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Pseudocoarctation of the aorta: A rare congenital aortic disease

CASE REPORT

INTRODUCTION

Aortic pseudocoarctation is a rare congenital aortic anomaly that causes elongation, stenosis and kinking of the aorta at the isthmus level. Although aortic coarctation and pseudocoarctation share a similar clinical spectrum, pseudocoarctation rarely results in a significant gradient in descending aorta and hemodynamic consequence. Therefore, it is critical to differentiate between coarctations. All the imaging modalities, especially cardiac CT angiography and catheterization, are very important in the differential diagnosis. Our case highlighted the differences between aortic pseudocoarctation and coarctation, with other cardiac anomalies accompanying pseudocoarctation and also emphasized the importance of cardiac imaging in the differential diagnosis of pseudocoarctation.

CASE REPORT

A 23-year-old male patient was admitted to the cardiology outpatient clinic to investigate the etiology of his hypertension. His medical and family histories were unremarkable. On physical examination of the patient, the blood pressure in the left arm was 164/96 mm Hg and in the right arm was 161/92 mm Hg; and there was no difference in the blood pressures between the lower and upper extremities. Peripheral pulses were bilaterally palpable, radio-femoral, the radio-radial delay was not observed. Electrocardiogram was normal sinus rhythm. Pathological findings on transthoracic echocardiography were bicuspid aortic valve (type 2, non-coronary cusp and right coronary cusp fusion) and in the suprasternal evaluation of descending aorta, peak systolic gradient was measured as 20 mm Hg on Doppler evaluation (Fig. 1a, 1b). Buckling of the aorta was seen on the patient's chest x-ray (Fig. 1c). CT angiography was performed for the preliminary diagnosis of aortic coarctation, and it was observed that the distal aortic arch had kinking at the level of the isthmus, and the diameter of the narrowest part was measured as 13×11 mm (Fig. 2, Video 1). In addition, it was observed that collateral circulation, which is the typical finding of coarctation on CT angiography, did not develop in this patient. A peak systolic gradient of 20 mm Hg was observed between the pre

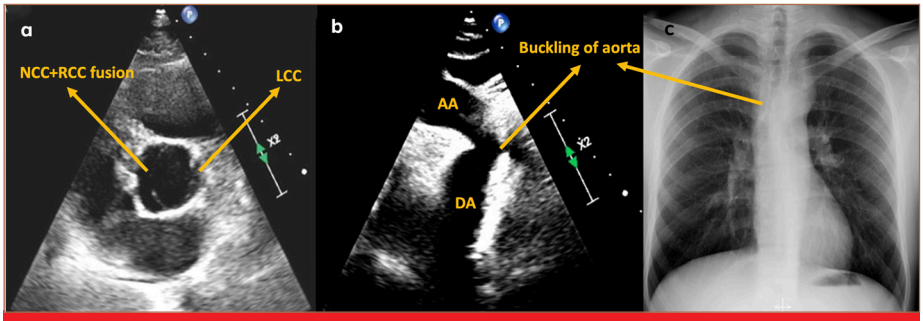

Figure 1. a) Transthoracic echocardiography, parasternal short-axis view, showing a bicuspid aortic valve with a fusion of the right and non-coronary cusps. b) Suprasternal view of transthoracic echocardiography and buckling of the descending aorta. c) Chest x-ray, buckling of the aorta

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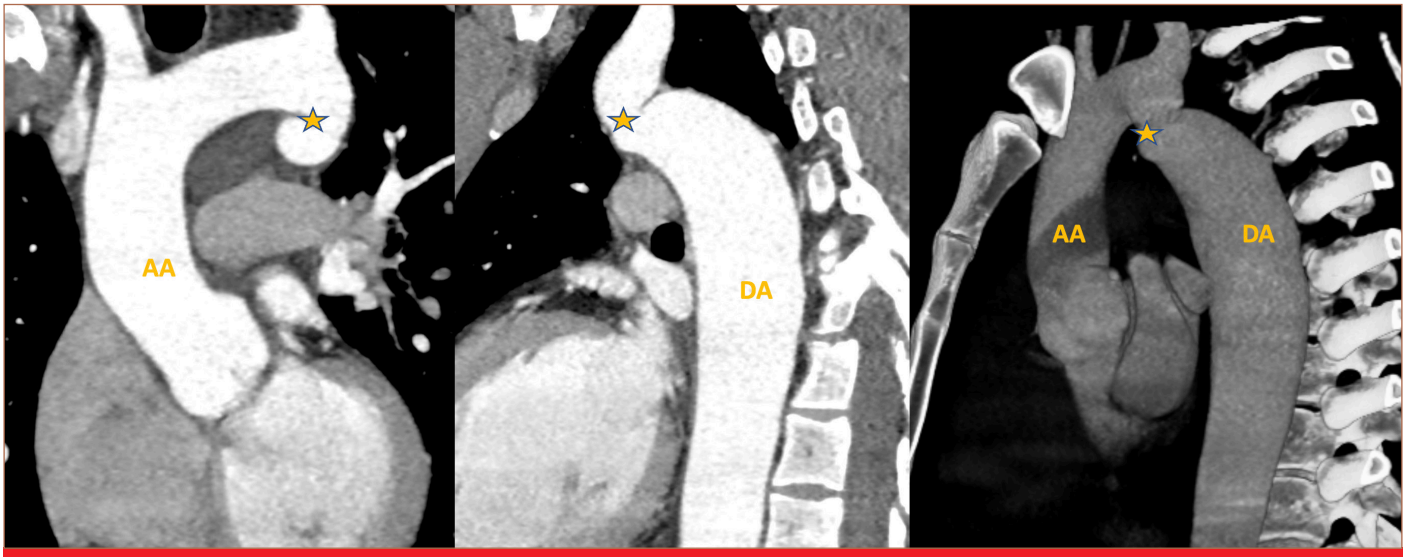

Figure 2. CT angiography showing an elongated and kinked aortic arch (*Elongation and buckling of the aorta)

and post pseudocoarctation segments in the catheterization study performed on the patient for aortic pressure study. On the basis of these clinical and imaging findings, the patient was diagnosed with aortic pseudocoarctation.

DISCUSSION

Pseudocoarctation of the aorta is a very rare congenital anomaly of the aorta that causes elongation, stenosis, twisting, and kinking of the aorta at the isthmus level as a result of compression of the 3rd and 7th dorsal aortic segments during the embryological period (1). The mean age at diagnosis is 43 years, and it is observed with equal frequency in men and women (1). The etiology of aortic pseudocoarctation is unknown; however, this anomaly has been associated with familial conditions like chromosomal abnormalities (18p–/18q+, Turner, Noonan, and Hurler) and congenital heart diseases such as the bicuspid aorta (2). There are very few cases of aortic pseudocoarctation reported worldwide. The last systematic review published in 2015 found that there have been at least 18 instances during the previous 20 years (1). It has been reported that approximately half of these patients present with hypertension and the rest with symptoms such as dyspnea and dysphagia owing to lung and esophageal compressions, and abdominal pain because of aortic dissection (1).

Gay and Young (3) defined diagnostic criteria in 1969 and included an abnormal posteroanterior chest roentgenogram, the absence of upper/lower extremity pressure difference, no signs of increased collateral circulation, and definitive aortogram images. Nowadays, the initial test for diagnosis is usually echocardiography owing to its easy accessibility and importance in the evaluation of descending aorta and associated congenital defects. A chest x-ray is also a simple test that can help in the diagnosis. The "3 sign" typical for aortic coarctation and notching of the ribs are not seen in patients with pseudocoarctation (4). Although there is no clear definition of chest x-ray findings in the literature, it has been reported in some case reports that a "pseudo 3 sign" and buckling of the aorta may be seen (4, 5). CT angiography and MRI angiography are important imaging modalities to evaluate the narrowed segment of the aorta and to rule out associated aortic aneurysm or aortic dissection (1). Cardiac catheterization is the gold standard for accurate measurement of pressure gradient if diagnostic uncertainty exists.

Aortic pseudocoarctation and coarctation are in the same disease spectrum, and their clinical presentations differ depending on whether there is hemodynamically significant stenosis in the descending aorta or not. Although there are significant gradient-related hemodynamic results in the aortic segment, such as the difference in blood pressure between the upper and lower extremities, inability to palpate lower extremity peripheral pulses, and delayed radiofemoral pulse in true coarctation; these findings are not observed in pseudocoarctation. The clinical findings in our patient, especially the absence of pulse delay and the blood pressure difference between the four extremities, suggested that kinking and an elongated aortic segment did not cause a significant blood flow obstruction in the descending aorta. This hypothesis was supported by the absence of collateral circulation on thoracic CT angiography and the absence of a significant gradient in the aortic catheterization study. In conclusion, we diagnosed our patient with aortic pseudocoarctation.

Specific guidelines on the management of pseudocoarctation are lacking. Conservative management of pseudocoarctation, which does not cause significant hemodynamic stenosis and aneurysm formation, are necessary (1, 6). Unnecessary surgical interventions should be avoided in these patients; however, the presence of a pseudoaneurysm or aortic aneurysm of the aorta adjacent to pseudocoarctation, which carries a high risk of rupture, requires prompt intervention (1, 7). The aortic diameters in our patient were within normal limits, and there was no aneurysmal enlargement. Antihypertensive treatment of our patient with hypertension, bicuspid aortic valve, and pseudocoarctation was adjusted to a blood pressure target of <130/80 mm Hg.

Avoidance of isometric exercise with high static load, annual follow-up, and bicuspid aortic valve screening for first-degree relatives were recommended.

CONCLUSION

Aortic pseudocoarctation is a rare congenital aortic disease. Because of its generally asymptomatic presentation and benign course, it is very important to distinguish it from coarctation of the aorta, which will prevent unnecessary interventional and surgical procedures and associated risks. There is no standard management algorithm, and limited literature is available for pseudocoarctation. More evidence-based case reports and studies on treatment modalities and timing are needed.

Declaration: This case report was presented as an oral presentation in 17. International Congress of Update in Cardiology and Cardiovascular Surgery on 5-7 November 2021.

Informed consent: Informed consent was obtained from the patients for the publication of the case report and the accompanying images.

Video 1. CT angiography with three-dimensional reconstruction showing an elongated and kinked aortic arch

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