Herz 2015 · 40:549–551
DOI 10.1007/s00059-013-4023-8
Received: 3 October 2013
Accepted: 2 November 2013
Published online: 18 December 2013

I. Rencuzogullari[1] · I.T. Ozcan[2] · A. Cirit[2] · S. Ayhan[2]
[1] Division of Cardiology, Mugla Sitki Kocman University Education and Research Hospital, Mugla
[2] Division of Cardiology, Department of Medicine, Mersin University School of Medicine, Mersin

Isolated interrupted aortic arch in adulthood

A case report

Case report

A 56-year-old man presented to our hospital with the complaints of dyspnea and chest pain during exercise over 6 months. He had a history of hypertension for several years, which had been kept under control with nifedipine plus valsartan. In addition, he had a hemorrhagic stroke 2 years earlier but he had no physical limitations. However, he had not undergone a complete evaluation at that time. His brother died at 41 years of age of myocardial infarction.

On physical examination, peripheral pulses were palpable over the carotid arteries and in the upper limbs, with a radial pulse stronger in the right arm than in the left arm. Bounding pulses in the neck were also detected. The blood pressure was 135/70 mmHg in the right arm and 95/60 mmHg in the left arm. Lower limb pulses were not palpable. There was 2/6 systolic ejection murmur on the second intercostal area at the left. A chest x-ray demonstrated cardiomegaly and increased vascularity. Transthoracic echocardiography from parasternal and apical views showed concentric hypertrophy and moderate aortic regurgitation, but a satisfactory image could not be obtained from the suprasternal view because of poor image quality.

During cardiac catheterization, the guidewire did not pass beyond the proximal part of the descending aorta. Aortography via the femoral artery showed a complete interruption of the aortic arch just distal to the origin of the left subclavian artery (**Fig. 1a**). We inserted a pigtail catheter distal to the subclavian artery via the right brachial artery and we could

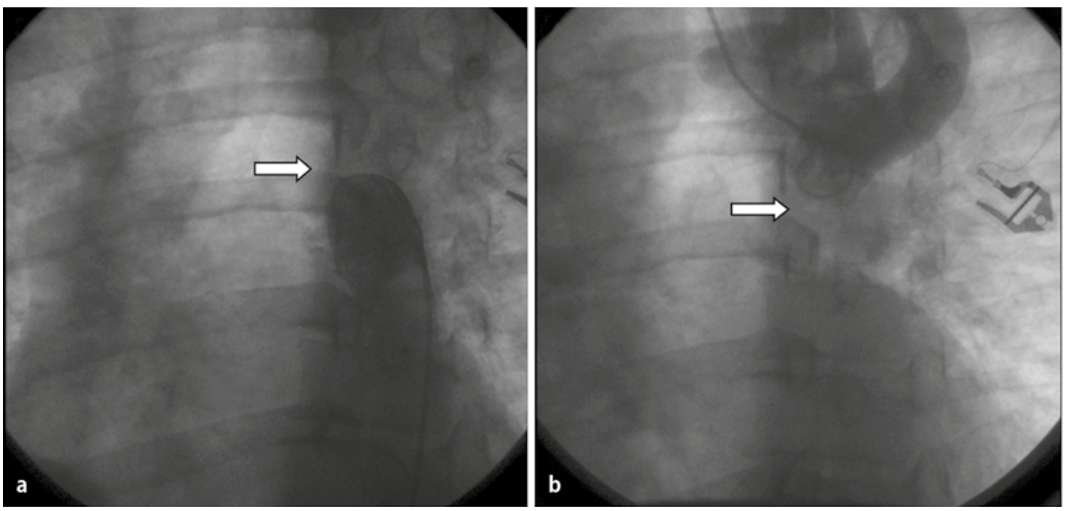

Fig. 1 ◀ Aortography showing the distal portion of the interruption (**a,** *arrow*) and the total occlusion of aorta distal to the left subclavian artery (**b,** *arrow*)

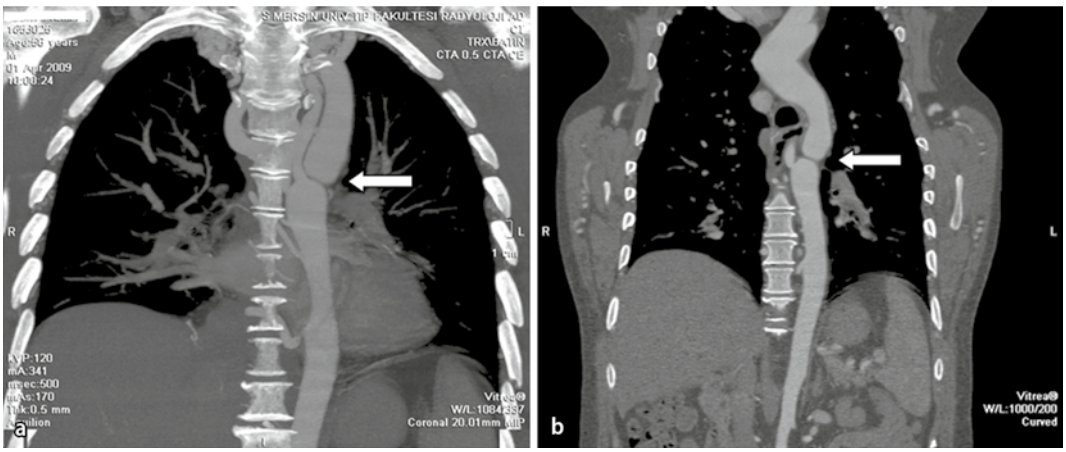

Fig. 2 ◀ **a, b** Contrast-enhanced, three-dimensional computed tomography angiogram reveals severe hypoplasia of the aortic arch (*arrow*) distal to the origin of the left subclavian artery (interrupted aortic arch, type A)

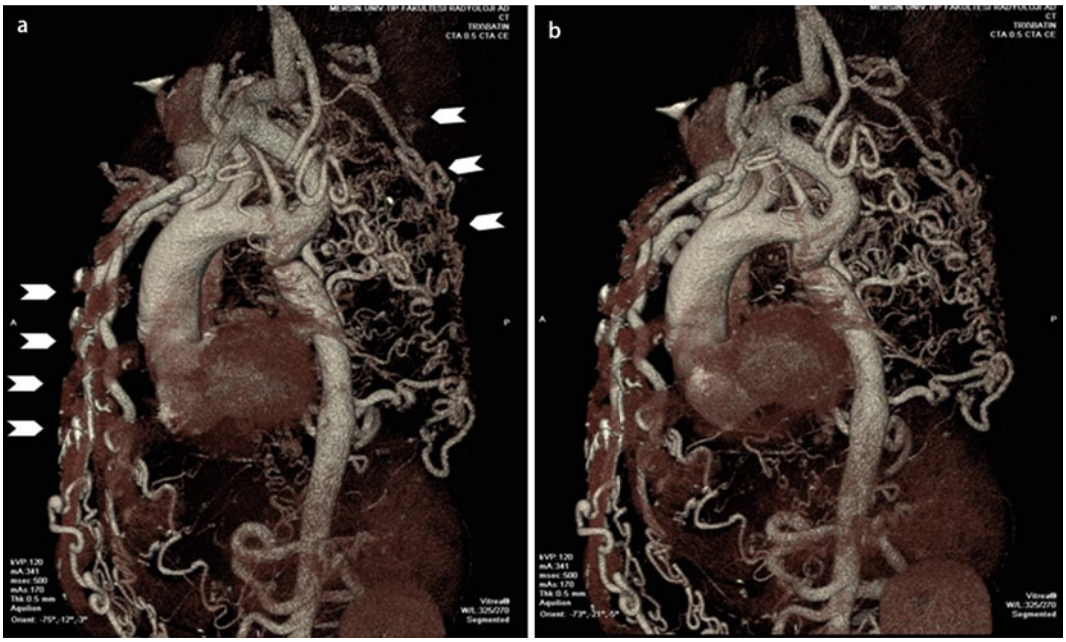

Fig. 3 ◀ **a, b** Contrast-enhanced, three-dimensional computed tomography angiogram. Extensive collateral vessels are evident in the paraspinal region and involve the vertebrobasilar system (*arrows*)

clearly demonstrate the interruption and collaterals (◘ **Fig. 1b**). Computed tomography angiography revealed severe hypoplasia of the transverse aortic arch proximal to the origin of the left subclavian artery (◘ **Fig. 2**). The bilateral common carotid arteries were dilated. The descending thoracic aorta was supplied by extensive collateral vessels from the vertebrobasilar system down to the posterior chest wall and the spine (◘ **Fig. 3**).

In accordance with the results of the cardiac catheterization, retrograde flow could be seen in the proximal left subclavian artery and the left vertebral artery. A single-stage extra-anatomic repair was made by placing a 16-mm Dacron graft between the ascending and descending portions of the thoracic aorta. The patient recovered uneventfully and was doing well at the first follow-up visit 1 month later.

Discussion

Interrupted aortic arch (IAA) is a rare congenital malformation that occurs in three cases per million live births [1]. It is defined as the absence of luminal continuity between the ascending and descending portions of the aorta [2]. In most cases, IAA is associated with intracardiac malformations such as ventricular septal defect, patent ductus arteriosus, bicuspid aortic valve, left ventricular outflow tract obstruction, or aortopulmonary window. In this anomaly, the prognosis, which depends on the associated congenital anomalies, is very poor unless there is surgical treatment [1, 2]. In infants, its clinical presentation involves severe congestive heart failure and 90% of affected infants die at a median age of 4 days if not treated [3]. In a few documented cases in adults, the presentation ranges from a lack of symptoms to limb swelling with blood pressure difference between the arms and legs. Substantial collateral circulation must be present to maintain flow, enabling survival. However, collateral vessels are prone to atrophy and atherosclerosis, which can lead to other challenging problems [4].

IAA was first described by Steidele in 1778 [5]. The first classification system, introduced by Celoria and Patton in 1959 [6], is still used today almost uni-

versally. This system describes and classifies the site of aortic arch discontinuity, which may be: distal to the left subclavian artery (type A); between the left carotid and left subclavian arteries (type B); or between the innominate and left carotid arteries (type C). The most common type is B (53%), followed by type A (43%), and C (4%).

Several methods can be used for the diagnosis of IAA. Two-dimensional echocardiography plays an important role in the delineation of IAA. This technique is also necessary for ruling out associated intracardiac anomalies that were not detailed at cardiac catheterization [7]. In general, cardiac catheterization is warranted for definitive anatomical evaluation in patients with IAA. However, it may be difficult to perform without prior knowledge of the vascular anatomy to ensure visualization of both proximal and distal segments [8]. Thoracic three-dimensional magnetic resonance angiography is a reliable, noninvasive diagnostic modality for the correct diagnosis of aortic coarctation and aortic arch anomalies as well as visualization of collateral vessels [9, 10].

Treatment of the IAA is definitely surgical correction [1, 2, 3, 4]. IAA is rarely encountered in an adult patient and the malformation may be repaired in a single-stage procedure by means of an extra-anatomic approach with a low risk of morbidity and mortality [4].

Corresponding address

I. Rencuzogullari
Division of Cardiology,
Mugla Sitki Kocman University Education and Research Hospital
Orhaniye Mahallesi,
İsmet Çatak caddesiMugla
Turkey
rencuzog@gmail.com

Compliance with ethical guidelines

Conflict of interest. I. Rencuzogullari, I.T. Ozcan, A. Cirit, and S. Ayhan state that there are no conflicts of interest.

Consent was obtained from all patients identifiable from images or other information within the manuscript. In the case of underage patients, consent was obtained from a parent or legal guardian.

References

1. Canova CR, Carrel T, Dubach P et al (1995) Interrupted aortic arch: fortuitous diagnosis in a 72-yearold female patient with severe aortic insufficiency [in German]. Schweiz Med Wochenschr 125(1–2):26–30
2. Backer CL, Mavroudis C (2000) Congenital Heart Surgery Nomenclature and Database Project: patent ductus arteriosus, coarctation of the aorta, interrupted aortic arch. Ann Thorac Surg 69(4 Suppl):S298–S307
3. Collins-Nakai RL, Dick M, Parisi-Buckley L et al (1976) Interrupted aortic arch in infancy. J Pediatr 88:959–962
4. Messner G, Reul GJ, Flamm SD et al (2002) Interrupted aortic arch in an adult, single-stage extraanatomic repair. Tex Heart Inst J 29:118–121
5. Steidele RJ (1778) Samml Chir u Med Beob (Vienna) 2:114
6. Celoria GC, Patton RB (1959) Congenital absence of the aortic arch. Am Heart J 58:407–413
7. Collins-Nakai RL, Dick M, Parisi-Buckley L et al (1976) Interrupted aortic arch in infancy. J Pediatr 88:959–962
8. Kosucu P, Kosucu M, Dinc H, Korkmaz L (2006) Interrupted aortic arch in an adult: diagnosis with MSCT. Int J Cardiovasc Imaging 22(5):735–739
9. Varghese A, Gatzoulis M, Mohiaddin RH (2002) Images in cardiovascular medicine: magnetic resonance angiography of a congenitally interrupted aortic arch. Circulation 106(3):E9–E10
10. Yucel EK, Anderson CM, Edelman RR et al (1999) Magnetic resonance angiography: update on applications for extracranial arteries. Circulation 100(22):2284–2301