Aortic coarctation accounts for 5–8% of congenital heart defects. When accompanied by other congenital heart defects, it is called complex aortic coarctation. Its association with bicuspid aortic valve is between 60% and 85%. Without correction of the defect, mean survival of these patients is 35 years. Survival greater than 65 years is extremely rare and few cases have been reported in the literature. We report the case of a 68-year-old woman with history of resistant hypertension since early adulthood who presented progressive dyspnea during the last two years.

Physical examination was remarkable for short stature, underdevelopment of lower extremities and laterally displaced apical impulse. Auscultation over the aortic area revealed a harsh meso-telesystolic crescendo-decrescendo murmur IV/VI radiating to carotids and apex with Gallavardin phenomenon. A loud systolic murmur was also heard over the left paravertebral area. Carotid pulse was parvus et tardus. Lower extremities pulses where markedly diminished. Blood pressure was 152/90 mmHg in the upper extremities and 64/40 mmHg in the lower extremities. Anklet...
was 198/60 mmHg. No significant coronary artery stenosis were found.

Based on the association of a bicuspid aortic valve and a coarctation of the aorta, she was diagnosed with a complex aortic coarctation and severe aortic valve stenosis. Considering the anatomic complexity for an endovascular treatment (Fig. 2E), the patient was proposed for a surgical coarctectomy and aortic valve replacement in a two stage surgery approach. The heart team accepted the surgical proposition but the patient refused the surgical intervention during the index hospitalization. She continues the follow up while considering the intervention. Notably, without a surgical correction, the survival likelihood of this patient is poor in the middle term, the mortality of patients with severe aortic stenosis presenting with heart failure is around 50% at 2 years, furthermore, the added risk of aortic rupture/dissection and cerebral hemorrhage in patients with aortic coarctation confers a bad prognosis.
Complex aortic coarctation and a bicuspid aortic valve with severe stenosis

Fig. 2  (A) Transesophageal echocardiography aortic short axis view showing a Type 1 bicuspid aortic valve (right–left coronary cusp fusion) with important thickening and color Doppler acceleration. (B) 3D imaging of the aortic valve. (C) Maximum intensity projection CT angiography and (D) volumetric reconstruction showing the severe aortic coarctation site (white arrow) measured 35 mm after the origin of the left subclavian artery along with marked collateral circulation (mammary and vertebral arteries). (E) Coarctation site angiography demonstrating the reduction of the luminal area (white arrow) and the anatomic complexity for an endovascular treatment.

References


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