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Rare Branching Variants of Arch of Aorta - A Case Series

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Abstract: ***Aortic arch branching anomalies encompass a wide spectrum of aortic arch malformations due to altered embryogenesis of branchial arches. Although most of these variants are asymptomatic, knowledge about such vascular variations is of utmost importance in the context of operative and interventional procedures of the thorax and head and neck region. Here we describe three rare patterns of aortic arch branching variants which were detected incidentally when contrast enhanced CT scans were acquired for unrelated indications – namely, right sided aortic arch with aberrant left subclavian artery arising from Kommerell diverticulum, truncus bicaroticus with aberrant right subclavian artery and bovine arch with aberrant left vertebral artery from arch of aorta. Knowledge of these anatomical variations can aid in diagnosis of their possible complications, and help predict technical difficulties while planning surgical and interventional procedures.***

Keywords: Aortic arch branching variant, Kommerell diverticulum, Truncus bicaroticus, bovine arch, Left vertebral artery

1. Introduction

Aortic arch develops during 3rd to 8th week of intrauterine life [1]. Altered embryogenesis of branchial arches lead to anomalous branching patterns of the arch of aorta and thus variations in the vessels supplying the brain, head and neck region and upper thorax. Although most of these variants are asymptomatic, knowledge about these variants may be crucial as they may also be associated with vascular rings, other congenital heart diseases, like Tetralogy of Fallot, truncus arteriosus and pulmonary stenosis and chromosomal anomalies and may cause symptoms owing to their pressure effects. They may also pose clinical dilemmas in the context of surgical or interventional procedures of the thorax and head and neck region [2]. When used pre operatively whenever indicated, Contrast enhanced Computer Tomogram (CECT) can detect of such variant branching patterns can help plan surgical or endovascular interventional procedures.

2. Case Presentation

Case 1:
46 yr old female who underwent Contrast enhanced CT neck with upper chest (pre-operative evaluation of multinodular goitre with Bethesda IV nodule with abnormal chest X- ray – Fig 1). The arch of aorta and proximal descending aorta were seen located to the right of midline with mild kinking without significant luminal narrowing. The branches arising from the arch were (from proximal to distal): left common carotid (LCC), right common carotid (RCC), right subclavian (RSCA), and aberrant left subclavian artery (LSCA) originating from Kommerell diverticulum of maximum diameter 2.6cm, arising from the left wall of the arch, passing posterior to the esophagus and trachea, causing minimal indentation (Figure). Echo evaluation was otherwise unremarkable and she successfully underwent total thyroidectomy and was advised follow up. The incidence of such a variant is 0.04 – 0.05% and less than 60 cases have been described in the literature [3], [4].

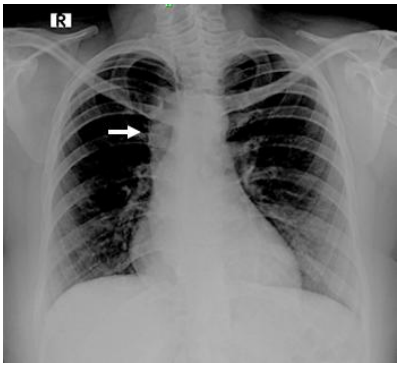


Figure 1: Chest X ray PA view showing right sided aortic arch (arrow)

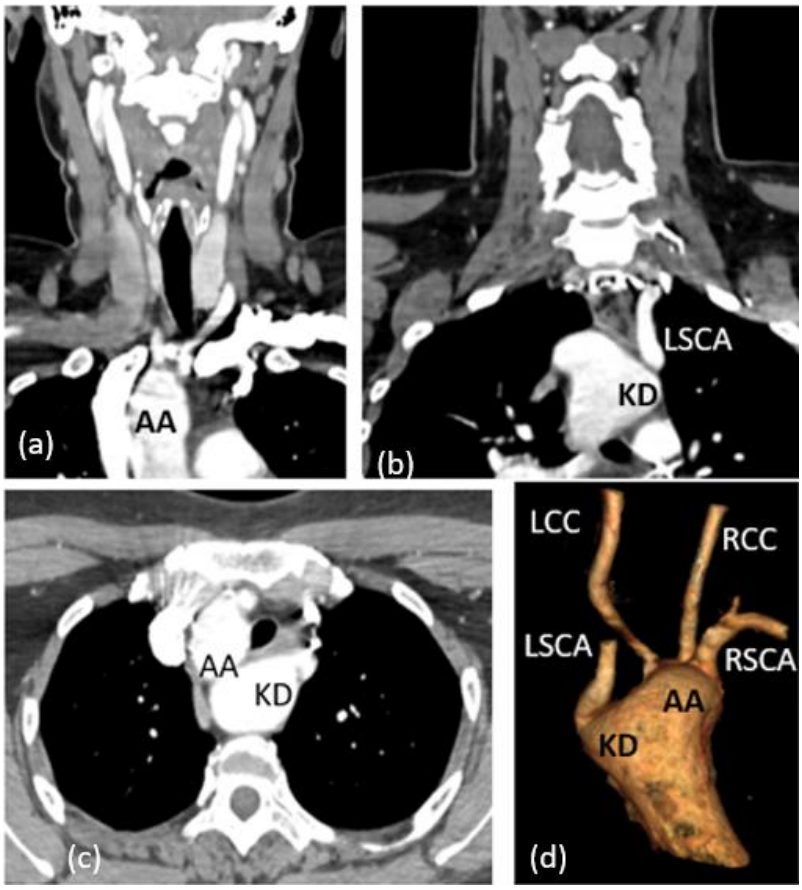
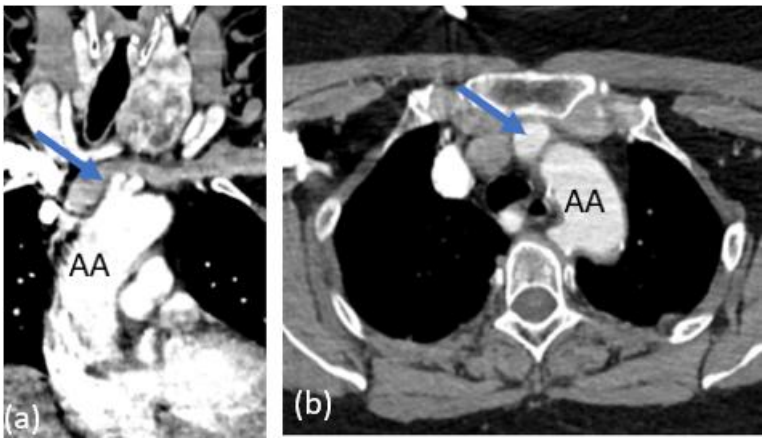


Figure 2: Contrast enhanced CT coronal (a), (b) and axial (c) views and 3D volume rendering image - posterior oblique view (d) demonstrating Right sided aortic arch (AA), Kommerell diverticulum (KD) and origins of right subclavian artery from the anterior aspect of arch (in axial image) and aberrant left subclavian artery from Kommerell diverticulum

Case 2:

68 yr old female who underwent CECT neck with upper chest as pre- operative evaluation of multinodular goitre with Bethesda IV nodules and suspected retrosternal extension. The branching pattern of the aortic arch was as follows. The first branch was a common trunk bifurcating as right and left common carotids (truncus bicaroticus), followed by left subclavian artery and lastly aberrant origin of right subclavian artery from the postero-medial aspect of arch, coursing posterior to esophagus before ascending into the upper limb (Fig 3). Minimal focal compression of esophagus was also seen. Such branching pattern is seen in approximately 0.16 – 0.3% of population [5], [6], [7]. Other investigations including echo were normal and patient underwent an uneventful surgery and recovery.



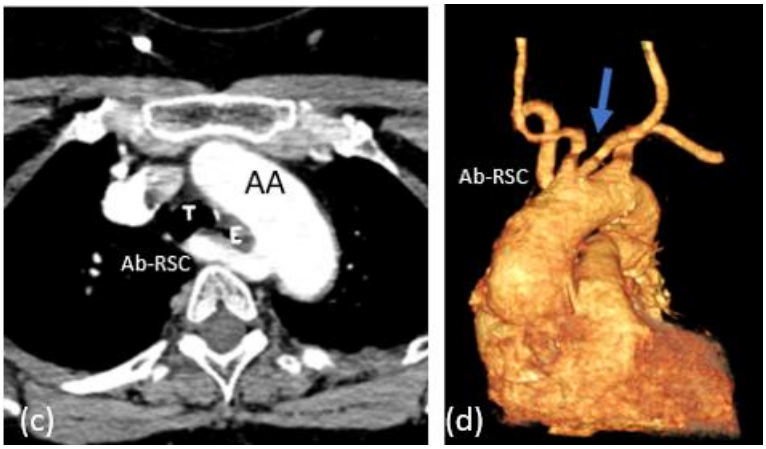


Figure 3: CECT coronal (a) and axial images (b), (c) and 3D volume rendering image - anterior oblique view (d) demonstrating arch of aorta (AA); truncus bicaroticus(arrow); Aberrant right subclavian artery (Ab RSC); trachea (T); esophagus (E)

Case 3:

85 yr old male, with history of hypertension and ischemic heart disease and known case of abdominal aortic aneurysm on follow up, presented with bleeding rectal polyp and underwent aortogram for aortic aneurysm follow up. A common trunk branching off as brachiocephalic trunk on the right side and left common carotid artery was the first branch of arch of aorta. The brachiocephalic trunk later branched into right subclavian and right common carotid arteries. Next, the left vertebral artery was seen arising directly from the arch of aorta immediately followed by origin of left subclavian artery (Fig 4). The abdominal aortic aneurysm showed no interval changes. Other investigations including echo were unremarkable. He underwent snare polypectomy for bleeding rectal polyp; and was advised follow up as size of the diverticulum was < 3cm and he was asymptomatic. The incidence of such branching pattern is 0.12 – 0.3% [8], [9], [10].

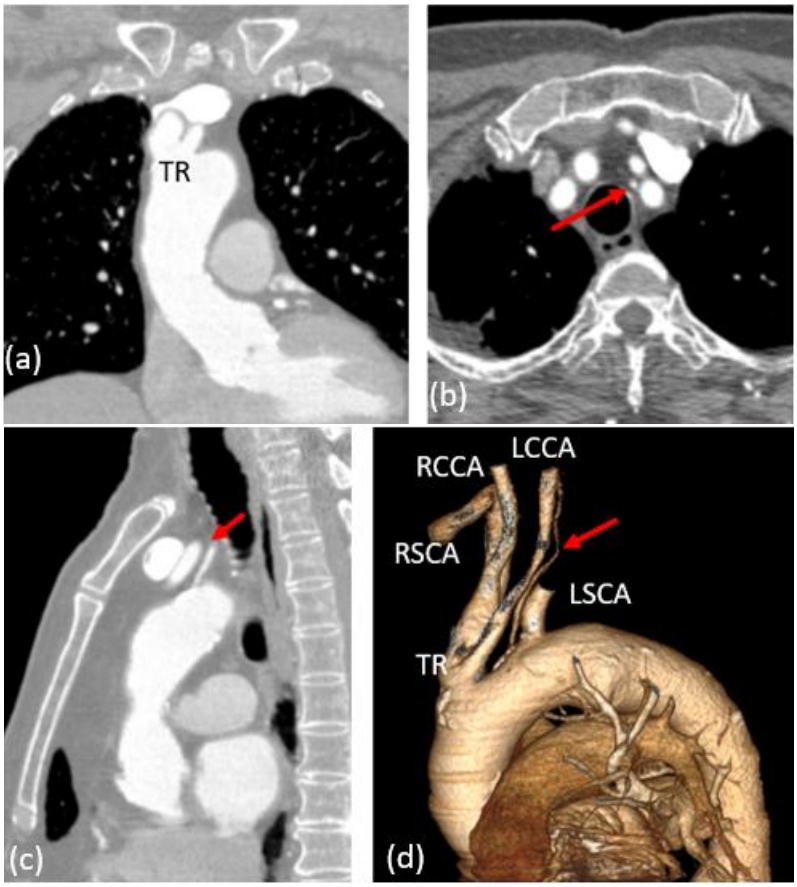


Figure 3: CECT coronal and axial images and 3D volume rendering image (anterior oblique view) demonstrating common trunk (TR) for brachiocephalic trunk and left common carotid arteries; Left vertebral artery originating from arch (arrow)

3. Discussion

Aortic arch and its branches develop from six pairs of pharyngeal arches between 3rd to 8th week of intrauterine life. The most common (80%) branching pattern of aortic arch consists of the brachiocephalic trunk, which then branches into the right subclavian artery and right common carotid artery, followed by the left common carotid artery and finally the left subclavian artery in that order from right to left [2]. In the present-day scenario, aortic arch anomalies are detected and evaluated commonly using echocardiography, CT and MR angiography [1]. CECT being easily available and widely used, may be the most likely modality which can first detect these anatomic variations; it can visualize even small vessels and provide relevant anatomic details as well. Incidental detection in CECT of these variants may suggest higher incidence than those suggested by literature. These variants may remain undetected or overlooked, but may cause symptoms owing to mechanical effects like dyspnoea, dysphagia and cough. Their clinical relevance apart from those due to pressure symptoms are summarised in table -1. Even though, bovine arch and truncus bicaroticus when present alone were considered benign normal variants, it is now believed that they will lead to increased pressure and stress in the left vertebral and subclavian arteries and descending thoracic aorta, ultimately leading to aneurysm formation. Prompt reporting and describing the anatomy of such variations is crucial for follow up and assessing possible complications [10], [11].

Table 1: Incidence and clinical relevance of aortic arch branching variants

Variant	Incidence	Associations and clinical implications
Aberrant right subclavian artery [4]	0.5 – 2 %	Rarely other congenital cardiovascular anomalies, Trisomy 21, non-recurrent right inferior laryngeal nerve Risk during esophagectomy,
Right sided arch with Kommerell diverticulum [3]	0.04 – 0.05 %	Early atherosclerosis, Aortic aneurysm (dyspnoea, dysphagia, cough)
Bovine arch [9]	8 – 25%	Difficulty encountered in carotid stenting via femoral approach Associated aortic dilatation, dissection or rupture, ischemic complications during surgery
Left vertebral artery arising from arch [3]	6 - 13%	Vertebral artery dissection (rare) Warrants extra caution during cervical spine surgery – cerebral ischemia
Bovine arch with left vertebral from arch [10]	0.12 – 0.3%	Brachiocephalic trunk may be deviated to right – injury during tracheostomy
Truncus bicaroticus [8]	5 - 10 %	Thoracic aortic aneurysm
Truncus bicaroticus with aberrant right subclavian artery [7]	0.16- 0.3%	Tetralogy of Fallot, DiGeorge syndrome, trisomy 13, trisomy 18, trisomy 2, oesophageal atresia

4. Conclusion

Contrast enhanced CT imaging provides a non- invasive acceptable opportunity for detection and characterization of aortic arch branching variants even in asymptomatic patients.

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