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Case Report

Congenital Saccular Aneurysm of Coarctation of Aorta: A Case Report

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Abstract

True saccular aneurysm is exceptional in coarctation of aorta in children. A 6-year-old girl with headache and systemic arterial hypertension referred to our center for cardiovascular evaluation. Physical examination revealed high blood pressure and weak lower extremity pulses. Two-dimensional and Doppler echocardiography and angiography demonstrated a saccular aneurysm of the descending aorta, 5×4 cm in size, associated with aortic coarctation. Aneurysmectomy was performed without cardiopulmonary bypass. The section of the aorta containing the coarctation and the aneurysm was resected and replaced with a 15-mm woven polyester graft. Histological examination revealed a thin walled aneurysm with hyalinosis changes and decreased elastic fibers. One-year echocardiographic follow-up revealed no gradient across of Dacron tube and hypertension decreased to normal level.

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Introduction

True saccular coarctation site aneurysms are extremely rare in children. Other non-saccular types are also very rare and are in consequence of infection, Marfan's syndrome, hypertension, and trauma. Coarctation of the aorta is found in approximately 7% of patients with congenital heart disease.[1] If untreated, approximately 90% will die by its sequel by the age of 50 years. Coarctation of the aorta is a common congenital anomaly and it may be associated with serious and rare anomalies such as aneurysms or other non-serious anomalies such as bicuspid aorta valves, double aortic arcs, left superior vena cavae, pulmonary venous return anomalies, ventricular septal defects, and disturbances of cardiac rhythms.[2, 3]

Aortic coarctation may cause systemic hypertension, coronary artery disease, myocardial infarction, congestive heart failure, infective endocarditis, intracranial berry aneurysms, subarachnoid hemorrhage, aneurysms, and dissection of the aorta. There are two types of aneurysms in coarctation: childhood type and adult type. The adult type is well known and is accompanied by the atherosclerosis of the aortic wall or other histochemical changes of the media. The childhood type is divided into two subtypes: subtype one (as was the case in our patient) occurs in the coarctation site and is extremely rare; (Our meticulous literature search yielded only two case reports on this subtype.) and subtype 2 occurs in the aortic wall in the pre- or post-coarctation site

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and, albeit very rare in children, is not rare in adults.

The most common cause of death in coarctation is aneurysm and rupture of the aorta and its branches. Resection of the aneurysmatic segment of the aorta is necessary to prevent the rupture of the aneurysm.[4]

Case Report

A 6-year-old girl was referred with headache and systemic arterial hypertension to our center. Physical examination revealed high blood pressure (160/10 mm Hg) and weak lower extremity pulses. Two-dimensional and Doppler echocardiography and angiography demonstrated a saccular aneurysm of the descending aorta, 5×4 cm in size, associated with aortic coarctation, with typical continuous wave Doppler display across the coarctation. The patient underwent left heart catheterization, which revealed a pressure gradient of 85 mm Hg across the coarcted segment. Lateral thoracotomy demonstrated a saccular aneurysm in the narrowing site. (Figure 1) She underwent surgical resection of the saccular aneurysm and replacement of the resected aortic segment (Figure 2) with a woven Dacron tube graft interposition. Postoperative period became eventful by atrial fibrillation, which was reverted to sinus rhythm with respiratory care and physiotherapy. Echocardiography, thereafter, showed a normal flow pattern. To our knowledge, this is the third reported case of aneurismal formation at a coarctation site in medical literature.

Discussion

In coarctation of the aorta, an aneurysm can occur in the ductal, pre-ductal, post-ductal, and intercostal arteries as well as in the narrowing site of the aortic coarctation. An aneurysm associated with coarctation is well known in adults and could remain asymptomatic for a prolonged time, whereas a saccular aneurysm of the coarctation site is exceedingly rare, and it may be detected in childhood during investigations for weakness, headache, or hypertension, or incidentally by chest radiography, or from symptoms caused by lower extremity pulselessness, as was the case in our patient.

In the Bobby et al. study,[5] the incidence of aneurysms in pre- and post-coarctation sites was 15.7%, all of the patients were adults, and none of the cases occurred in the narrowing site of the coarctation. In the Cohen series,[6] the incidence of pre-coarctation aneurysms was 33% and 73% of the cases occurred in other sites such as distal, ductal, and collateral intercostal arteries.

The cause of aneurysm formation in association with coarctation is attributed to cystic medial necrosis and mucopolysaccharide change of the media. Be that as it may, hypertension and endocarditis (by eddy currents in low-flow areas) might play a role in aneurysm formation. In adulthood, atherosclerosis, dissection, and infection of the aortic wall; and in childhood, cystic media necrosis of the aortic wall have been previously seen in pathological examinations of aneurysms. A definitive diagnosis, however, needs histopathological and immunohistochemical studies of a specimen from the aneurysmal wall.

Non-specific inflammation can occur in the aortic wall in the presence of atherosclerosis. Mitchel et al.[7] found that an inflammatory process might cause progressive destruction of the lamina elastica of the media and that the destruction of the media plays a particularly significant role in the aneurismal formation of the aortic wall. Clarkson et al.[8] reported an asymptomatic boy with coarctation of the thoracic aorta. Although the patient had no history of endocarditis or trauma, the authors identified a large saccular aneurysm involving the coarctation site on angiography, which was subsequently

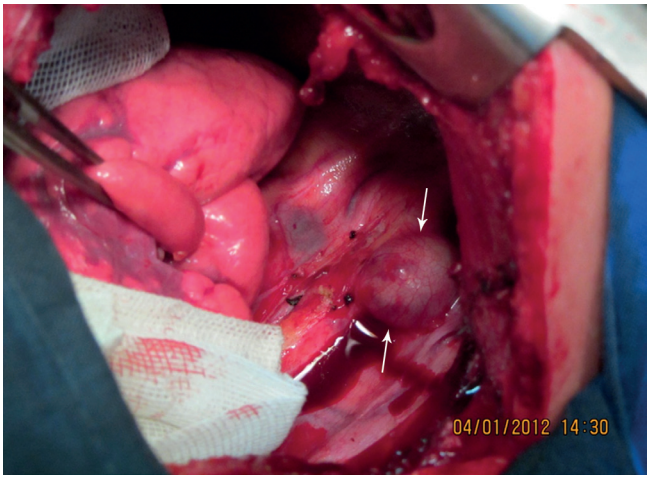

Figure 1. Left lateral thoracotomy in the 5th intercostal space. White arrows show the upper and lower margins of the aneurysm in the descending aorta

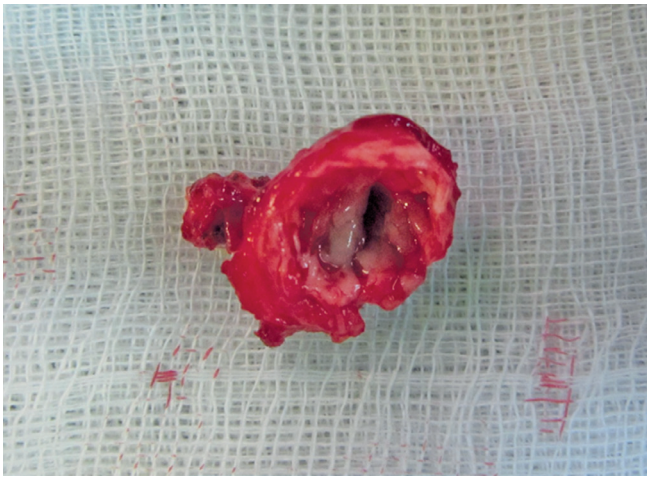

Figure 2. Gross anatomy of the resected aneurysm

resected surgically. Okabayashi[9] reported an 8-year-old girl with coarctation of the aorta associated with aortic aneurysm formation. She underwent surgery under cardiopulmonary bypass, during which a 7-cm section of the aorta containing the aneurysm and coarctation was resected and replaced with a 16-mm woven Dacron tube. Histologic sections of the resected portions of the thoracic aorta revealed intimal hyperplasia, medial fibrosis, and reduced elastic fibers, especially in the aneurismal wall. Hiller et al.[10] found that true aneurysms of the aorta accompanied by coarctation were rare in comparison with other aneurysms.

Pathogenesis for true aneurysms includes atherosclerosis, post-stenotic dilatation, infective endocarditis, and connective tissue disorder. Roy et al.[11] described the case of a infantile mycotic thoracic aneurysm secondary to a highly placed infected umbilical catheter. The rapid expansion of the infection and unsuccessful antibiotic therapy prompted the authors to replace the thoracic aorta with a woven graft, using cardiopulmonary bypass and hypothermic arrest. The patient had an uneventful recovery and was discharged home after a prolonged antibiotic course. Stewart et al.[12] reported two cases of post-coarctation descending thoracic aneurysms in 2 teenagers; the patients were treated via aneurysmectomy but one of them expired due to surgical complications. Ogawa et al.[13] described tuberculous pseudoaneurysms of the aorta as rare mycotic aneurysms with a fatal outcome if not treated properly. The authors reported a case of a recurrent tuberculous of the vertebra and a false aneurysm of the descending thoracic aorta that was treated surgically with excision and primary repair of the lesion.

Pre- or postoperative atrial fibrillation is not an uncommon rhythm in congenital heart disease and as Sabzi et al.[3] stated, its occurrence in the postoperative period relates to multiple factors such as atelectasis and hypoxia (as was the case in our patient). Atrial fibrillation can be converted to sinus rhythm through the correction of respiratory causes.

Conclusion

To the best of our knowledge, this is the third reported case of a saccular aneurysm after a successful surgical treatment. In contrast with our case, descending aortic aneurysms in children are usually associated with connective tissue defect syndromes such as Marfan's. Indications for surgery as well as surgical options differ according to the diagnosis and type of coarctation.

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