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Saccular aneurysm formation of the descending aorta associated with aortic coarctation in an infant

Formação de aneurismas saculares da aorta descendente associados com coarctação aórtica em criança

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Abstract

Aneurysm of the descending aorta associated with CoA is an extremely rare congenital abnormality. In this report, we present a 16 months old female patient in whom cardiac catheterization had been performed which had revealed a segment of coarctation and saccular aneurysm in the descending aorta. The patient was operated and a 3x2 centimeters aneurysm which embraces the coarcted segment in descending aorta was resected. In summary, we present a case of saccular aortic aneurysm distal to aortic coarctation in an infant without any history of intervention or vascular inflammatory disease. Our case report seems to be the youngest patient in literature with this pathology.

Descriptors: **Aortic Aneurysm, Thoracic. Aortic Coarctation. Infant.**

Resumo

Aneurisma da aorta descendente associada à coarctação aórtica é uma anomalia congênita extremamente rara. Neste relato, apresentamos uma paciente de 16 meses de idade, nos quais o cateterismo cardíaco foi realizado, que havia revelado um segmento de coarctação e aneurisma sacular na aorta descendente. A paciente foi operada e um aneurisma de 3x2 centímetros, englobando o segmento coarctada na aorta descendente foi ressecado. Em resumo, apresentamos um caso de aneurisma sacular distal à coarctação da aorta em uma criança sem histórico de intervenção ou doença inflamatória vascular. Nosso caso parece ser o do paciente mais jovem na literatura com esta afecção.

Descritores: **Aneurisma da Aorta Torácica. Coarctação Aórtica. Lactente**

INTRODUCTION

Coarctation of the aorta (CoA) is defined as the hemodynamically significant narrowing of the descending thoracic aorta, usually just distal to the left subclavian artery where the ligamentum arteriosum originates[1]. CoA accounts for 6.5% of all congenital heart defects[2]. CoA is usually classified into three categories: I.Isolated CoA, II. CoA with ventricular septal defect (VSD) and III. CoA with complex cardiac anomalies[3]. Aneurysm of the descending aorta associated with CoA is an extremely rare congenital abnormality. In this report, we present the youngest patient per our literature search with CoA and saccular descending aortic aneurysm.

CASE PRESENTATION

A 16 months old female patient was admitted to our clinic with diagnosis of aortic coarctation and descending aortic aneurysm. She presented with poor feeding and failure to thrive (body weight: 8 kg). The prenatal and postnatal history was unremarkable; any umbilical vascular catheterization was not performed. Physical examination revealed

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Abbreviations, acronyms & symbols	
CoA	Coarctation of the aorta
VSD	Ventricular septal defect

80 mmHg systolic pressure difference between upper and lower extremities. Femoral pulses were weak but palpable. There was a systolic murmur (3/6) at the left sternal border which was also heard at the interscapular area. The leukocyte count, C-reactive protein level and erythrocyte sedimentation rate were all within normal limits. The patient had been admitted to another clinic a few months ago and transthoracic echocardiography had demonstrated aortic coarctation with 60mmHg peak systolic gradient. Any intracardiac pathology had not been encountered. Cardiac catheterization had been performed in order to confirm the diagnosis and perform a balloon angioplasty if possible; however the catheterization had revealed a segment of coarctation and saccular aneurysm in the descending aorta (Figure 1). Therefore, the patient was referred to our clinic for surgical repair.

The patient was operated under general anesthesia and a left posterolateral thoracotomy was performed at the 4th intercostal space. A 3x2 cm aneurysm which embraces the coarcted segment in descending aorta was encountered (Figure 1). Descending aorta was mobilized, resection and end-to-end anastomosis was performed with 6/0 polyprolene suture. The sutures were continuous at the posterior wall, whereas they were interrupted at the anterior wall of the anastomosis, in order to provide potential for growth. Residual gradient was less than 5 mmHg. The resected specimen was examined by the pathology department. Gross and microscopic cross sectional examination of the coarctation area revealed the segments of both coarctation and aneurysm formation. Basophilic degeneration and vacuole formation were remarkable (Figure 2). The postoperative period was uneventful and the patient was discharged at the sixth postoperative day without any complication. At the sixth postoperative month, a second intervention was performed because of recoarctation. The MR angiography revealed a recoarctation at the descending aorta, in which balloon dilatation was performed with a residual gradient of 25 mmHg (Figure 3). The patient is followed up by echocardiographic evaluations with regular intervals.

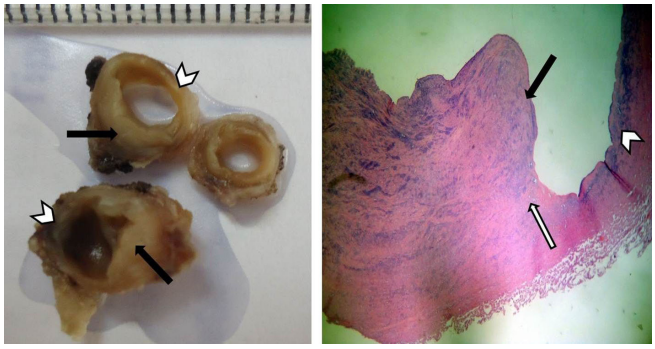

Fig. 2 - Gross cross sectional examination of the area of coarctation and aneurysm formation (left side) (black arrows indicate the area of coarctation and white arrow heads indicate the area of aneurysm formation). Hematoxylin eosin stained section revealing the zone of transition between the area of coarctation and aneurysm (right side) (x40 magnification) (black arrow indicates the area of coarctation and white arrow head indicates the area of aneurysm formation. Basophilic degeneration and vacuole formation were remarkable indicated with white arrow).

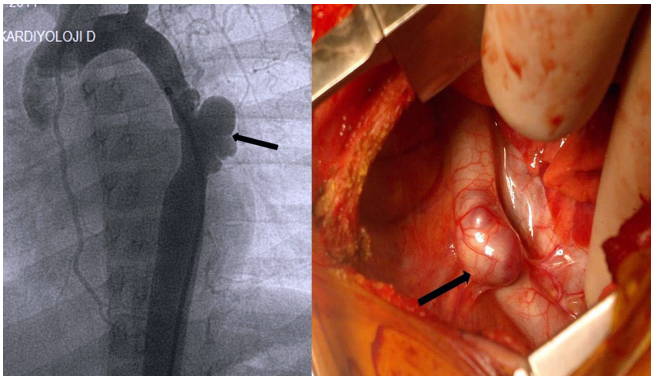

Fig. 1 - Coarctation and saccular aneurysm formation in the descending aorta in cardiac catheterization (left side - arrow). Saccular aneurysm embracing the segment of coarctation in the descending aorta, operative view (right side - arrow).

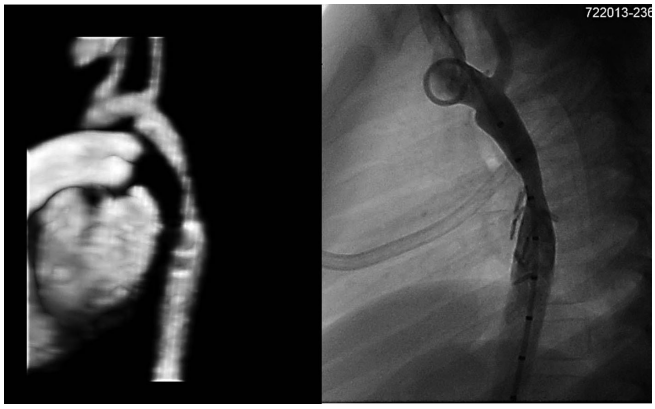

Fig. 3 - MR angiography revealing the recoarctation at the descending aorta (left) which was dilated with percutaneous intervention (right).

DISCUSSION

Surgical repair of CoA mostly depends on the accompanying cardiac anomalies in the patient. The main treatment target is providing a non stenotic aortic continuity with efforts to enhance the growth potential of the native vascular tissues, with or without the repositioning of the left subclavian artery[4]. Although rarely seen, aortic wall pathology such as aneurysm formation, aortic dissection and rupture are mainly the presenting symptoms of adult CoA. When pediatric age group is concerned, the saccular aortic aneurysms distal to coarcted segment are very rare and data about these patients are only confined to limited case reports[5]. Our case report seems to be the youngest patient in literature with this pathology. In such cases, recoarctation may be encountered in the follow up which may be managed by percutaneous balloon dilatation as presented in our case. Extensive mobilization of the aorta and its branches with a meticulous surgical technique is mandatory in order to reduce the tension at the anastomosis site.

In summary, we present a case of saccular aortic aneurysm distal to aortic coarctation in an infant without any history of intervention or vascular inflammatory disease. We speculate that localized weakness of the aortic wall may be responsible for aneurysm formation, since basophilic degeneration and vacuole formation were remarkable at the transition zone between the coarcted and aneurysmatic segments under microscope.

Authors' roles & responsibilities	
AO	Included in surgical team, preparation of the manuscript
EC	Included in surgical team, preparation of the manuscript
AD	Preoperative diagnostic workup of the patient, preparation and final control of the paper
AA	Head of surgical team, preparation and final control of the paper

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