failure despite maximal medical treatment and intraaortic balloon pumping (IABP) was initiated. After significant improvement, he was weaned from the IABP and transferred to the ward. Unfortunately, he had a ventricular fibrillation attack, which was followed by cardiac arrest. He survived with effective cardiopulmonary resuscitation and no neurological deficit was present. Berlin Heart Excor biventricular assist device was implanted in May 2010. The procedure was uneventful including the postoperative period and he was discharged home in June 2010.

During periodical visits, skin infection on cannulation sites was diagnosed. There was significant amount of pus around the cannula. He was re-hospitalized and daily wound care with silver patch dressings was applied. These are commercially available wound dressing patches combined by silver, alginate and maltodextrin (Algidex Ag® Silver Alginate Wound Dressing, DeRoyal Industries, U.S.A.). Systemic treatment with antibiotics was consulted with the department of infectious