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Available Online Date/Çevrimiçi Yayın Tarihi: 18.09.2012

doi:10.5152/akd.2012.225

Acute aortic dissection in a 10-year-old boy with bicuspid aortic valve

Biküspit aort kapağı olan 10 yaşında erkek çocukta akut aort disseksiyonu

Introduction

Acute dissection is an unusual complication of ascending aortic aneurysm in childhood. Although progressive dilatation of the ascending aorta has been described in pediatric patients with bicuspid aortic valve (BAV), the occurrence of aortic dissection is rare (1-5).

Case Report

A 10-year-old boy presented with severe "tearing" type chest pain for 2 hours. The patient had no trauma, infection, previous cardiothoracic surgery or systemic disorder. Blood pressure and heart rate were 146/88 mmHg and 90 beats/minute, respectively. Heart sounds were diminished on auscultation. Electrocardiogram showed sinus rhythm without ischemia and low-voltage in all derivations. Chest X-ray demonstrated an enlarged mediastinum. Transthoracic echocardiography (TTE) showed dilatation of the ascending aorta and mild pericardial effusion without tamponade (Fig. 1). The diameters of the aortic annulus, sinotubular junction (STJ) and ascending aorta were 18, 28 and 44 mm, respectively. The patient was immediately transferred to intensive care unit, and transesophageal echocardiography (TEE) revealed a dissection flap above the STJ (Fig. 2, Video. See corresponding video/movie images at www.anakarder.com). The dissection extended from the STJ to the origin of the brachiocephalic artery (BCA). The aortic arch and origins of the coronary arteries were normal. The aortic valve was bicuspid with normal function. Therefore, an emergent surgery was performed. During operation, we observed giant aneurysm of the ascending aorta (Fig. 3A). Cardiopulmonary bypass was established via the right axillary and femoral arteries, and femoral vein. After cardiac arrest and aortic incision, true and false lumens were clearly exposed (Fig. 3B). We observed that the intimal tear was above the STJ and extended up to the origin of the BCA. The aortic sinuses and coronary orifices looked normal. We explored the aortic arch during total circulatory arrest and confirmed that the dissection did not extend distally to the BCA. Supracoronary graft interposition with a Dacron graft was performed uneventfully. The patient was discharged on postoperative day 14.

At 6-month follow-up, the patient remained normotensive under medical treatment, and TTE showed normal aortic valve functions. In chromosomal analysis, there was no genetic syndrome as an underlying cause for dissection.

Discussion

The etiology of aortic dissection in children and young adults includes mostly hypertension in up to 80%, followed by BAV in 7-14% (1). The prevalence of BAV is 4.6 in 1000 live born neonates with a higher prevalence in male neonates than in female neonates (6). In a

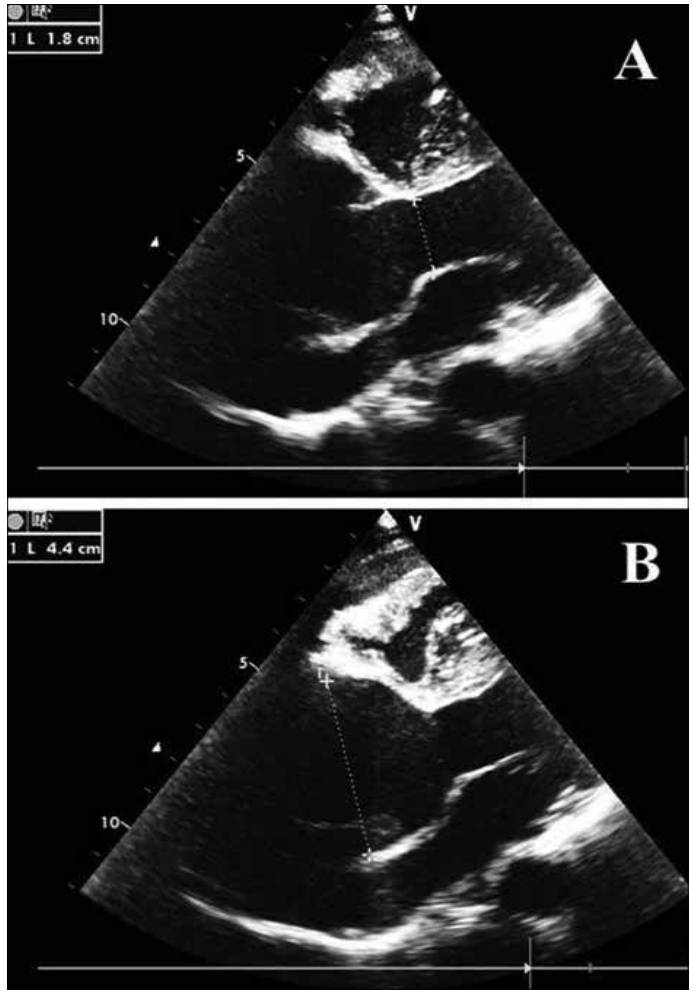

Figure 1. Transthoracic echocardiography demonstrates dilatation of the ascending aorta and pericardial effusion

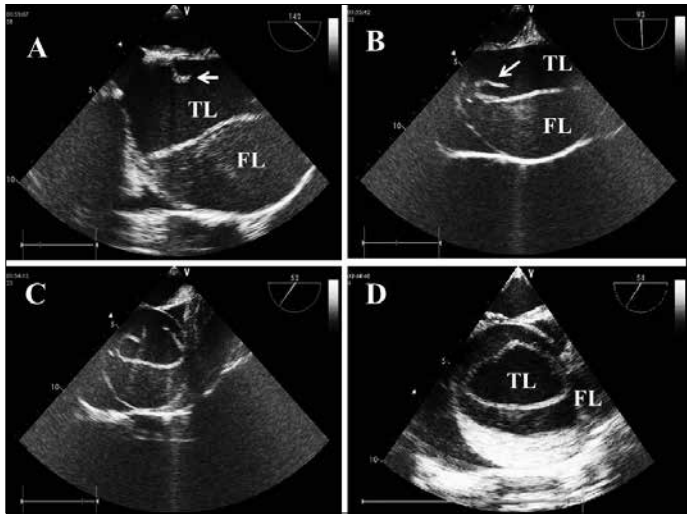

Figure 2. Transesophageal echocardiography demonstrates dissection flap (white arrow) in the ascending aorta. This reveals true and false lumen after dissection of the aorta in parasternal long (A, B) and short axis (C, D) views. Note posterior localization of intimal tear in parasternal short axis view at the level of sinotubular junction (C)
FL-false lumen, TL- true lumen

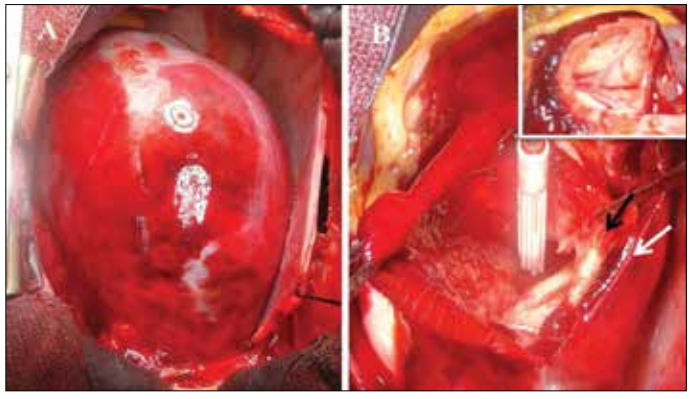

Figure 3. Operative view demonstrates enlargement of the ascending aorta after pericardiotomy incision (A). Macroscopically, aortic dissection caused separation of the intimal (black arrow) and adventitial (white arrow) layers from each other (B). The tip of the sucker was inside the aortic tear in the posterior aortic wall. Bicuspid aortic valve (inset) was confirmed during operation

survey with healthy primary-school children, BAV presents in 0.5% of the study population (7). Patients with BAV have greater increases in aortic dimensions than those with trileaflet valves and have a higher risk of dissection (2, 3, 8). Although the clinical course of ascending aortic dilatation is relatively benign, follow-up for progressive dilatation is recommended (3). Children with Marfan syndrome, genetic disorders such as Ehler-Danlos or Turner's syndrome, fibrillin gene mutations, chest trauma and giant cell arteritis should also be followed-up.

On diagnosis of dissection, computed tomography or TEE can be the initial modality of choice. Transesophageal echocardiography is highly diagnostic in aortic dissection. It helps to visualize separate lumens and intimal tear with differential blood flow. Although TTE is useful in identifying proximal aortic dissection, it can be limited in visualizing the distal ascending, transverse and descending aorta.

Urgent surgery is indicated in acute type-A aortic dissection to avoid lethal complications such as complete rupture, pericardial tamponade and myocardial ischemia due to coronary obstruction. Composite graft implantation with mechanical valve (Bentall procedure) can be performed if the aortic root, coronary orifices and aortic valve are involved. In pediatric population, this procedure carries 2 disadvantages including long-term anticoagulation and difficulty in implanting an adult-size composite conduit. In such cases, Ross operation is an alternative approach that provides freedom from thromboembolism, without anticoagulation, and potential of progressive growth of neo-aortic valve. However, this approach may change single-valve disease (aortic) to 2-valve disease (aortic-pulmonary). Valve-sparing surgery with supracoronary graft interposition can be another alternative. The results of valve-sparing operations for BAV were comparable to those for tricuspid valves (9). Nevertheless, function of native aortic valve affects decision upon preservation of either bicuspid or tricuspid valves.

Conclusion

Acute type-A dissection is an unusual event in childhood. Clinical suspect in patients with chest pain and hypertension establishes early diagnosis. Regular follow-up of pediatrics with BAV and aortic dilatation is recommended.

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Video 1. Transesophageal echocardiography examination shows dissection of the ascending aorta

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Available Online Date/Çevrimiçi Yayın Tarihi: 18.09.2012

doi:10.5152/akd.2012.226