Pseudocoarctation with saccular aneurysms, left sided SVC and aberrant right subclavian artery - A case report

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ABSTRACT

Pseudocoarctaion is a rare congenital anomaly due to elongation of aortic arch. The exact etiology is still uncertain. It may be associated with other congenital cardiac and vascular anomalies. We report an unusual case of pseudocarctation associated with aberrant right subclavian artery, left SVC and multiple saccular aneurysms in the kinked arch and we feel that this is the first documented case in literature.

CASE REPORT

INTRODUCTION

Pseudocoarctation of aorta is defined as kinking of aorta with normal blood flow dynamics [1, 2, 3]. It is usually detected due to abnormal mass seen on chest radiograph performed for other reasons.

CASE REPORT

35-year-old female presented with 3 weeks history of productive cough. Chest radiograph demonstrated a superior mediastinal mass. (Figure 1) She was normotensive with no radio-femoral delay. This was investigated further with a CT (Toshiba Aquilion 64, 100ml of omnipaque 350 (contrast) using bolus trigger on descending aorta). CT revealed elongated, kinked and high riding aortic arch in keeping with pseudocoarctation of aorta. (Figure 2) In our case, there were further exceptional associations with aberrant right subclavian artery and left SVC draining into coronary sinus (Figures 3, 4, 5). The right SVC drained into the right atrium. There were no collaterals or rib notching. Multiple saccular aneurysms were also demonstrated in the aortic arch (Figures 6, 7).

DISCUSSION

Dotter and Rob described pseudocoarctation in 1951 [1, 3]. Unlike coarctation, pseudocoarctation does not affect the blood flow and is not associated with collaterals. (Table 1) The aortic arch is higher than normal and this may mimic as a superior mediastinal mass as in our case [4, 5].

Pseudocoarctation is thought to be due to failure of normal regression of the third through the seventh segments of the dorsal aortic roots and the fourth arch segment [6]. The resultant vascular anomalies can be exquisitely demonstrated on CT angiogram. CT findings, which are suggestive of pseudocoarctation, include high riding and kinked aortic arch, isthmic portion of aortic arch is surrounded by aerated lung

and is ventral to the spine and the origin of left subclavian artery is more caudal. [7, 8]

Aberrant right subclavian artery is the last branch of aortic arch. The major vessels of the aorta are derived from dorsal aortic roots. During embryonic life there are 6 paired aortic arches which develop from the dorsal aortic roots. Some of these eventually form the major branches of aorta while others disappear. There are seven pairs of intersegmental arteries which arise from dorsal aortic roots. The right seventh intersegmental artery and right fourth aortic arch are involved in the formation of right subclavian artery. Aberrant right subclavian artery is a consequence of abnormal regression of the fourth aortic arch. [9]There is no previously documented association with pseudocoarctation.

Left SVC is due to failure of regression of left sinus horn and left anterior cardinal vein [10]. Left subclavian and left internal jugular vein form the left SVC which drains into the right atrium via the coronary sinus. It is lateral to the aortic arch and anterior to the left hilum. The incidence of left superior vena cava is higher in patients with congenital heart anomalies (2.8 - 4%) in comparison to 0.3% in general population. [11]. Left sided SVC is associated with septal defects, coarctation, tetralogy of fallot and anomalous pulmonary venous return. However there is no reported case of association with pseudocoarctation in the literature.

The etiology of pseudocoarctation has not been established with certainty. Although most cases have been described in adults, this has also been reported in children. A congenital theory with accentuation by hypertension and atherosclerosis has been proposed. [12]This theory is supported by high incidence of associated congenital cardiac defects. [12, 13, 14] The other theories which have been proposed include acquired theory and fibroelastosis. [15, 16]

Aneurysm is defined as localised dilatation of an artery containing all three layers, adventitia, media and intima. Thoracic aortic aneurysm in patients who are younger than 40 years are associated with congenital bicuspid valve and connective tissue disorders (Ehler-Danlos and Marfan syndrome). Mortality is due to dissection and rupture. There are few case reports, which describe aneurysms and aneurysmal dilatation of thoracic aorta in pseudocoarctation [17, 18, 19], but there is no previously reported association with multiple saccular aneurysms of the aortic arch. Coexistence of coarctation with pseudocoarctation has been reported however the etiology is probably different as the male predominance is more in coarctation (4:1) than pseudocoarctation (1.5:1)

So, the currently proposed embryologic theory for pseudocoarctation does not explain all the associated anomalies seen in this patient [5, 8].

To the best of our knowledge, there have been no previous reports of pseudocoarctation of the aorta associated with the aberrant right subclavian artery, left SVC and multiple saccular aneurysms in the aortic arch.

TEACHING POINT

Pseudocoarctation is thought to be due to failure of normal regression of embryologic aortic arches and can be associated with other exceptional vascular anomalies such as aberrant right subclavian artery and left SVC.

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FIGURES

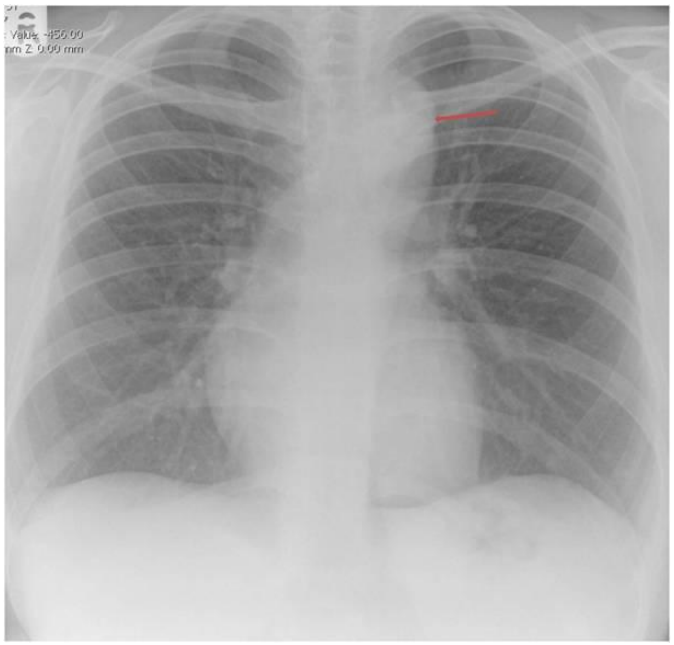

Figure 1. 35 year old female with pseudocoarctation of aorta. Chest radiograph (PA view) showing superior mediastinal widening (red arrow).

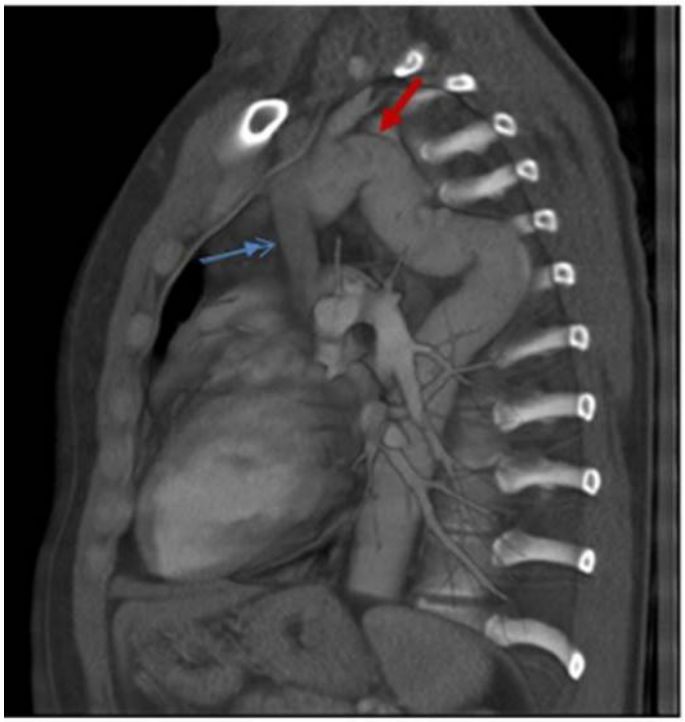

Figure 2. 35 year old female with pseudocoarctation of aorta. Minimum intensity projection sagittal reformatted image shows kinked and high riding aortic arch (red arrow) and left sided SVC (blue arrow).

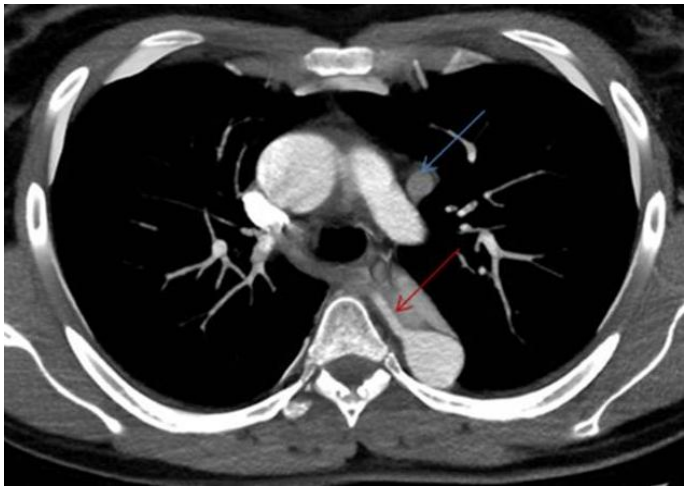

Figure 3. 35 year old female with pseudocoarctation of aorta. MDCT post contrast angiogram axial showing aberrant right subclavian artery (red arrow) and left sided SVC (blue arrow).

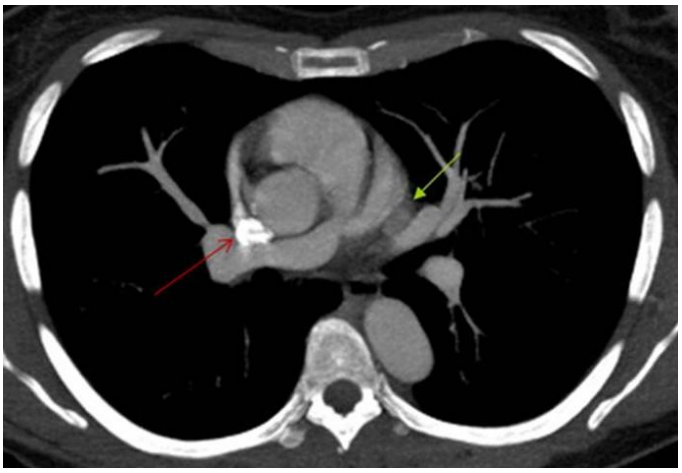

Figure 4. 35 year old female with pseudocoarctation of aorta. MDCT axial shows left SVC entering coronary sinus (green arrow) and right SVC draining into right atrium (red arrow).

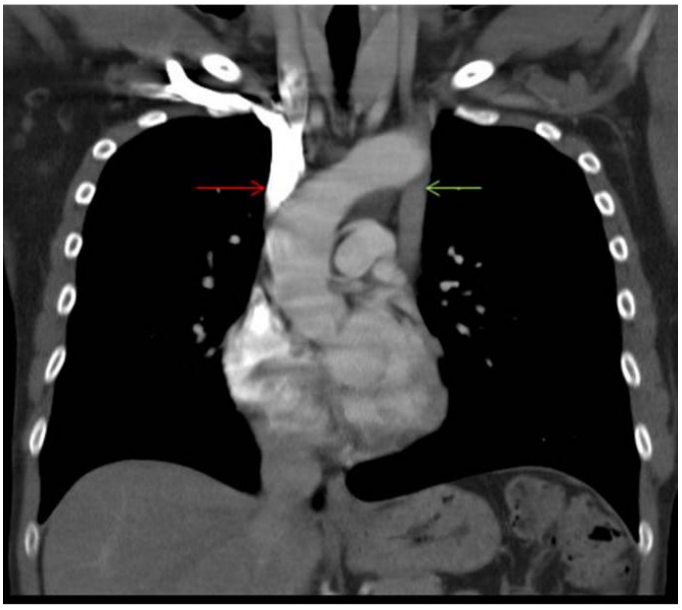

Figure 5. 35 year old female with pseudocoarctation of aorta. Minimum Intensity Projection coronal reformatted image showing left SVC (green arrow) and right SVC (red arrow).

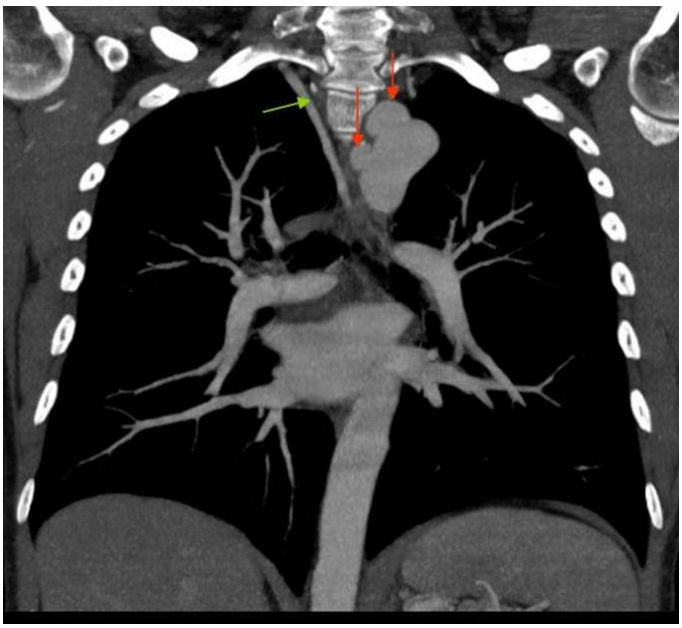

Figure 6. 35 year old female with pseudocoarctation of aorta. Minimum Intensity Projection coronal reformatted image showing 2 saccular aneurysms from aortic arch (red arrows) and aberrant right subclavian artery (green arrow).

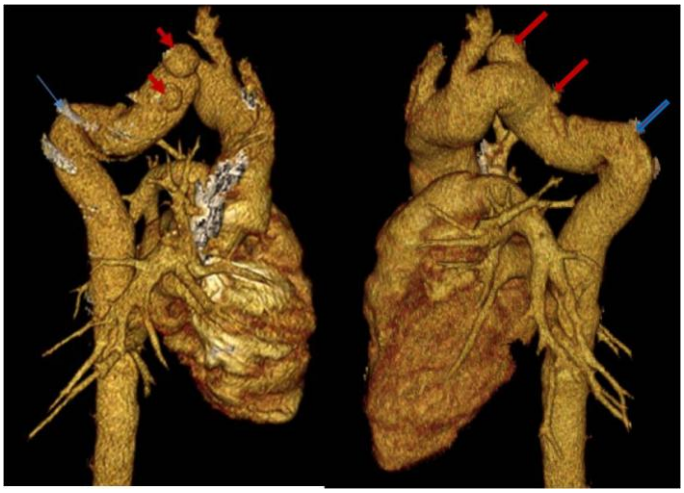

Figure 7. 35 year old female with pseudocoarctation of aorta. Volume rendered 3D reformat right lateral and left lateral showing kink in aortic arch (blue arrow) with multiple saccular pseudoaneurysms. (red arrow)

Coarctation	**Pseudocoarctation**
5-10% of congenital cardiac conditions	Rare (approximately 150 cases reported in literature)
Male: female 4:1	Male: female 1.5:1
Hypertension	Normotensive
Radio-femoral delay	No radio-femoral delay
Alteration of blood flow dynamics	No alteration of blood flow dynamics
Collaterals	No collaterals
Rib Notching	No rib notching

Table 1: Comparison table – Coarctation versus Pseudocoarctation

Right sided SVC	**Left sided SVC**
Drains into right atrium	Drains into coronary sinus
Normal	0.3% of general population
No association with congenital defects	Associated with coarctation and septal defects (incidence (2.8-4.3%)

Table 2: Comparison table – Right sided SVC versus left sided SVC

ABBREVIATIONS

MDCT = Multidetector computed tomography
SVC = Superior vena cava

KEYWORDS

Pseudocoarctation, left SVC, aberrant, subclavian

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