

A rare combination of vascular anomalies: Hypoplastic aortic arch, coarctation of the aorta and poststenotic aneurysm

Nadir bir damar anomalisi kombinasyonu: Hipoplastik aort yayı, aort koarktasyonu ve poststenotik anevrizma

Nermin Bayar, M.D., Şakir Arslan, M.D., Çağın Mustafa Üreyen, M.D.,
Selçuk Küçükseymen, M.D., Bekir Erol, M.D.#

Department of Cardiology, Antalya Training and Research Hospital, Antalya

#Department of Radiology, Antalya Training and Research Hospital, Antalya

Summary– Coarctation of the aorta is the fifth most common congenital cardiac anomaly encountered in adults. It is important for prognosis to diagnose and treat this anomaly early. An aneurysm might develop due to tunica media abnormalities in patients with coarctation of the aorta. We hereby present an adult case with a very rare combination of vascular anomalies including ascending aorta aneurysm, hypoplastic aortic arch, coarctation of the aorta and poststenotic aneurysm.

Coarctation of the aorta constitutes 5-8% of all congenital cardiac anomalies.^[1] Moreover, bicuspid aortic valve and ventricular septal defect may accompany coarctation of the aorta.^[2] An aortic aneurysm may also develop owing to medial abnormalities in the aortic wall of the ascending aorta and/or the para-coarctation region.^[3]

We report a 41-year-old male patient with an ascending aorta aneurysm, hypoplastic aortic arch, coarctation of the aorta and poststenotic aneurysm demonstrated with three-dimensional (3D) reconstructed computed tomography (CT).

CASE REPORT

A 41-year-old male patient was admitted to the cardiology outpatient clinic with a complaint of exertional dyspnea. The patient had been taking metoprolol 50 mg and ramipril 10 mg for 5 years and his blood

Özet– Aort koarktasyonu erişkinlerde beşinci sıklıkta görülen doğumsal kalp anomalisidir. Prognoz için erken tanı ve tedavi önemlidir. Aort koarktasyonu bulunan olgularda tunika medya anormallikleri anevrizma gelişimine neden olabilmektedir. Bu yazıda, çıkan aort anevrizması, hipoplastik aort yayı, aort koarktasyonu ve poststenotik anevrizmayı içeren çok nadir damar anomalisi birlikteliği olan erişkin bir hasta sunuldu.

pressure was regulated. Physical examination revealed a II/IV diastolic murmur in the meso-cardiac area. Lower extremity pulses were weak and there was a 20 mmHg difference between upper extremity systolic blood pressures. His electrocardiogram was unremarkable. Left ventricular diffuse hypokinesia with an ejection fraction of 35%, left ventricular hypertrophy and left heart chambers dilatation were ascertained on transthoracic echocardiography (TTE). The aortic valve was tricuspid and there was third degree aortic insufficiency with an ascending aorta aneurysm of 52 mm. Left heart catheterization was intended, but the guidewire could not be advanced through the thoracic aorta to the arcus aorta via the left femoral artery. A stenotic segment between the arcus aorta and thoracic aorta was detected by means of contrast media injec-

Abbreviations:

3D	Three-dimensional
CT	Computed tomography
TTE	Transthoracic echocardiography

Received: November 18, 2014 Accepted: January 08, 2015

Correspondence: Dr. Nermin Bayar. Öğretmenevleri Mahallesi, 914. Sokak, 19. Cadde, Fetih Konakları, B Blok, Daire: 5, Konyaaltı, Antalya, Turkey.

Tel: +90 242 - 249 44 00 e-mail: dr.nermin@mynet.com

© 2015 Turkish Society of Cardiology



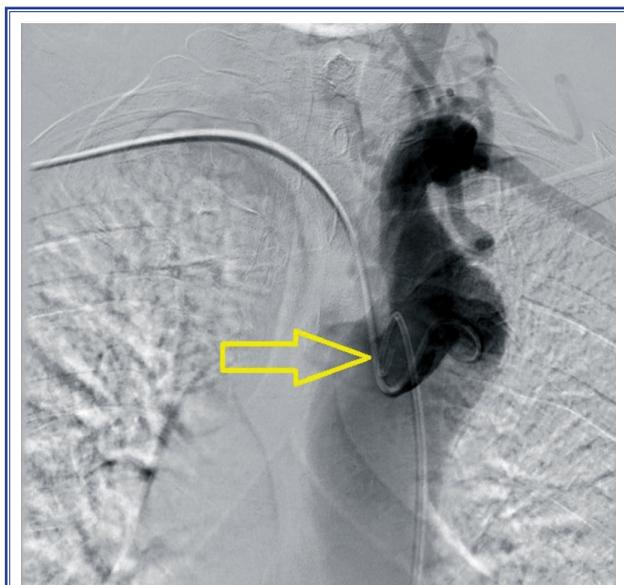


Figure 1. Stenotic segment of aorta demonstrated by synchronous injection of contrast agent from proximal and distal of coarctation.

tion from the thoracic aorta. Therefore, another catheter was forwarded via the right brachial artery to the proximal stenotic segment and synchronous contrast agent injection was performed through the proximal and distal segments. Digital subtraction angiography (DSA) of the aorta revealed the coarctation (Figure 1). The peak-to-peak pressure gradient of the coarctation was 15 mmHg. No significant lesion was as-

certained on coronary angiography. An aneurysm of the ascending aorta, coarctation of the aorta following the hypoplastic segment of the distal arcus aorta, and an aneurysm of diameter of 51 mm distal to the coarctation were confirmed with 3D-reconstructed CT (Figure 2a, b). Percutaneous intervention was not an option due to the proximal-distal diameter mismatch and presence of the ascending aortic aneurysm. The patient refused the recommended surgery, and he was medically treated and followed up.

DISCUSSION

Coarctation of the aorta is the fifth most common congenital cardiac anomaly encountered in adults.^[1] A decline of aortic elasticity and an increment of stiffness due to abnormalities of the tunica media of the aortic wall could lead to ascending aorta dilatation, aneurysm and aortic valvular insufficiency.^[4] Moreover, an aneurysm in late term may also develop in patients who have undergone an operation for coarctation of the aorta. Due to the impact of genetic or environmental factors, poststenotic aneurysms may develop distal to the coarctation in patients without any intervention. Various studies have reported combined aortic anomalies in patients with genetic disorders such as Klippel-Feil syndrome.^[5] We could not perform any genetic study concerning our patient. However, we considered that asymmetric and enhanced shear stress

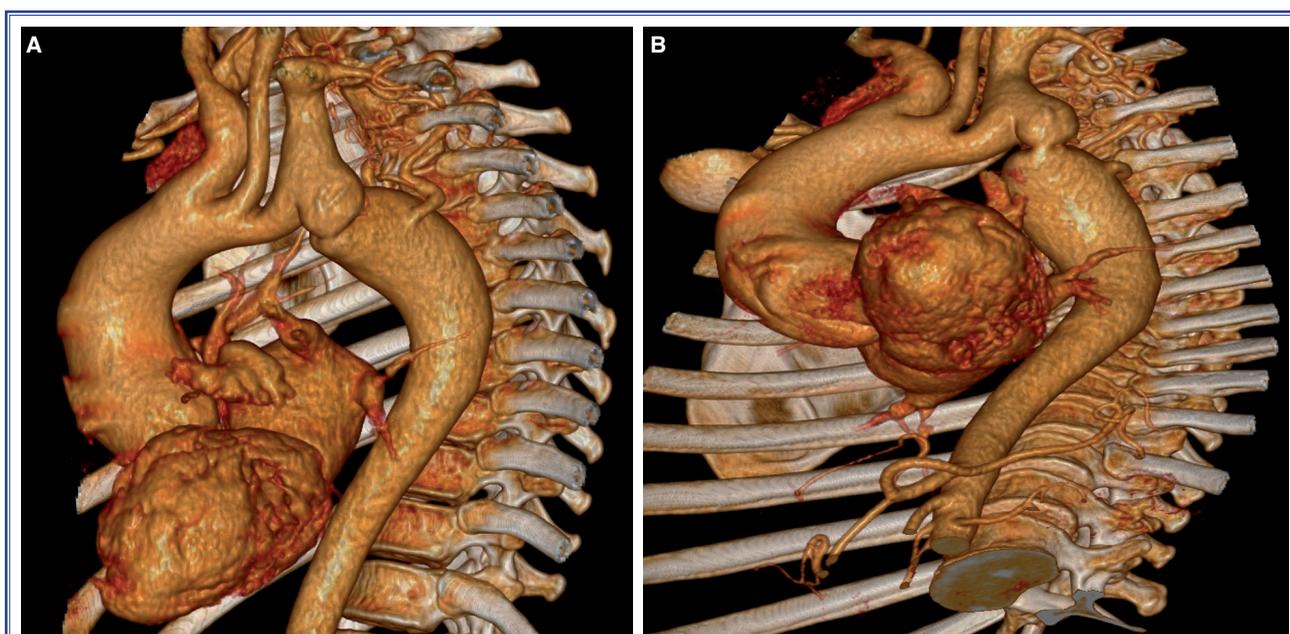


Figure 2. (A, B) Hypoplastic aortic arch, coarctation of the aorta and poststenotic aneurysm depicted by 3D-reconstructed CT.

caused by the hypoplastic aortic arch might contribute to poststenotic aneurysm development.

Coarctation of the aorta is generally diagnosed by means of suprasternal window of TTE or cardiac catheterization. On the other hand, a 3D-reconstructed CT or MRI could make a major contribution to diagnosis in patients with low echogenity or, as in our patient, with accompanying complicated anomalies around the coarctation zone which may cause difficulty in diagnosis.

Intervention of coarctation of the aorta is a class 1C indication for patients with at least a 20 mmHg gradient in the coarctation region, or in patients with a lower gradient with well-developed collaterals, demonstrated according to ACC guidelines.^[6] Surgical treatment was chosen for our patient by virtue of well-developed collaterals with a gradient of 15 mmHg and the presence of a concomitant aortic aneurysm. Overall hospital mortality in the literature is approximately 1%. However, accompanying anomalies and diminished left ventricular ejection fraction are expected to augment this percentage.^[7] Furthermore, various articles refer to the enhanced risk of development of an aneurysm around the operated segment in late term in patients with concomitant hypoplastic aortic arch.^[8]

Detailed examinations aimed at etiology and early treatment of hypertension and heart failure in young patients may be vital for treatment and prognosis of patients before irreversible changes occur. In this regard, utilisation of a 3D-reconstructed CT or MRI may provide substantial contribution to diagnosis.

Conflict-of-interest issues regarding the authorship or article: None declared.

REFERENCES

1. Cetta F, Lichtenberg RC, Clark SE. Adults with congenital heart disease. *Compr Ther* 1992;18:33-7.
2. Barik R, Patnaik AN, Mishra RC, Kumari NR, Gulati AS. A tetrad of bicuspid aortic valve association: A single-stage repair. *J Cardiovasc Dis Res* 2012;3:143-6. [CrossRef](#)
3. Roberts CS, Roberts WC. Dissection of the aorta associated with congenital malformation of the aortic valve. *J Am Coll Cardiol* 1991;17:712-6. [CrossRef](#)
4. Niwa K. Aortopathy in Congenital Heart Disease in Adults: Aortic Dilatation with Decreased Aortic Elasticity that Impacts Negatively on Left Ventricular Function. *Korean Circ J* 2013;43:215-20. [CrossRef](#)
5. Sabol F, Kolesar A, Toporcer T, Bajmoczy M. Surgical management of a hypoplastic distal aortic arch and coarctation of aorta in a patient with Klippel-Feil syndrome, ascending aortic aneurysm and bicuspid aortic valve. *Interact Cardiovasc Thorac Surg* 2014;19:708-10. [CrossRef](#)
6. Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, et al. ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (writing committee to develop guidelines on the management of adults with congenital heart disease). *Circulation* 2008;118:714-833.
7. Sakurai T, Stickley J, Stümper O, Khan N, Jones TJ, Barron DJ, et al. Repair of isolated aortic coarctation over two decades: impact of surgical approach and associated arch hypoplasia. *Interact Cardiovasc Thorac Surg* 2012;15:865-70. [CrossRef](#)
8. Bogaert J, Gewillig M, Rademakers F, Bosmans H, Verschakelen J, Daenen W, et al. Transverse arch hypoplasia predisposes to aneurysm formation at the repair site after patch angioplasty for coarctation of the aorta. *J Am Coll Cardiol* 1995;26:521-7. [CrossRef](#)

Key words: Aneurysm; aortic coarctation; hypoplastic aortic arch.

Anahtar sözcükler: Anevrizma; aort koarktasyonu; hipoplastik aort yayı.