Pseudocoarctation of the Aorta: A Rare Congenital Aortic Disease.

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Abstract

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 haemodynamic consequence. Therefore, it's critical to differentiate in coarctation. All imaging modalities but especially cardiac CT angiography and catheterization are very important in the differential diagnosis. Our case highlighted to differences between aortic pseudocoarctation and coarctation, with other cardiac anomalies accompanying pseudocoarctation. And also the importance of cardiac imaging in the differential diagnosis of pseudocoarctation was emphasized.

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Running title: Aortic Pseudocoarctation

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Abstract

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spectrum, pseudocoarctation rarely results in a significant gradient in descending aorta and haemodynamic consequence. Therefore, it's critical to differentiate in coarctation. All imaging modalities but especially cardiac CT angiography and catheterization are very important in the differential diagnosis. Our case highlighted to differences between aortic pseudocoarctation and coarctation, with other cardiac anomalies accompanying pseudocoarctation. And also the importance of cardiac imaging in the differential diagnosis of pseudocoarctation was emphasized.

Case presentation

A 23-year-old male patient was admitted to the cardiology outpatient clinic to investigate the aetiology of hypertension. His medical and family history was unremarkable. In the physical examination of the patient, the blood pressure taken from the left arm was 164/96 mmHg, the blood pressure taken from the right arm was 161/92 mmHg, and there was no difference in blood pressure between the lower and upper extremities. Peripheral pulses were bilaterally palpable, radio-femoral, radio-radial delay was not observed. Electrocardiogram was in normal sinus rhythm. Pathological findings in transthoracic echocardiography were bicuspid aortic valve (type 2, NCC+RCC fusion) and in the suprasternal evaluation of descending aorta, peak systolic gradient was measured as 20 mm Hg in doppler evaluation **(Figure 1A-B).** Buckling of the aorta was seen on the patient's chest x-ray (**Figure 1C).** CT angiography was performed for the preliminary diagnosis of aortic coarctation, and it was observed that the distal aortic arch had king formation at the level of the isthmus, and the diameter of the narrowest part was measured as 13*11 mm **(Figure 2** -Video 1). In addition, it was obtained that collateral circulation, which is the typical finding of coarctation on CT angiography, did not develop in this patient. A peak 20 mm Hg systolic gradient qas observed between the pre and post psedocoarctation segment in the catheterization study performed on the patient for aortic pressure study. In the light of these clinical and imaging findings, the patient was evaluated as 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, and it is observed with equal frequency in men and women [1]. The etiology of aortic pseudocoarctation is unknown This anomaly has also 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 reported worldwide of aortic pseudocoarctation. The last systematic review published in 2015 ffound that there have been a at least 18 instances during the previous twenty years [1]. It has been reported that approximately half of these patients present with hypertension, the rest with symptoms such as dyspnea and dysphagia due to lung and esophageal compressions, and abdominal pain due to aortic dissection [1].

Gay and Young defined diagnostic criteria in 1969 and include the abnormal posteroanterior chest roentgenogram, the absence of an upper/lower extremity pressure difference, no signs of increased collateral circulation, and definitive aortogram images[3]. Nowadays, the initial test for diagnosis is usually echocardiography due to its easy accessibility and important in evaluation of descending aorta and associated congenital defects. Also, chest X-ray is 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 pseudocoarctation patients [4]. Although there is no clear definition of chest X-ray findings in the literature, It has been reported in some case reports that the '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. While 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 blood pressure difference between the four extremities, suggested that king and elongated aortic segment not caused significant blood flow obstruction in desendan aorta. This hypothesis was supported by the absence of collateral circulation in 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, is recommended[1,6]. Unnecessary surgical interventions should be avoided in these patients. However, the presence of a pseudoaneurysm or aortic aneurysm of aorta adjacent to pseudocoarctation, which carries a high risk of rupture, requires prompt intervention[1,7]. Our patient's aortic diameters were within normal limits, 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 mmHg. 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. Due to its generally asymptomatic presentation and benign course, it is very important to distinguish it from coarctation of the aorta and proper diagnosis of pseudocoarctation 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 research on treatment modalities and timing are needed.

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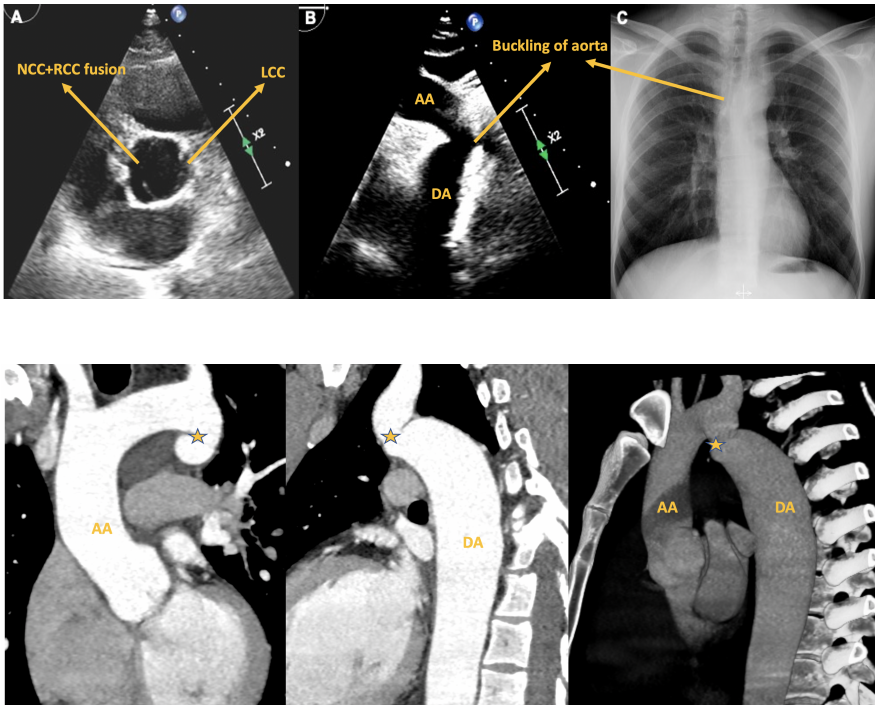
Figure 1A-C: A, Transthoracic echocardiography parasternal short-axis view, a bicuspid aortic valve with a fusion of the right and non-coronary cusps. B, Suprasternal view of transthoracic echocardiography and buckling of the descending aorta. C, Chest X-ray, buckling of the aorta. Abbreviations:NCC, Non-coronary cusp, RCC, right coronary cusp, LCC, Left coronary cusp, AA, Ascending aorta, DD, Descending aorta.

Figure 2: CT angiography, showing an elongated and kinked aortic arch (* Elongation and buckling of the aorta). Abbreviations: AA, Ascending aorta, DD, Descending aorta

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



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