Cardiology in the Young

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Large saccular aneurysm of the ascending aorta in a 5-year-old patient

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Brief Report

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Abstract

Saccular aneurysms of the aorta in childhood are rare, and the low incidence of aortic aneurysms among children limits our understanding of their aetiology and surgical indications. In this case report, we describe the successful surgical treatment of a 5-year-old boy with severe aortic valvular stenosis, supra-valvular aortic stenosis, and a large saccular aneurysm in the anterior wall of the ascending aorta, without any connective tissue disorder.

Aneurysms of the aorta in children have been reported in association with various conditions such as aortic valvular stenosis, bicuspid aortic valves, coarctation of the aorta, abnormality of sinus Valsalva, or congenital connective tissue disorders (such as Marfan, Ehlers-Danlos, and Turner syndromes).[1,2] However, saccular aneurysms of the aorta in childhood are rare, and the low incidence of aortic aneurysms among children limits our understanding of their aetiology and surgical indications. In this case report, we described the successful surgical treatment of a 5-year-old boy with severe aortic valvular stenosis, supra-valvular aortic stenosis, and a large saccular aneurysm in the anterior wall of the ascending aorta, without any connective tissue disorder.

Case report

A 5-year-old boy was admitted to our clinic with a diagnosis of severe aortic valvular stenosis, a large saccular aneurysm in the anterior wall of the ascending aorta, and supra-valvular aortic stenosis. Physical examination revealed high blood pressure (140/60 mmHg) and a systolic murmur (grade 3/6) on the upper right side of the sternum. The rest of the examination did not reveal any abnormalities. Transthoracic echocardiography and catheter angiography confirmed the presence of severe aortic valvular stenosis with a peak systolic gradient across the aortic valve of 135 mmHg, a large saccular aneurysm in the anterior wall of the ascending aorta, and supra-valvular aortic stenosis (Fig. 1). Propranolol treatment was initiated after the transthoracic echocardiography examination to prevent the progression of left ventricular hypertrophy caused by advanced aortic stenosis. CT angiography showed that the diameter of the aneurysm was 28 x 22 mm (Fig. 2). The diameter of the ascending aorta was 16 mm, while at the level of the junction of the ascending aorta and arcus aorta, it was 21 mm, and the diameter of the descending aorta was 10 mm. We also checked the patient for cerebral aneurysms. The cranial CT angiography showed that there was no cerebral aneurysm. The surgical plan included resection of the aneurysm, aortic valvotomy, and single patch repair of the supra-valvular aortic stenosis. The patient provided written consent, and preparations for the operation were made. Following a median sternotomy, subtotal pericardiectomy was performed due to pericardial adhesions of unknown aetiology. Cardiopulmonary bypass was established via the innominate artery for arterial cannulation, and the superior and inferior caval vein were cannulated in a standard fashion for venous cannulation. A 6 mm polytetrafluoroethylene vascular graft was anastomosed end-to-side to the innominate artery to provide an additional site for placing an aortic cross-clamp using a 7.0 polypropylene suture. A 16 Fr. aortic cannula was inserted into the vascular graft for distal perfusion under moderate hypothermia (28°C). An aortic cross-clamp was applied, and antegrade cold blood cardioplegia (8°C) was administered and repeated every 20 minutes. The saccular aneurysm was resected, and a longitudinal incision was made on the anterior surface of the ascending aorta from the aneurysm into the non-coronary sinus. Aortic valvotomy was performed, and the bovine pericardium was used as a single patch for repairing the supra-valvular aortic stenosis and aneurysm. The cross-clamp and cardiopulmonary bypass times were 86 and 135 minutes, respectively. Pathological examination of the aneurysm specimen revealed an increase in soft connective tissue in the intimal layer, fibrosis of the media, and congestion and chronic inflammation in the adventitia. The specimens were stained using periodic acid-Schiff, Elastica van Gieson, and trichrome stains. No accumulation of material was observed in

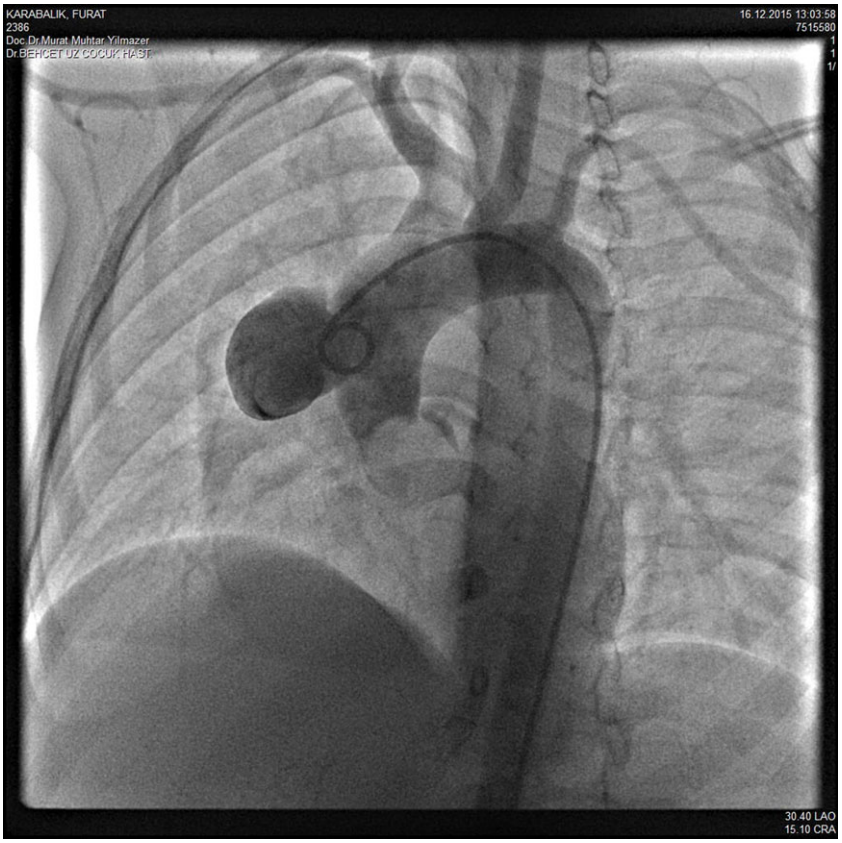

Figure 1. Angiography image of the aneurysm.

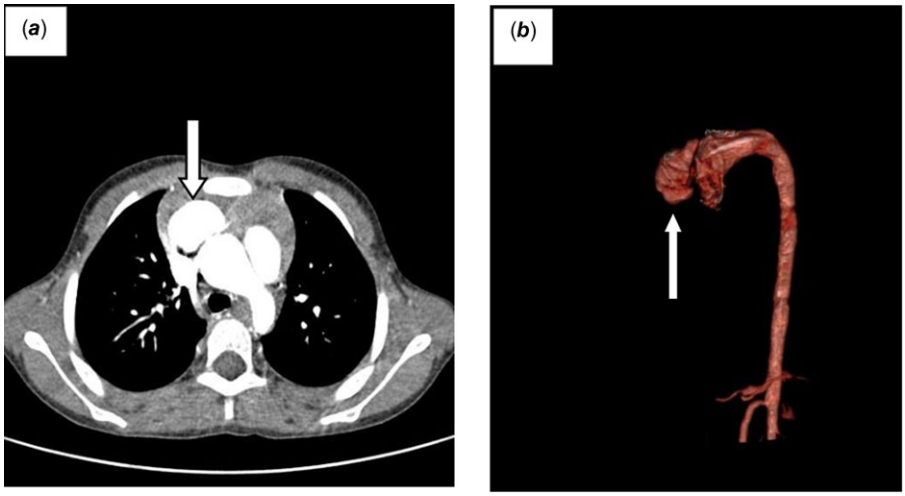

Figure 2. (***a***,***b***) Pre-operative CT angiography image of the aorta.

the periodic acid-Schiff stain. The elastic fibre arrangement appeared to have disappeared in the Elastica van Gieson stain. An increase in myointimal fibrosis was observed in the trichrome stain. The microbiological culture of the aneurysm tissue yielded negative results. The patient was extubated in the third hour of the post-operative period and was discharged from the hospital on the fifth post-operative day. Post-operative control echocardiography revealed a peak systolic gradient across the aortic valve of 20 mmHg and grade 1 aortic regurgitation. Post-operative CT angiography showed no remaining aneurysm of the aorta (Fig. 3). The long-term follow-up was uneventful.

Discussion

The association between the aortic aneurysm and aortic valvular stenosis was first reported in adults by Frederick et al.[1] Hypertension, infection (mycotic aneurysm or aortitis), and underlying abnormalities of the aortic wall (such as cystic medionecrosis) contribute to the pathogenesis of aortic aneurysms in both children and adult patients.[3,4] Coarctation of the aorta and aortic valvular stenosis have been reported as the most common congenital cardiac abnormalities associated with aortic aneurysms. In our patient, the presence of a large saccular aneurysm in the anterior wall of the ascending aorta, without any

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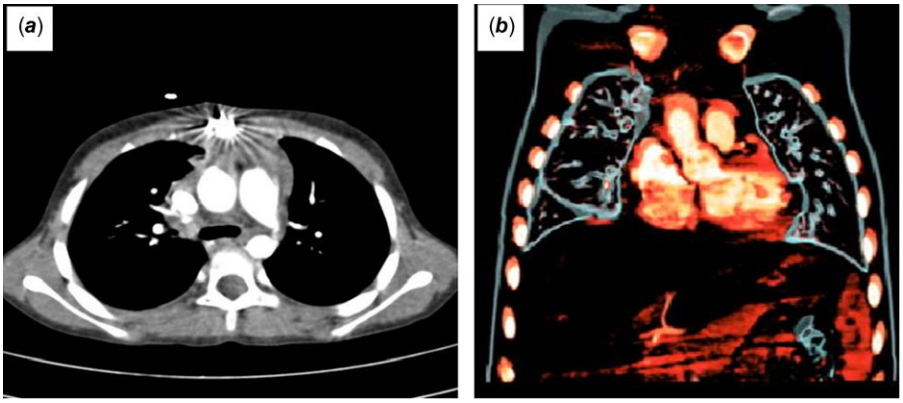

Figure 3. (*a*,*b*) Post-operative CT angiography.

connective tissue disorder, required urgent treatment due to the risk of rupture. However, the association between pericardial thickening and fibrosis is not easily explained. Perhaps, this fibrosis is caused by the rupture of the aneurysm. The association of clinically significant abnormalities of the ascending aorta, including aortic dilatation, aneurysm, dissection, and coarctation of the aorta, suggests a common underlying developmental defect involving the aortic valve and the wall of the ascending aorta.[5] The common neuroectodermal origin of both structures is important, as experimental neural crest ablation has demonstrated its impact on cardiac outflow tract formation.[6,7] Despite ascending aortic aneurysms in children typically being associated with underlying congenital abnormalities such as aortic valvular stenosis, coarctation of the aorta, and Marfan syndrome, saccular aneurysms with unknown aetiology, like the one in our case, are rarely observed. Traumatic aortic aneurysms have been almost exclusively reported in adults,[1] and there was no history of chest trauma in our patient.

In this case, the early diagnosis of the rarely seen aneurysm, without any association with connective tissue defect syndromes, allowed for a favourable post-operative outcome. The treatment of a large saccular aneurysm in the anterior wall of the ascending aorta was successful.

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Competing interests. The author declares that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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