Journal of the
Bahrain Medical Society

CASE REPORT

Congenital kinking of aorta – A rare aortic arch anomaly

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Received date: July 21, 2021; **Accepted date:** December 02, 2021; **Published date:** December 31, 2021

Abstract

Congenital kinking of aorta or pseudocoarctation is a rare anomaly of the aortic arch with elongation and kinking of aorta at the level of ligamentum arteriosum. This anomaly is not a true coarctation as there is no significant hemodynamic obstruction or pressure gradient across the lesion. We report two cases with this anomaly diagnosed during the evaluation for incidental finding of murmur. Both patients had flow gradient across the kinked segment of aorta by echocardiography but did not require any intervention. Long term complication is aneurysmal dilatation and rupture beyond the kinked segment. Computed tomography of the aorta is a simple noninvasive diagnostic modality for the definitive diagnosis. Surgical repair should be performed for all symptomatic individuals. Regular periodic follow up is mandatory for all asymptomatic patients.

Keywords: Aortic arch syndromes, Aortic coarctation, Congenital heart defects, Echocardiography, Thoracic aorta

Introduction

Congenital variants and anomalies of the aortic arch are uncommon. It is important to recognize these anomalies as they may be associated with vascular rings, congenital heart disease chromosomal abnormalities and can have important implications for prognosis and management. Congenital kinking of the aorta is a rare condition due to congenital

elongation of the aortic arch. The elongation leads to redundancy and kinking of the aorta at the level of the ligamentum arteriosum.[1] This condition is also termed as pseudocoarctation of aorta. Although there is echocardiographic pressure gradient across the arch of aorta like simple coarctation of aorta (COA) there is no significant hemodynamic obstruction or pressure gradient across the lesion (<25 mm Hg), and therefore no collateral vessel formation.[2] Only a few isolated case reports are published in the literature. We report two cases of this rare anomaly diagnosed during evaluation for incidental finding of murmur.

Case Presentation

Case 1

An 8-year-old asymptomatic female child was referred for evaluation of murmur which was detected during routine pediatric examination. On examination she was found to have weak femoral pulses with no radio femoral delay. The clinical blood pressure gradient between upper and lower limb was 14 mmHg which is less than the significant gradient of >20 mmHg. There was grade 3/6 ejection systolic murmur in inter scapular region. Chest radiography (X-ray) was normal. Transthoracic echocardiogram (TTE) showed normal left aortic arch and a tortuous segment with narrowing distal to left subclavian artery (LSCA). There was turbulent flow across the narrowed segment with significant peak pressure gradient (PG) of 60 mmHg and mean gradient of 30 mmHg with diastolic tailing. The pulse wave doppler flow across the abdominal aorta was also continuous instead of pulsatile flow. All these features were suggestive of COA. The left ventricle (LV) was of normal size and there was no left ventricular hypertrophy, and the LV function was normal.

Computed tomography of aorta (CT Aortogram) showed tortuous kinking of aortic arch just after the origin of LSCA with two very acute angulations, first 1mm distal to LSCA and the second at the junction of arch to descending aorta (Figure 1). The narrowest diameter at the acute angulation of was 9 mm with transverse diameter of 18 mm. The length of tortuous segment was 14mm (Figure 2). Ascending aortogram by cardiac catheterization also showed tortuous kinking of aorta 15 mm distal to LSCA (Figure 3). The invasive pressure gradient between transverse arch and descending aorta was only 13 mmHg. Hence no intervention was attempted, and the patient was advised for periodic follow up.

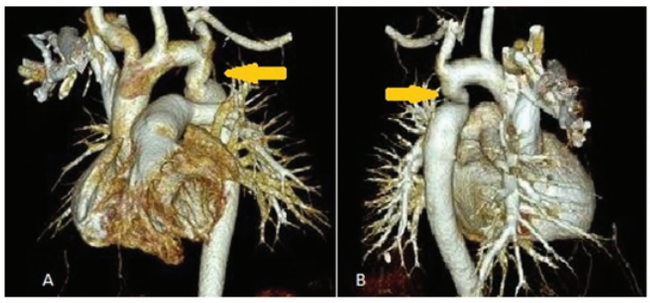

Figure 1: CT Aortogram – 3-Dimensional Volume rendering technique (3D VRT) image- Anterior (A) and posterior (B) views with arrow showing tortuous kinking of aorta distal to left subclavian artery

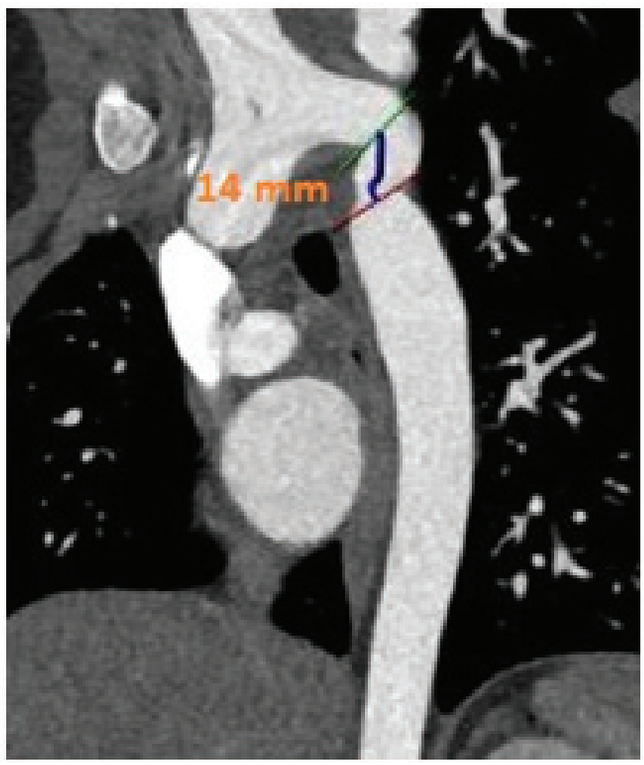

Figure 2: CT Aortogram – 2-dimensional (2D) image showing the length of the tortuous segment

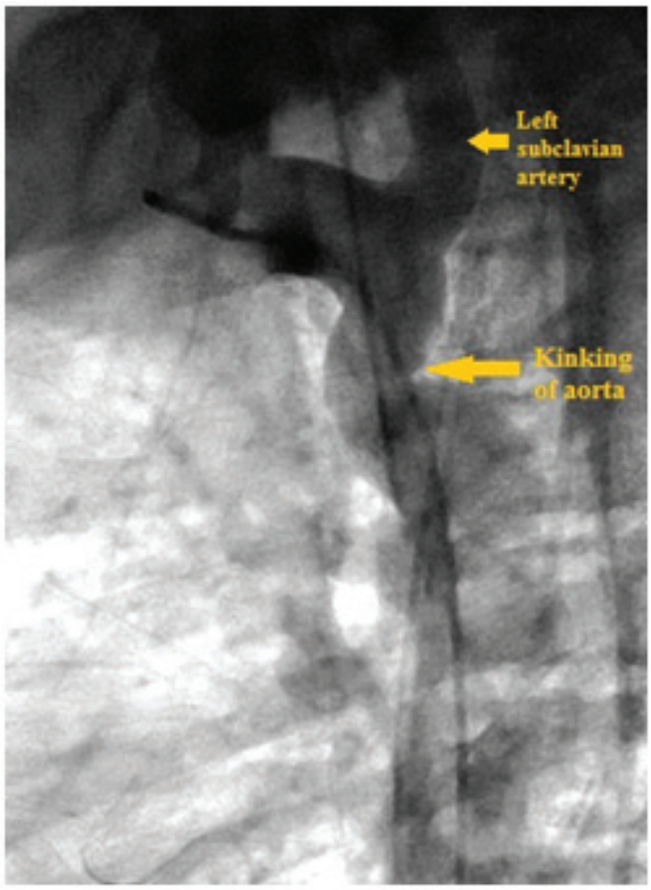

Figure 3: Cardiac catheterization – Aortogram – Arrow showing the kinking of aorta distal to left subclavian artery

Case 2

A 3-year-old male child was referred in view of incidental finding of murmur. On physical examination he was found to have feeble femoral pulses with no radio femoral delay. The clinical blood pressure gradient between upper and lower limb extremities was only 8 mmHg which was insignificant. On auscultation there was grade 2 ejection systolic murmur in the inter scapular region. Chest X-ray showed mildly widened superior mediastinum. TTE showed normal left aortic arch. There was tortuous kinking with narrowing of aorta at the level of isthmus just distal to LSCA with PG of 40mmHg and diastolic tailing suggestive of COA. Abdominal aorta pulse wave doppler was also continuous. Left ventricle was of normal size with normal function. CT aortogram showed tortuous kinking of aorta at the level of isthmus with mild post stenotic dilatation (Figure 4). Since there was no significant clinical blood pressure gradient between upper and lower limb extremities no intervention was attempted, and the patient was advised for regular follow up.

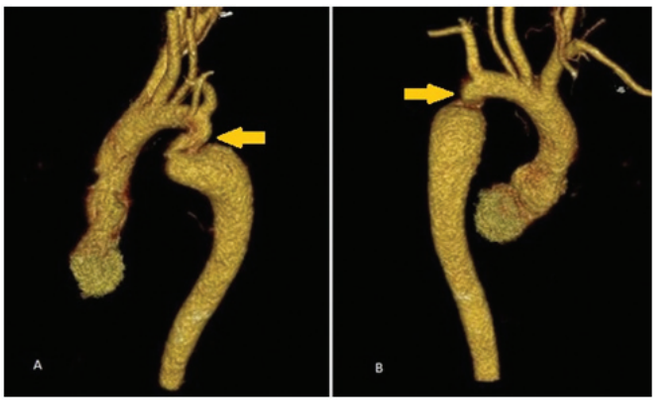

Figure 4: CT Aortogram – 3-Dimensional Volume rendering technique (3D VRT) image – Anterior (A) and posterior (B) views with arrow showing kinking of aorta distal to left subclavian artery

Discussion

Congenital kinking of aorta is caused by an abnormally long aortic arch extending into superior mediastinum above the clavicle. The elongation of the arch results in increase in distance between the origins of left common carotid artery and left subclavian artery. This causes kinking or buckling at the isthmus at the level of attachment of ligamentum arteriosum.[1]

During embryogenesis at 7th week of gestational age, failure of regression and fusion of 3rd through 7th segments of the dorsal aortic roots and 4th left aortic arch segment occurs. There is also associated cephalic shift of 7th dorsal intersegmental artery in the dorsal aorta. Hence, the aortic arch becomes longer. The redundant long aortic arch is kinked at the insertion point of ligamentum arteriosum.[3] Unlike true COA, there is no significant hemodynamic pressure gradient across the kink. Therefore, upper extremity hypertension, lower extremity hypotension, rib notching, delayed femoral pulses, and collateral blood flow are not present.

It is usually a benign asymptomatic condition as in our cases. Various presentations of pseudocoarctation reported in the literature were hypertension, dyspnea, dysphagia due to aneurysm formation and compression of esophagus, back pain due to dissection of aorta and as incidental finding of CT imaging of the chest.[4,5]

The most frequent radiological presentation of congenital kinking of aorta before the advanced diagnostic modalities was abnormal left superior mediastinal opacity on the postero- anterior chest X-ray.Souders, *et al* were the first to appreciate this entity of congenital kinking of the aortic arch without coarctation in three cases.[6] Their first case was a 58-year-old man with enlarged mediastinum on chest X-ray. The radio opacity above the aortic knob was misinterpreted as superior mediastinal tumor of Hodgkin's disease. Radiotherapy was initiated and repeat chest X-ray showed a still enlarged mediastinum. Hence thoracotomy was done which showed sharply kinked aortic arch. The apex of the kink was sited at the insertion of a short, taut, patent ductus arteriosus. There was no evidence of narrowing of the aorta. Division of the ductus did not alter the appearance. Repeat Chest X- ray three months later showed no change from the pre-operative films. Several patients have been subjected to therapeutic irradiation and to exploratory thoracotomy.[7] The mistaken interpretation of the radio opacity as a superior mediastinal tumor is a real and dangerous hazard.

Congenital kinking with pseudocoarctation can also be associated with bicuspid aortic valve, aortic stenosis, ventricular septal defect, atrial septal defect, patent ductus arteriosus, and sinus of Valsalva aneurysms.[3] It can also occur in Turner's syndrome, Noonan's syndrome, and Hurler's syndrome.[7]

Long term complication of pseudocoarctation is the occurrence of aortic aneurysm but the precise incidence of aneurysm formation is not clear. The pathogenesis of the aneurysmal dilatation may be due to turbulent flow beyond the kinked segment.[8] Turner and associates reported successive angiograms showing development of an aneurysm in a 36-year-old patient 12 years after the diagnosis of kinking of aorta.[9] Makani, *et al* reported a case of pseudocoarctation with cervical aortic arch complicated by multiple aneurysms requiring surgical correction.[10] Gay and Young reported the death of a patient caused by a ruptured thoracic aorta two years after the diagnosis of pseudocoarctation was made.[2] Surgical repair should be the treatment of choice for all the symptomatic patients and or those with associated aneurysm formation.[11,12]

Conclusion

Congenital kinking of aorta is a benign asymptomatic lesion. A thorough clinical examination and high clinical suspicion is needed in diagnosis of pseudocoarctation. Although there is flow gradient measured in the narrowed segment of aortic arch by Echocardiography other findings of true coarctation such as significant clinical gradient of > 20 mmHg between upper and lower limb extremities, hemodynamically significant obstruction are absent. Long term periodic follow up is mandatory because of risk of aneurysmal dilatation. This may cause sudden rupture or dissection which can be lethal. With the advent of high resolution CT Aortogram, the diagnosis and follow up of this lesion is feasible with less radiation exposure.

Declaration of Patient consent

The authors certify that they have obtained appropriate patient consent. The legal guardians have kindly consented to share the images and other clinical information for publication in a journal. The guardians understand that names and initials will not be published, and due efforts will be made to conceal the patient's identity.

Financial support and Sponsorship

Nil.

Conflicts of Interest

No Conflict of Interest.

Presentation at a meeting

Nil.

References

1. Steinberg I, Engle MA, Holswade GR, *et al*. Pseudocoarctation of the aorta associated with congenital heart disease: report of ten cases. *Am J Roentgenol Radium Ther Nucl Med*. 1969; 106:1-20.
2. Gay WA Jr, Young WG Jr. Pseudocoarctation of the aorta. A reappraisal. *J Thorac Cardiovasc Surg*. 1969;58(5):739–745.
3. Lavin N, Mehta S, Liberson M, *et al*. Pseudocoarctation of the aorta: an unusual variant with coarctation. *Am J Cardiol*. 1969; 24(4):584–590.

4. Amico A, Michaud JL, Baron O, *et al*. Pseudocoarctation as cause of a refractory hypertension. *J Cardiovasc Med*. 2007; 8(4):289–90.

5. Singh S, Hakim FA, Sharma A, *et al*. Hypoplasia, pseudocoarctation and coarctation of the Aorta – a systematic review. *Heart, Lung Circ*. 2015; 24(2):110–118

6. Souders CR, Pearson CM, Adams, HD. An aortic deformity simulating mediastinal tumor: a subclinical form of coarctation. *Dis of Chest*. 195l;20(1): 35 -45

7. Hoeffel JC, Henry M, Mentre B, *et al*. Pseudocoarctation or congenital kinking of the aorta: radiologic considerations. *Am Heart J* 1975; 89(4):428–436.

8. Kessler RM, Miller KB, Pett S, *et al*. Pseudocoarctation of the aorta presenting as a mediastinal mass with dysphagia. *Ann Thorac Surg*. 1993; 55(4):1003-1005.

9. T urner AF, Swenson BE, Jacobson G, *et al*. Kinking or buckling of the aorta: case report with complication of aneurysm formation. *Am J Roentgenol Radium Ther Nucl Med*. 1966; 97: 411-415

10. Makani S, Mitchell J, Metton O, *et al*. Surgical repair of a pseudocoarctation with cervical aortic arch complicated by multiple aneurysms of the aorta: a case report. *Pan Afr Med J*. 2017; 26:236.

11. Shindo S, Katsu M, Kojima A, *et al*. Thoracic aortic aneurysm associated with pseudocoarctation of the aorta. *Jpn Thorac Cardiovasc Surg*.2002; 50(12):520–522.

12. Sakai O, Yamagishi M, Shuntoh K, *et al*. Critical aneurysmal dilatation of congenital kinking of the aorta. *J Thorac Cardiovasc Surg*. 2001; 122:832-833