A case of aortic coarctation mimicking an interrupted aorta

Kesintili aort izlenimi veren aort koarktasyonu

İbrahim Özdoğru, M.D., Özgür Günebakmaz, M.D., Mehmet Güngör Kaya, M.D. Ali Doğan, M.D.

Erciyes University, Faculty of Medicine, Department of Cardiology, Kayseri

An asymptomatic, healthy, 19-year-old male patient was examined for aortic coarctation upon detection of a heart murmur and hypertension on routine physical examination. Transthoracic echocardiography (TTE) showed rupture of the sinus of Valsalva, and bicuspid aortic valve. Findings of aortography and computed tomography (CT) angiography were compatible with an interrupted aorta. For further delineation, transesophageal echocardiography (TEE) was performed and color Doppler imaging showed passage at the site of the descending aorta, which was suggestive of interruption by other imaging methods. The patient underwent surgery for aortic coarctation. At surgery, severe aortic coarctation was noted and corrected. Although TTE is usually adequate for the diagnosis of aortic coarctation, even aortography and CT angiography were misleading in this particular case, and differentiation from interrupted aorta was only possible by TEE.

Key words: Adult; aorta, thoracic/abnormalities; aortic coarctation; echocardiography, transesophageal; heart defects, congenital/diagnosis.

On dokuz yaşında, asemptomatik, sağlıklı erkek hasta, rutin fizik muayenesinde kardiyak üfürüm ve hipertansiyon saptanması üzerine aort koarktasyonu düşünülerek araştırıldı. Transtorasik ekokardiyografide (TTE) Valsalva sinüsü yırtığı ve biküspit aort saptanan hastada, aortografi ve bilgisayarlı tomografi (BT) anjiyografi bulguları kesintili aort ile uyumlu bulundu. Tanıyı kesinleştirmek için transözofageal ekokardiyografi (TÖE) yapıldı ve inen aortta, daha önceki incelemelerde kesintili olarak saptanan bölgede renkli Doppler ile geçiş saptandı. Hasta aort koarktasyonu öntanısıyla ameliyat edildi. Ameliyatta ciddi aort koarktasyonu görülerek darlık bölgesi düzeltildi. Aort koarktasyonu tanısında sıklıkla TTE yeterli olmakla birlikte, sunulan olguda aortografi ve BT anjiyografi dahi tanıda yanıltıcı olmuş, kesintili aort ile aort koarktasyonu ayrımı TÖE ile yapılabilmiştir.

Anahtar sözcükler: Erişkin; aort, torasik/anormallik; aort koarktasyonu; ekokardiyografi, transözofageal; kalp defekti, doğuştan/tanı.

Aortic coarctation was first defined by Morgagni in 1760 as narrowness in a part of the descending aorta.[1] In 98% of the cases, narrowness in aortic coarctation is located immediately distal to the exit point of the left subclavian artery from the aortic arch and just opposite the point of entry of the ductus arteriosus to the aorta. In addition, it may also occur in the transverse aortic arch or the abdominal aorta. 50% of the patients have several cardiac and noncardiac disorders. The most common concomitant pathologies are bicuspid aortic valve (50%), ventricular septal defect (30%), transverse arch hypoplasia (30%) and aortic stenosis (15%).[2-4]

Aortic coarctation is known to result from defects in the fourth and sixth aortic arches during embryonic development. However, the underlying mechanism has not been fully understood, despite the proposed ductal tissue theory and the reduced flow theory.[5]

The prevalence of aortic coarctation per 10.000 live births is 3-4 and it forms 5-7% of congenital heart diseases.[6-8] Patients reaching adult age with aortic coarctation may have a wide range of complaints associated with the extent of the stenosis and accompanying lesions. Detection of hypertension or murmur on routine physical examination may suggest aortic coarctation in asymptomatic patients. Patients may present with mild to moderate symptoms including headache, weakness in the lower extremities, and cold legs, while some may have severe complaints such as back pain and shock associated with aortic dissection, loss of consciousness due to intracranial hemorrhage or high fever due to infective endocarditis.[7]

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Corresponding address: İbrahim Özdoğru, M.D., Erciyes Üniversitesi Tıp Fakültesi, Kardiyoloji Anabilim Dalı Yılmaz ve Mehmet Öztaşkın Kalp Hastanesi, 38039 Kayseri
Tel: +90 - 352 437 49 37 e-mail: iozdogru@erciyes.edu.tr

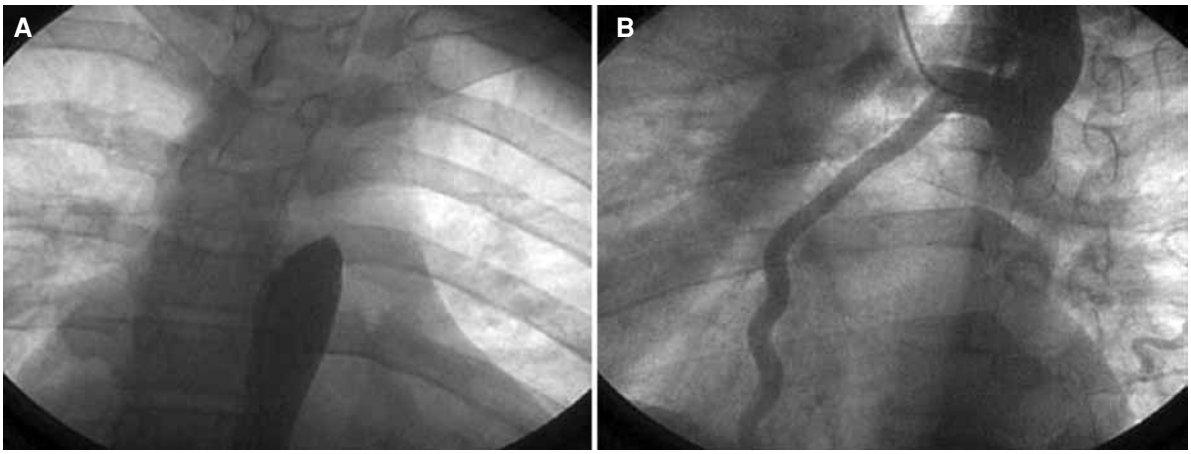

Figure 1. (A) Image of interrupted aorta in aortography which was performed through the right femoral artery entry pathway. **(B)** Image of interrupted aorta in aortography which was performed through the right radial artery pathway and significantly dilated right internal mammarian artery.

CASE REPORT

A-19-year-old male patient visiting the Erciyes University Faculty of Medicine, Cardiology outpatient clinic in order to get a health report was hospitalized for further investigation due to the presence of cardiac murmur and hypertension. Physical examination of the asymptomatic patient without any growth retardation revealed a 1-2/6 degree systolic murmur in the apex and back, and 2/4 degree diastolic murmur in the pulmonary focus. Blood pressure of upper extremity was 160/80 mmHg and of lower extremity was 80/50 mmHg. The two-sided femoral and dorsalis pedis pulses were weak, while the tibialis pedis pulse was nonpalpable. The electrocardiogram showed sinus rhythm and met the criteria of left ventricular hypertrophy. Telecardiography also showed coastal notches. Transthoracic echocardiography (TTE) which was performed with the pre-diagnosis of aortic coarctation, demonstrated rupture of the sinus of Valsalva and bicuspid aortic valve as well as left ventricular hypertrophy. Cardiac catheterization was considered when the echocardiography showed no gradient in the aortic valve and descending aorta. An interrupted image of the aorta was observed at the level where a 0.38-inch guide wire could not pass immediately distal to the left subclavian artery during catheterization (Figure 1a). Following the failure of passage with a *floppy* wire at this area, the catheterization procedure was maintained by using the right radial artery. Aortography confirmed rupture of the sinus of Valsalva. Imaging of the aortic arch revealed an interrupted aorta located immediately distal to the subclavian artery of descending aorta, and collaterals to lower arterial system on both sides of the internal mammary artery with significant dilatation (Figure 1b). The catheterization procedure was terminated considering the diagnosis of interrupted aorta findings. 16-slice computed tomography (CT) angiography also demonstrated similar findings (Figure 2). For further delineation, a transesophageal echocardiography (TEE) was performed to confirm the diagnosis and find out other possible pathologies overlooked due to the low prevalence of interrupted aorta in asymptomatic adult patients. Additionally, an open foramen ovale was found, while rupture of the sinus of Valsalva and bicuspid aortic valve were confirmed (Figure 3a). In addition to the results of TEE, color Doppler showed a passage of the descending aorta at the site where the interrupted aorta was observed during catheterization and CT angiography.

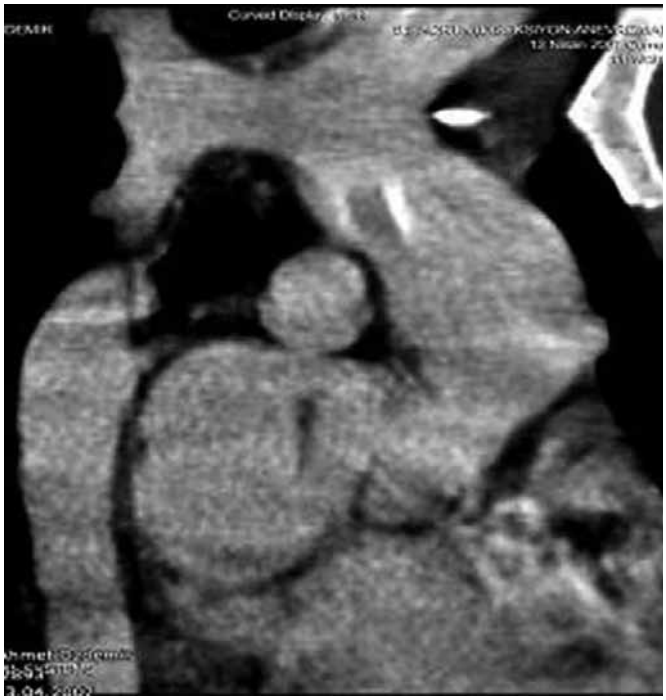

Figure 2. Computed tomography angiography showing interrupted aorta. Collateral observed between the ascending and descending aorta.

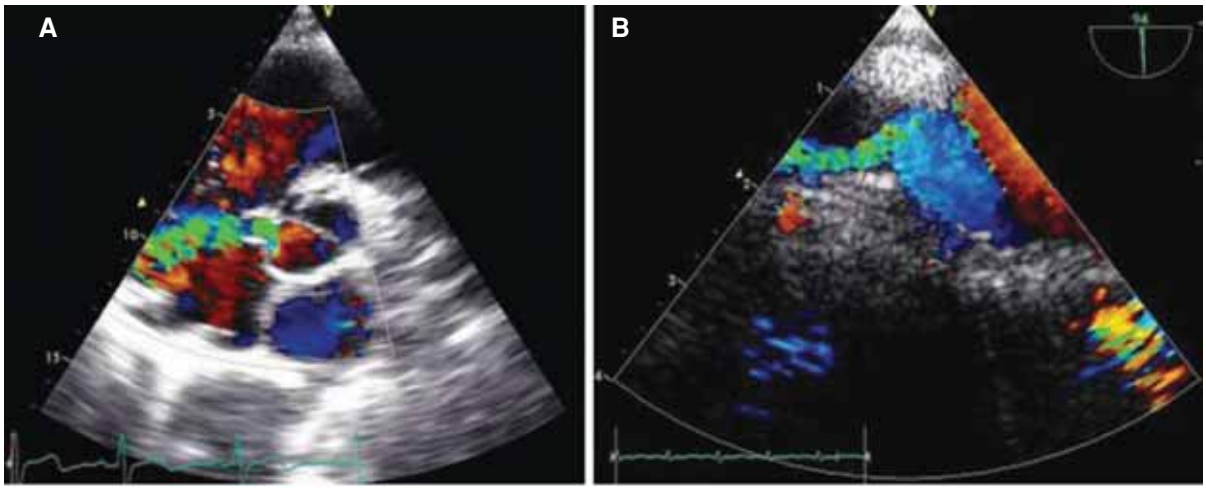

Figure 3. (A) Transesophageal echocardiography showing rupture of the sinus of Valsalva to the right atrium and bicuspid aortic valve. **(B)** The region of interruption observed in the aortography and CT angiography, showing passage by color Doppler imaging.

The patient was scheduled for surgery due to a diagnosis of aortic coarctation (Figure 3b). Patent ductus arteriosus, a common pathology accompanying interrupted aorta, was not observed by the performed imaging methods. At surgery, severe aortic coarctation was noted and corrected by grafting after removal of the area of narrowness and the rupture of the sinus of Valsalva was corrected.

DISCUSSION

Aortic coarctation is one of the leading congenital anomalies in adults. It is also one of the correctable causes of hypertension in adult population. 60% of untreated infants with severe stenosis, depending on the severity of the stenosis and concomitant cardiovascular disorders die within the first year of their lives; 25% of those who survive die before the age of 20 and 50% of the survivors die before the age of 32[9] Mean life span is 34 years.[10] Heart failure (26%), aortic rupture (21%), infective endocarditis (18%) and intracranial hemorrhage (18%) were the most common causes of mortality.[11]

Aortic coarctation is termed as 'complex coarctation' when it presents with severe cardiovascular diseases such as ventricular septal defect and obstruction of left ventricular exit pathway. Bicuspid aorta and aneurysm in the circle of Willis may be seen in simple coarctation, which is generally seen in adults; and serious disorders usually don't accompany simple coarctation.[10] In our case, rupture of the sinus of Valsalva is a rare pathology, accompanying aortic coarctation, and has led to the diagnosis of complex coarctation mostly seen in adults. We considered that hypertension could have contributed to the rupture of sinus of Valsalva.

End to end anastomosis if possible or graft interposition, patch aortoplasty or subclavian flap angioplasty may be used in the surgical correction of aortic coarctation.[12] Singer et al. (1982) were the first to use the technique of balloon angioplasty in aortic coarctation, and stent application has been started 10 years after.[13] Though there is lack of data from randomized trials, the stent procedure appears to be superior to balloon angioplasty. Both methods may be used as alternatives to surgical interventions in experienced centers when lesions are suitable.[14] In our case surgical therapy with graft interposition was performed since an interventional procedure was not deemed appropriate.

An interrupted aorta is a condition where by there is no link between both parts of the aortic arch. In 1959, Celoria and Patton categorized this anomaly according to location as follows:[12] Type A, interruption distal to the left subclavian artery; Type B, interruption between the left common carotid artery and left subclavian artery, and Type C, interruption between the brachiocephalic trunk and the common carotid artery. Patients generally die at an early age and the anomaly is often accompanied by complex cardiovascular disorders. In addition, although rare, asymptomatic adult patients may be also seen. These patients are generally diagnosed while investigating the etiology of hypertension or incidentally like in the case of mild symptomatic patients with aortic coarctation.[15,16]

Transthoracic echocardiography is used in daily clinical practice for the evaluation of the origin of embolism, diagnosis and the follow up of infective endocarditis, detection of aortic dissection and aneurysm, assessment of mitral regurgitation, diagnosis of congenital lesions and the assessment of prosthetic valves.[17] Transthoracic echocardiography, catheterization, computed tomography and magnetic resonance imaging are used to diagnose aortic coarctation and interrupted aorta. Transesophageal echocardiography is generally performed to investigate concomitant lesions. Of note, we did not find any data in the literature related to the use of TEE in the differential

diagnosis of aortic coarctation and interrupted aorta. In the differential diagnosis, TEE was performed in our case since we found that it to be superior to CT angiography and aortography. In particular, TEE can be used when a definitive diagnosis is not obtained.

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