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Case report / Olgu Sunumu

The single-stage extra-anatomic repair of an isolated interrupted aortic arch in an adult and a review of the literature

Yetişkin bir hastada izole kesintiye uğramış arkus aortun tek aşamalı ekstra anatomik baypas ile onarımı ve literatür incelemesi

Murat Özeren, Barlas Aytaçoğlu, Kerem Karaca, Özden Vezir, Nehir Sucu

Department of Cardiovascular Surgery, Medical Faculty of Mersin University, Mersin, Turkey

Interrupted aortic arch is a rare and severe congenital heart defect characterized by a complete loss of luminal and anatomic continuity between the ascending and the descending aorta. This malformation has a poor prognosis without surgical treatment. In this article, we present a 56-year-old male patient with interrupted aortic arch type A who was treated successfully with surgery, and review the literature regarding this topic.

Key words: Adult; congenital; extra-anatomic bypass; interrupted aortic arch.

Kesintiye uğramış arkus aort, çıkan ve inen aort arasında lümenin tamamen kaybı ve anatomik sürekliliğin olmaması ile karakterize, nadir ve ciddi bir doğuştan kalp defektidir. Bu malformasyonun prognozu, cerrahi olarak tedavi edilmez ise kötüdür. Bu yazıda tip A kesintiye uğramış arkus aortu olan ve cerrahi ile başarılı bir şekilde tedavi edilen 56 yaşında erkek hasta, konu ile ilgili literatür gözden geçirilerek sunuldu.

Anahtar sözcükler: Erişkin; doğuştan; ekstra anatomik baypas; kesintiye uğramış arkus aort.

The interruption of the aortic arch (IAA) characterized by complete luminal and anatomic discontinuity between the ascending and descending aorta is an extremely rare entity in the adult population.[1] It is often diagnosed and repaired during the neonatal period. The prognosis for this anomaly depends on the associated congenital anomalies, but the outcome is usually very poor unless there is surgical treatment. We found 18 adult cases (18 years and older) of IAA which had been surgically treated by extra-anatomic bypass in the literature. Herein we describe a successful extra-anatomic repair in a patient who had an isolated type A aortic interruption.

CASE REPORT

A 56-year-old male patient arrived at the cardiology clinic of our hospital complaining of chest pain and shortness of breath. He also had a history of stroke due to an intracranial hemorrhage and hypertension, which had been treated with angiotensin-converting enzyme (ACE) inhibitors. On his physical examination, the peripheral pulses were normally palpable in the upper extremities, but they were reduced in the lower limbs. Cardiac auscultation revealed a rhythmic tachycardia with no additional heart sounds. Electrocardiography showed sinus tachycardia and signs of left ventricular hypertrophy and ST-T wave changes. Chest radiography showed an increased cardiothoracic ratio. Laboratory results were within normal ranges.

An angiography was performed via the femoral approach for coronary artery imaging, but the catheter could not be inserted to the arcus aorta (Figure 1a). Interruption of the thoracic aorta was suspected. The angiography was then repeated via the brachial approach, and the interruption of the arcus aorta was diagnosed with normal coronary arteries (Figure 1b). An angiotomography of the aorta was then requested which disclosed an aortic occlusion 2 cm after the origin of the left subclavian artery (Figure 2a). This

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Correspondence: Murat Özeren, M.D. Mersin Üniversitesi Tıp Fakültesi, Kalp ve Damar Cerrahisi Anabilim Dalı, 33343 Mersin, Turkey.
Tel: +90 324 - 337 43 00 e-mail: mozeren@yahoo.com

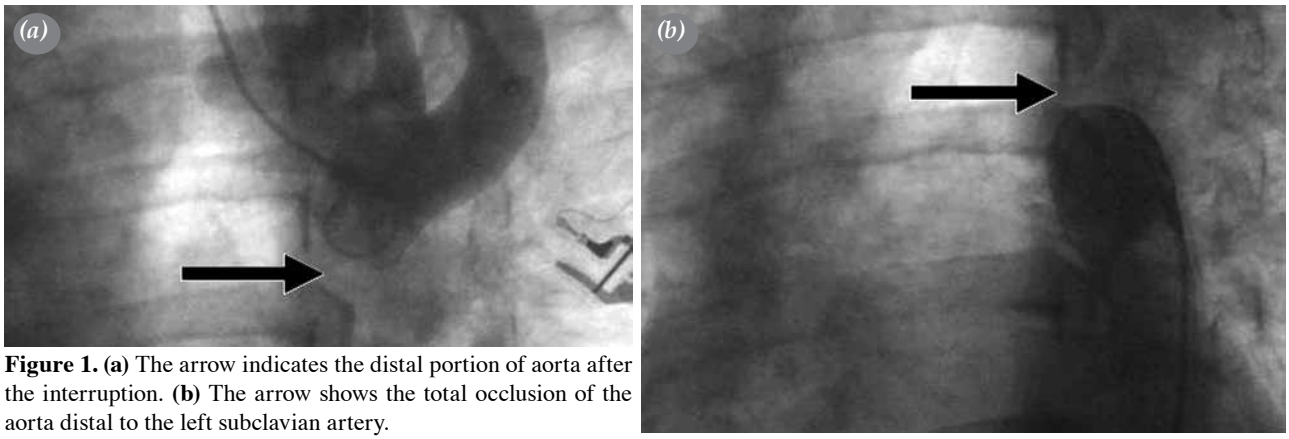

Figure 1. (a) The arrow indicates the distal portion of aorta after the interruption. **(b)** The arrow shows the total occlusion of the aorta distal to the left subclavian artery.

was associated with an extensive collateral network that retrogradely filled the descending aorta and the iliac arteries with normal calibrations (Figure 2b). The clinical and radiological findings revealed the diagnosis of aortic arch interruption type A.

Elective surgery was planned for the patient, and a single-stage operation was performed via a left posterolateral thoracotomy. The left subclavian artery, arcus aorta, and descending aorta were exposed. An extra-anatomic bypass was performed between the arcus and descending aorta using a 22 mm Dacron tubular graft (Vaskutek®, Gelsoft TM Terumo Cardiovascular Systems, Inchinnan, Renfrewshire, Scotland) (Figure 3). The proximal and distal anastomoses were done with running 4-0 polypropylene sutures using an end-to-side technique. Bleeding from the anastomotic sites was controlled with tissue glue and Surgicel® (Ethicon, Inc., a Johnson & Johnson company, Somerville, New Jersey, USA). The early postoperative period was uneventful until postoperative day three when rebound hypertension and arterial mesenteritis ensued. The hypertension was controlled with oral administration of 50 mg of metoprolol and 60 mg of nifedipine. The arterial mesenteritis was followed closely, and it regressed in conjunction with the reduction of the blood pressure in three days time. The patient was discharged from the hospital without any complications on postoperative day 14.

DISCUSSION

Interruption of the aortic arch is an extremely rare congenital malformation that occurs in 3/1.000.000

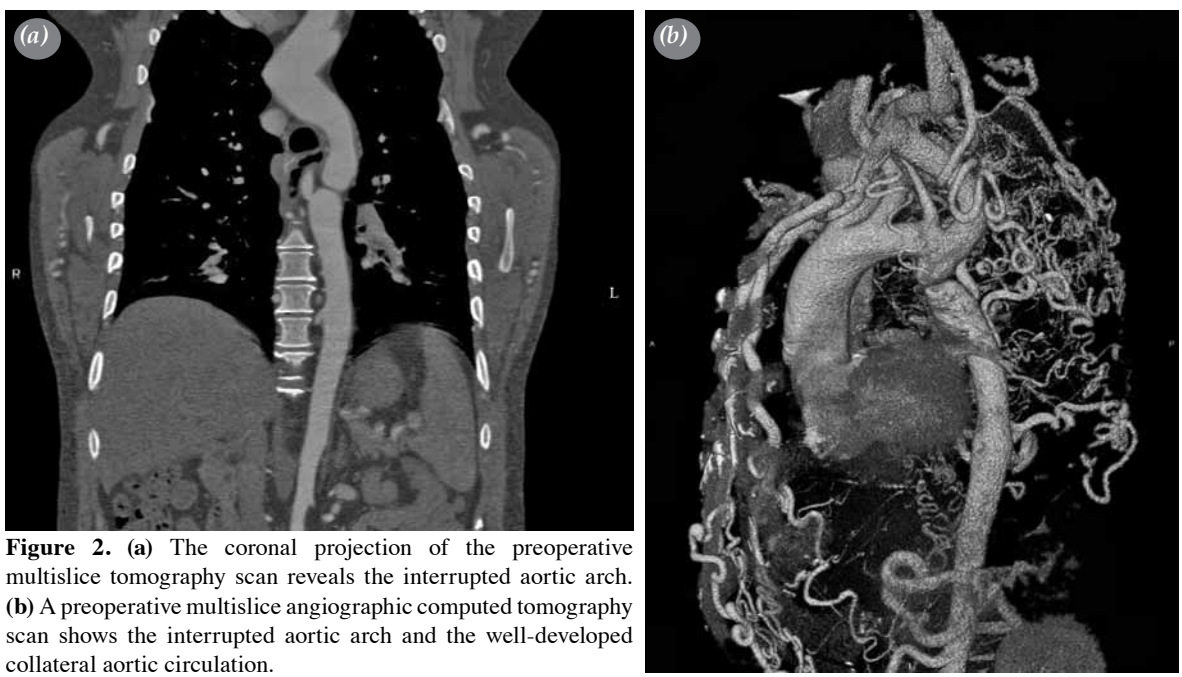

Figure 2. (a) The coronal projection of the preoperative multislice tomography scan reveals the interrupted aortic arch. **(b)** A preoperative multislice angiographic computed tomography scan shows the interrupted aortic arch and the well-developed collateral aortic circulation.

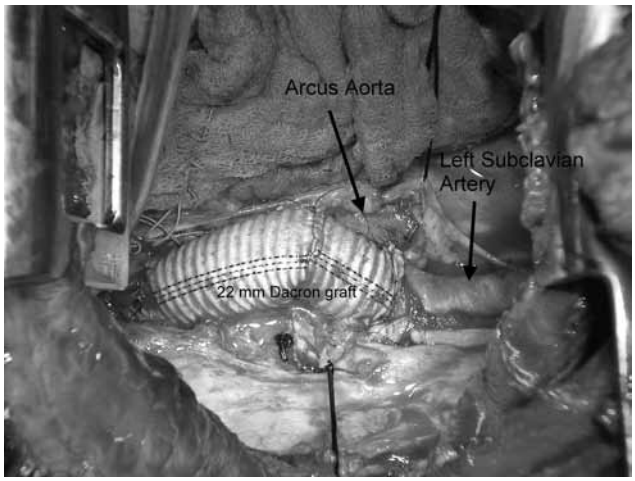

Figure 3. An intraoperative view via a left thoracotomy approach shows the 22 mm Dacron graft conduit anastomosed between the arcus aorta and descending aorta using an end-to-side technique.

live births and accounts for 1% of all congenital heart disease.[1] It was described for the first time in 1778 by Steidele,[2] and Celoria and Patton[3] defined the first classification system of IAA in 1959, which is still popular today. According to this classification system, the interruption is identifieded as type A when the site of the aortic arch discontinuity is distal to the left subclavian artery (as in our case), as type B when the interruption site is between the left subclavian artery and the left carotid artery, and as type C when this segment is between the left carotid artery and the innominate artery. Type B is the most common (53%), followed by type A (43%) and type C (4%).

Interruption of the aortic arch clinically presents with severe congestive heart failure in the neonatal period, and 90% of the affected neonates die at a median age of four days[4] if the condition is left untreated. The anomaly is extremely rare in adults, and the presentation varies from asymptomatic status to differential blood pressure recordings in the extremities and systemic arterial hypertension with its attendant complications. Stroke due to cranial hemorrhage was the first serious symptom in the past history of our patient. He only had severe hypertension and weak femoral pulses; however, he had no claudicating related to reduced peripheral perfusion. Survival into adulthood is dependent upon the development of substantial collateral circulation, as happened with our patient. These collateral vessels are subject to atrophy, atherosclerosis, and even spontaneous rupture, resulting in secondary complications,[5] but these conditions were not found in our case.

Several methods can be used for the diagnosis of IAA. Although it has some limitations, echocardiography is the procedure of choice for the initial diagnosis of IAA in almost all cases.[6] Cardiac catheterization is the most widely used technique for the definitive diagnosis; however, it may be difficult to perform this in patients without a prior knowledge of their vascular anatomy to ensure visualization of both the proximal and distal segments.[7] In the presented case, aortography via brachial artery demonstrated the IAA, the site of interruption, and the branching pattern of the great arteries. Angiographic reconstructions of the images obtained with a 64-slice multidetector computed

Table 1. Adult patients with surgical repair of isolated interrupted aortic arch

No	Reference	Age/gender	Interruption type	Size of extraanatomic Dacron tube
1	Morgan et al.[11]	19/M	B	10 mm
2	Kauff MK et al.[12]	36/M	B	Size not stated;
3	Kauff MK et al.[12]	49/M	A	Size not stated;
4	Le Page et al.[13]	56/M	A	Size not stated;
5	Milo S et al.[14]	65/M	B	10 mm
6	Todoric M et al.[15]	19/M	A	10 mm
7	Canova CR et al.[16]	72/F	A	16 mm
8	Burton BJ et al.[17]	32/M	A	18 mm
9	Ogino H et al.[18]	32/M	A	16 mm
10	Messner G et al.[19]	42/F	B	7 mm and 16 mm
11	Riess FC et al.[20]	64/M	A	16 mm
12	Krishna SC et al.[10]	40/M	B	18 mm
13	Krishna SC et al.[10]	46/F	A	16 mm
14	Krishna SC et al.[10]	34/M	B	20 mm
15	Krishna SC et al.[10]	40/M	A	18 mm
16	Krishna SC et al.[10]	23/M	A	20 mm
17	Celkan MA et al.[21]	35/M	A	Size not stated
18	Casati V et al.[22]	35/M	A	16 mm

tomography unit are a reliable noninvasive diagnostic modality for the precise diagnosis of aortic coarctation and aortic arch anomalies. In addition, as in our case, they are useful for the visualization of the collateral vessels.[8]

The surgical reparation of IAA cases in a single- or multi-staged setting had been a matter of debate until recently; however, a single-stage repair is now considered to be the best method of treatment in infants.[9] Sai Krishna et al.[10] published their three-year surgical experience in treating IAA in seven adult cases. They used a single-stage extra-anatomic bypass in nearly all of their cases through a midline sternotomy. There were only two exceptions in which other procedures were performed. Most adult patients with IAA have been repaired by using a single-stage extra-anatomic approach (Table 1).[10-22] For our case, the correction was done in a single setting by using an extra-anatomic bypass with a tubular Dacron graft. Different sizes (10-20 mm) of extra-anatomic Dacron tubular grafts have been used in the literature for this procedure, but we preferred a 22 mm Dacron graft, the largest graft we saw in the literature, in an effort to obtain a better distal blood flow. However, this increased flow could have been the reason for the mesenteric arterial inflammation in our case.

In conclusion, isolated IAA is a rarely encountered, congenital aortic anomaly in adult patients. Surgeons have a tendency to treat these patients via a single-stage extra-anatomic repair in most cases. This method carries lower morbidity and mortality risks. Unfortunately, there is insufficient data currently available to compare one-stage versus two-stage repairs.

Declaration of conflicting interests

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