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An asymptomatic double aortic arch with separate right vertebral artery and left subclavian artery originating from Kommerell Diverticulum combined with congenital asplenia and absence of celiac trunk

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Abstract

This report describes the unique case of a completely patent Double Aortic Arch (DAA) combined with Kommerell Diverticulum (KD), absence of celiac trunk and congenital asplenia. The anatomical variants described were identified and assessed in a 51-year old female from a computed-tomography angiography (CTA) images with 3D-volume rendered reconstructions during her follow-up after hysterectomy. The reconstructed CTA images showed a DAA with the left common carotid artery stemming from the hypoplastic left aortic

arch while the left subclavian artery originated from a KD in the descending thoracic aorta. A symmetric arrangement of the aortic arch branches was demonstrated, comprising a full vascular ring. Since the patient had been completely asymptomatic and with no symptoms of compression of the esophagus or trachea, no surgical management was advised. The abdomen CTA imaging revealed absence of the celiac trunk with direct origin of the common hepatic and the left gastric artery from the superior mesenteric artery as well as asplenia. We presented a case of asymptomatic DAA of completely patent arches with the right vertebral artery branching separately of and the left SCA originating from KD in the descending aorta. The term KD can be identified also in other arch anomalies than the one originally described. Since anatomical anomalies can be frequently combined, thorough imaging inspection with CTA of both thorax and abdomen is suggested.

Keywords: gross anatomy, vascular rings, double aortic arch, aortic congenital anomalies, anatomical variations

INTRODUCTION

Aortic arch anomalies can result either from the persistence of two completely patent aortic arches (Double Aortic Arch, DAA) or the presence of the anatomical continuity of both with an atretic segment in one of them (usually present as a fibrous band or ligament) [1,2]. The variations emerging during the development of the aortic arch can lead a wide variety of anomalies, the acknowledge of which has a significant meaning not only or for cardiothoracic surgery but also for the rapidly emerging field of complex endovascular aortic surgery.

The commonest aortic arch variations include: right aortic arch with aberrant left subclavian artery (SCA) originating from a Kommerell diverticulum (KD), left arch with aberrant right SCA stemming from KD, double aortic arch (DAA) with mirror imaging of the great vessels, right-sided aortic arch (RAA) with mirror imaging of the great vessels, right arch with aberrant left innominate artery off the KD and circumflex aorta [1,2].

Although DAA has been associated mainly with intracardiac malformations, the literature lacks reports DAA with other vascular and extravascular anomalies. We present a case of asymptomatic DAA forming a vascular ring in an adult female combined with absence of

celiac trunk and congenital asplenia. The need to scan for concurrent anatomic anomalies when detecting initially an aortic one is discussed.

CASE PRESENTATION

We illustrate the case of an asymptomatic 51-year old female who underwent a computed-tomography angiography (CTA) as a standard follow-up control after hysterectomy. All CTA imaging examinations were conducted in a 128-slice CT scanner (PHILIPS INGENUITY CORE 128) with spiral technique of 0.625mm thickness after intravenous injection of contrast-agent. The CT acquisition parameters were prescribed as follows: 268 mAs, 120 kVp, 5sec scan time, 22.1 mm feed/rotation ration, 2mm slice thickness, 0.75–1.5 mm reconstruction spacing/ increment, 0.5 mm slice overlap, and a 512x512 image matrix size. CT angiography was triggered at 150 Hounsfield units.

The chest X-ray had showed a RAA (Figure 1A, arrow) with the Computed Tomography revealing mild deviation of the trachea and the esophagus (Figure 1B,C). A symmetric encirclement of the trachea by vascular structures was revealed (Figure 1D) causing no strictures or symptoms. Her cardiac examination excluded any cardiac anomalies (and, specifically, intracardiac defects) while her past medical history documented hysterectomy for endometrial cancer and inflammatory mesenteric panniculitis managed with corticosteroids.

The CTA revealed absence of splenic artery (congenital, since there was no history of trauma or splenectomy) (Figure 2A) and direct origin of the common hepatic artery (CHA) and the left gastric artery (LGA) from the superior mesenteric artery (SMA) (Figure 2B). Moreover, the 3D volume rendering view (Figure 3A) showed a morphology accordant with a DAA with the left common carotid artery (CCA) of 5mm stemming from the hypoplastic left aortic arch of 6.5mm while the left SCA originated from a 12mmx16mm KD (Figure 3B). Moreover, a right isolated right vertebral artery (VA) was documented in a 3D volume rendering view of the superior thoracic inlet (Figure 3C). The left common carotid artery (CCA), was tethered and deflected posteriorly (red arrow) to approximate the left SCA as better depicted (Figure 3C). A symmetric arrangement of the aortic arch branches was demonstrated in the three-dimensional (3D) volume-rendered images, comprising a full vascular ring. Since the patient had been completely asymptomatic and with no symptoms of

compression of the esophagus or trachea, no surgical management was advised. The patient is followed-up on an annual basis.

DISCUSSION

Vascular rings occur in less than 1% of all cardiovascular malformations and can cause symptoms related the trachea and esophagus compression, being diagnosed very early in life (infants and childhood) [1,2]. After conducting routine first trimester ultrasound examination in 33.202 pregnancies, Vigneswaran et al. identified 5.4 and 1.5 per 10.000 cases of isolated RAA cases and DAA, respectively, comprising a 2-3 fold higher incidence than that reported in postantal studies [3].

Embryologically, the paired dorsal and ventral aorta are connected by six paired branchial arches, the development of which begins by the second week of gestation and continues till the seventh week [4]. All of these arches are not present at the same time, but appear and undergo regression sequentially from cranial to caudal direction. The first, second, and the fifth arches involute while the third, fourth and the sixth form the aortic branches. The third arch forms common carotid and a segment of internal carotid arteries while the fourth arch forms the definitive aortic arch. The ventral portion of the sixth arch forms the distal segments of pulmonary artery while the dorsal portion of the arch gives the ductii arteriosus. The ventral aorta forms truncus arteriosus dividing later into the ascending aorta and the proximal segment of main pulmonary artery. The paired dorsal aorta fuses to form a single trunk. The seventh intersegmental artery (branch of dorsal aorta) forms a part of the subclavian arteries. Normal left aortic arch is formed by the regression of the right aortic arch, right sided ductus and right dorsal aorta. The proximal portion of right dorsal aorta forms part of right subclavian artery. The left dorsal aorta forms the distal aortic arch and the descending thoracic aorta [4].

DAA results from persistence of both the left and right fourth embryonic aortic arches and is implicated in nearly half of the symptomatic cases. Three morphologies of DAA are recognized according to the dominance of the right or the left arch or their equivalence. Moreover, the ligamentum arteriosus running between the aorta and the left pulmonary artery is an additional factor that can cause compression to the trachea [5].

The term KD was initially introduced to describe an outpouching or dilatation of the proximal portion of an aberrant right SCA near its origin from a left aortic arch or the

descending aorta with the diameter of the aberrant artery near its origin measuring twice the size of its distal diameter. Likewise, the term has been also used to describe the bulge in a RAA where the aberrant analogue (i.e., left SCA) branches from. Yu et al. described also the origin of a “nonaberrant-aberrant” left SCA arising from a KD located on the left side of a LAA, while Komiyama and Yasui documented the origin of a left SCA from a KD located on the right side of a LAA [6,7]. Therefore, KD is used to describe a specific form of outpouching met also in normal arches which is associated with an anomaly in the SCA development. It is not generally appreciated that patients with vascular rings due to DAA can also have a KD, since “aberrant” arteries do not -by definition- exist in this anomaly [8]. However, in current clinical practice the KD term has been also adapted to describe a relevant bulge accompanying -less commonly- DAA [6].

DAA can be associated with segmental atresia of one of its arches; while the location of the atretic part either between the descending thoracic aorta and the left ligamentum arteriosus or between the latter and the SCA artery are mentioned commonly, the atresia between the left subclavian artery and the left CCA (i.e., the “C” segment) is reported extremely rare [9]. Interestingly, it seems prudent to recognize that modern advanced imaging techniques can identify cases of incomplete DAA which would be otherwise angiographically mistaken previously as RAA with aberrant left SCA, lowering the index of suspicion for KD in patients with DAA [5,10-14]. Modern documentation of these variations should acknowledge the difference between a DAA with atretic left dorsal aortic arch from a normal mirror-image RAA, where the left dorsal aortic arch has been interrupted [15].

The presence of a KD in our case is rather unusual, since KD has been associated with RAA with dorsal left arch atresia or with aberrant left SCA [5,15]. Backer et al. have recently addressed the presence of KD in a series of operated patients with symptomatic DAA; yet, all the distal left aortic arch was atretic in all patients [5]. The presence of the particular variant in our case cannot be explained based on the existing proposed schemes in the literature.

Since the patient was asymptomatic no surgical management was undertaken. DAA can be asymptomatic and, therefore, documented for the first time in the elderly life [11-13]. Occasionally, they can present late symptoms owing to atherosclerotic changes of the aorta (e.g. becoming tortuous or aneurismal), age-related changes of the chest or further dilatation of the KD. Some authors suggest a KD >1.5 times the aberrant left SCA origin as an operative indication for primary resection [5,16].

DAA can be accompanied also by anomalies in its branches with respect to their morphology or origin [14]. Our case demonstrates separate take-off of the right CCA, SCA and VA. Embryologically, involution of the 1^{st}-5^{th} cervical intersegmental arteries with persistence of the 6^{th} and 7^{th} and involution of longitudinal connection between these led to separate origin of the right SCA [17]. Moreover, the absence of an innominate artery is anticipated since both aortic arches have been preserved with maintenance of the dorsal segments between the 7^{th} cervical intersegmental artery and the descending aorta.

The existence of DDA in our case was combined with congenital asplenia and absence of the celiac trunk; rather, the common hepatic and left gastic artery originated directly from the superior mesenteric artery while the inferior mesenteric artery branched normally from aorta. The celiac trunk is absent in 0.1-4% of cases [18-20]. The celiac axis, the superior mesenteric artery and their major aortic branches frequently show diverse anomalies in their origins. Wang et al. classified systematically complex variant origins of the celiac axis, the superior mesenteric artery and their major branches based on a retrospective evaluation of 1.500 abdominal multidetector CTA images, identifying a hepatomesenteric trunk in 4.47% of cases [20].

Notably, Natsis et al. identified eight types of aortic arch variations with respect to the origins and anatomical relations of the vessels. The generated symptoms can include dyspnea, dysphagia but also misinterpretation of radiological examinations and complications during neck and thorax surgery [21]. More interestingly, the Authors underline that such variations are frequently accompanied by other congenital abnormalities [21]. Indeed, it is our belief that when a “single” vascular anomaly is identified -whether symptomatic or not- the patient should be also examined for other anomalies, not exclusively detected in the vascular system. This tack is nowadays significantly facilitated by the evolved, sophisticated and easily utilized imaging modalities providing detailed anatomic information. Although DAA has been reported to accompany intracardiac anomalies, its association with asplenia has been reported only scarcely whereas the particular combination in our case (i.e., DAA with asplenia and absence of celiac trunk) has been never reported [22].

3D volume-rendered imaging modalities not only provide thorough morphological inspection of anatomical variations and anomalies of the aortic arch but also enable preoperative planning in the modern era of staged or hybrid endovascular treatments where the detailed mapping of the size, origin sites and take-off angles of aortic branches are of paramount importance [23-25]. Besides, the incidence of aortic arch variants such as bovine

aortic arch, isolated left VA and aberrant right SCA is significantly increased in patients undergoing surgical management for thoracic aortic disease; for example, a recent study by Dumfarth et al. recorded a prevalence of 33.5% of arch anomalies among 556 patients operated for thoracic aortic disease when compared with the general population (18.2%) [26]. In this study, the patients with aortic branch variations were significantly younger and needed aortic arch intervention more frequently than patients with normal arch anatomy. Therefore, and atypical branching variants should no longer be considered clinically irrelevant or innocent -let alone aortic arch anomalies or combinations of these entities- but rather markers for thoracic aortic disease.

CONCLUSIONS

We presented a case of asymptomatic DAA with completely patent arches depicting a separate origin of the right VA and the presence of KD in the descending aorta giving rise to the left SCA. The term KD can be applied also in other arch anomalies than the one originally described since these analogue morphologies share the same potential complications and pose the same surgical difficulties when receiving the traditional surgical treatment or modern endovascular options. Since anatomical anomalies can be frequently combined, thorough imaging inspection with CTA of both thorax and abdomen is suggested.

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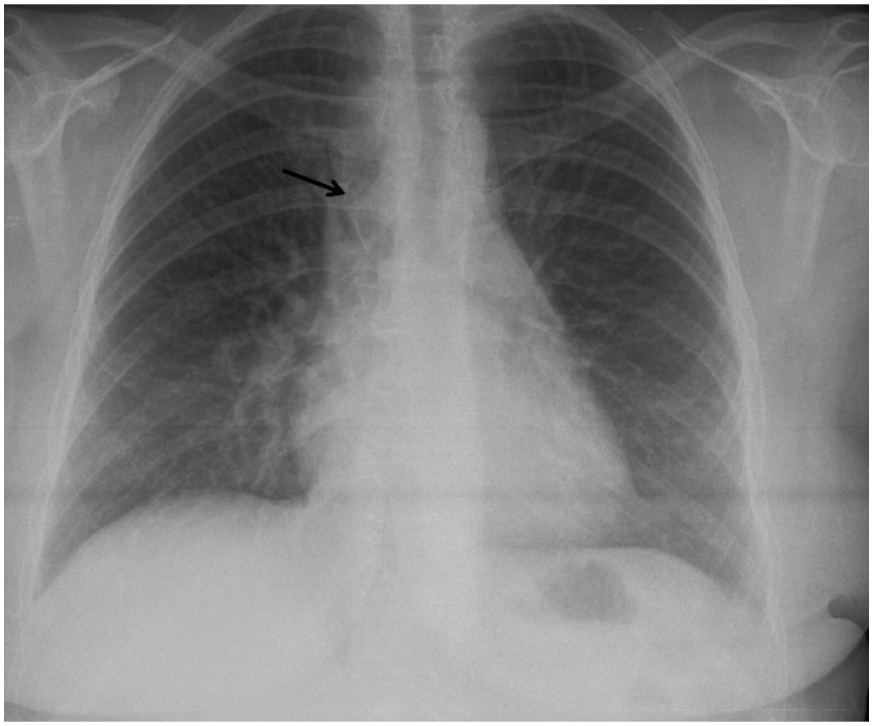
Figure 1. **A**. Chest X-ray showing right-sided aortic arch (*arrow*) with mild deviation of the trachea (**B**) and the esophagus (**C**). Computed tomographic scan showing the trachea encircled by an wide aortic structure on the right and a left sided-vessel in the form of a vascular ring (**D**).

Figure 2. A. Computed tomography showing complete absence of spleen. **B**. Absence of celiac trunk with direct origin of the gastric artery (LGA) and common hepatic artery (CHA) from the superior mesenteric artery (SMA). Note the normal origin of the inferior mesenteric artery (IMA) and the right renal artery (RRA). 1; SMA, 2; CHA, 3; LGA, 4; RRA, 5; IMA.

Figure 3. **A**. 3D reconstruction of the computed tomography angiography shows the left common carotid artery (LCAA) stemming from a rudimentary left aortic arch (LAA). **B**. The left subclavian artery (LSA) originates from Kommerell's diverticulum (KD) and is connected to the LAA. (Note: the clavicles and sternum have been removed)**. C.** While the left vertebral artery (LVA) originates from LSA, the right vertebral artery (RVA) originates separately between the right common carotid artery (RCCA) and the right subclavian artery (RSA). Note: the proximal right clavicle has been removed.

1; LCCA, 2; LAA, 3; LSA, 4; KD, 5; LVA, 6; RCCA, 7; RVA, 8; RSA.



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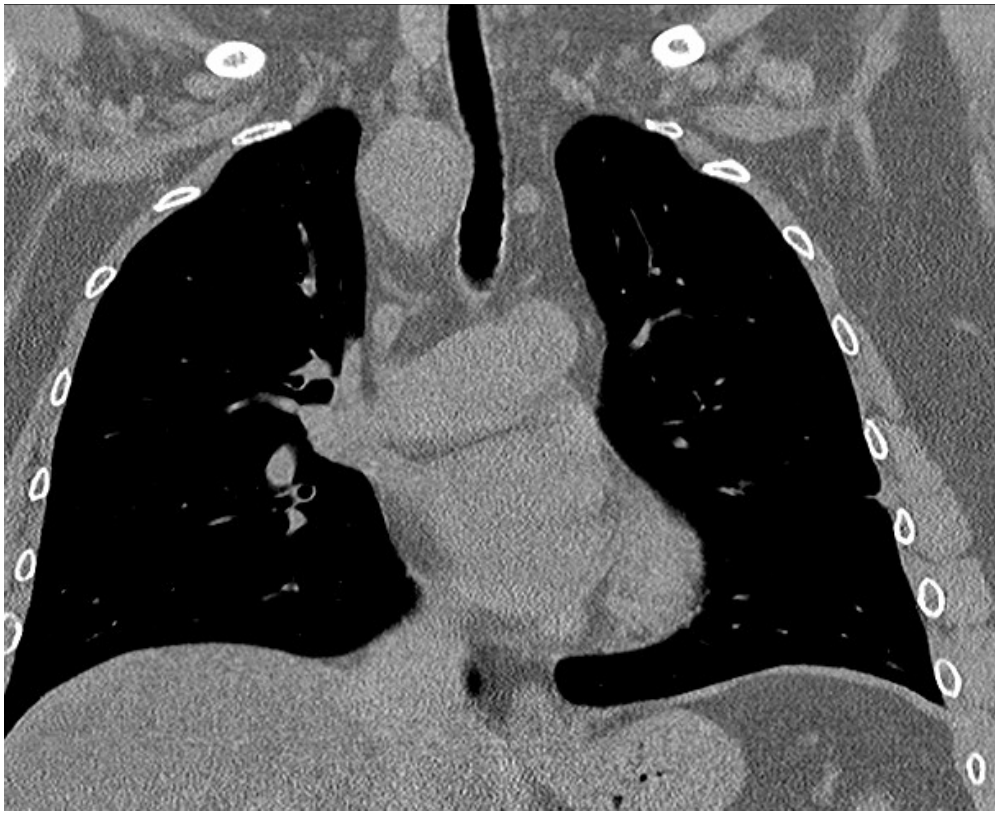

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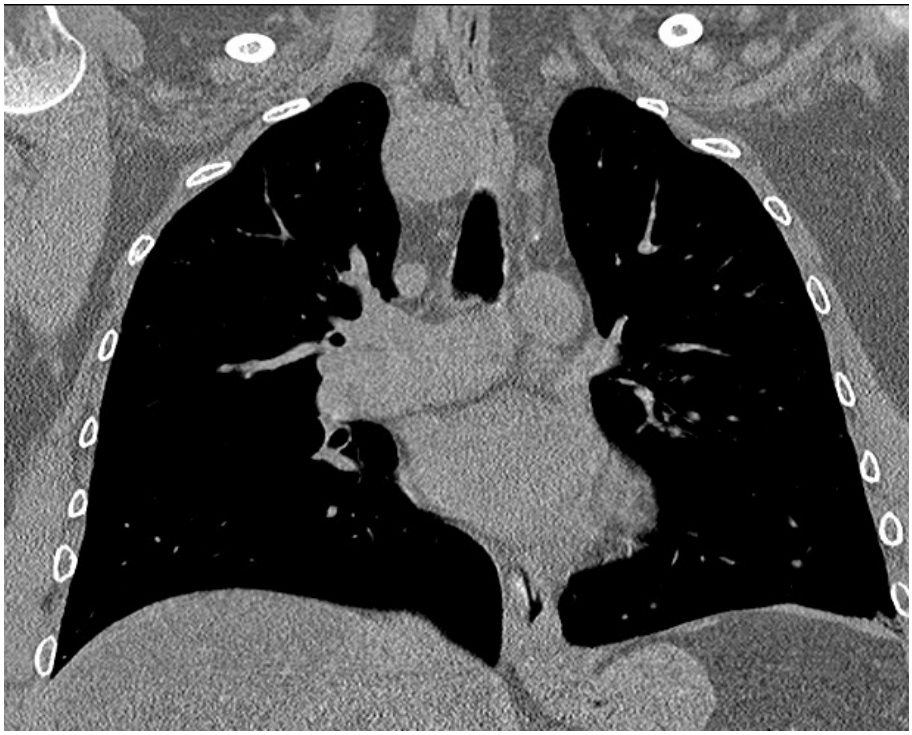

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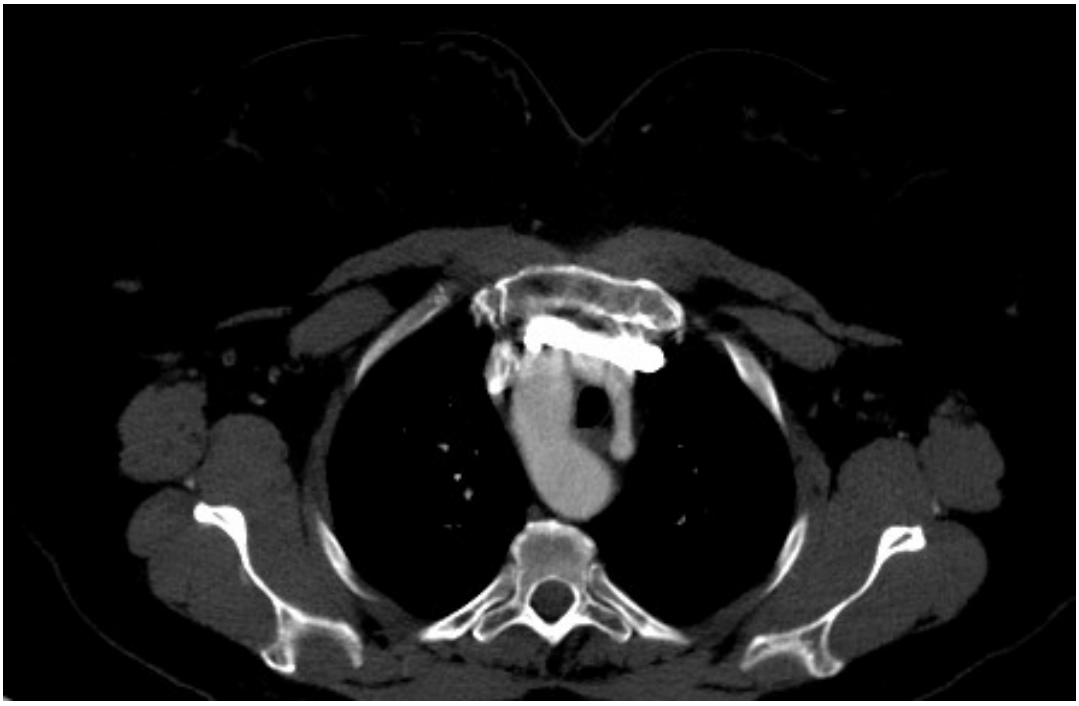


Figure 1D

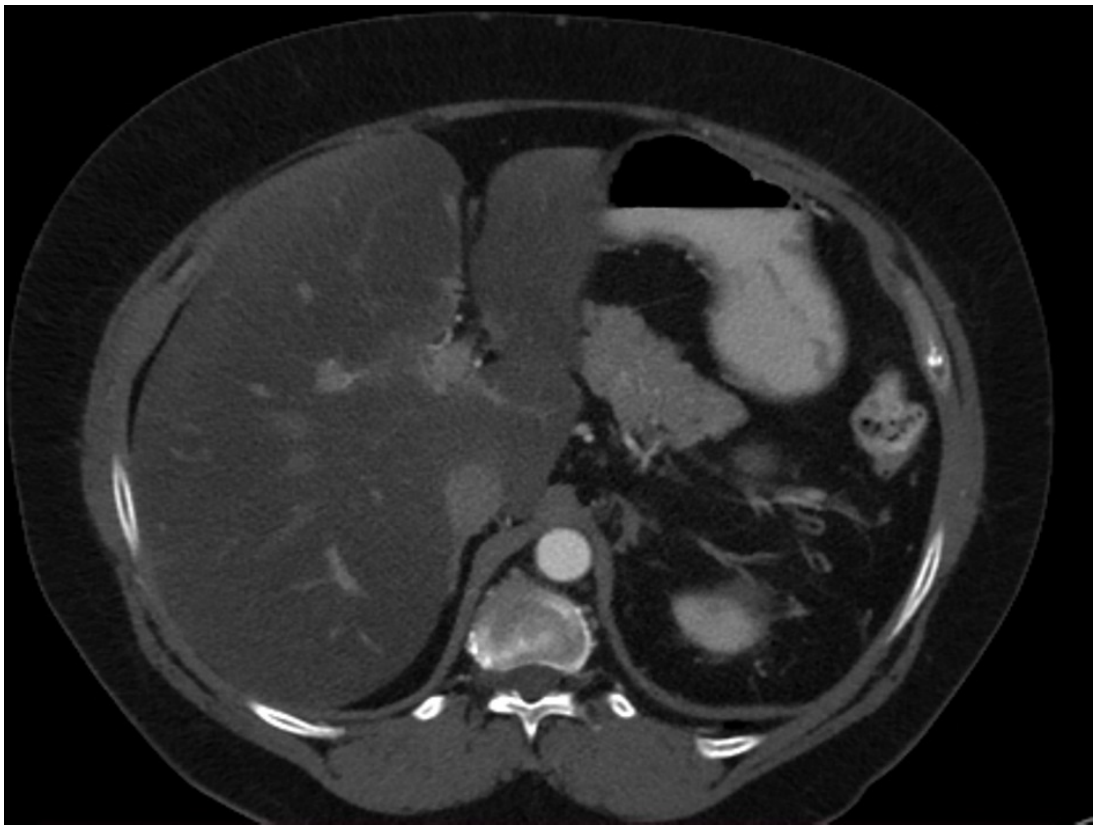
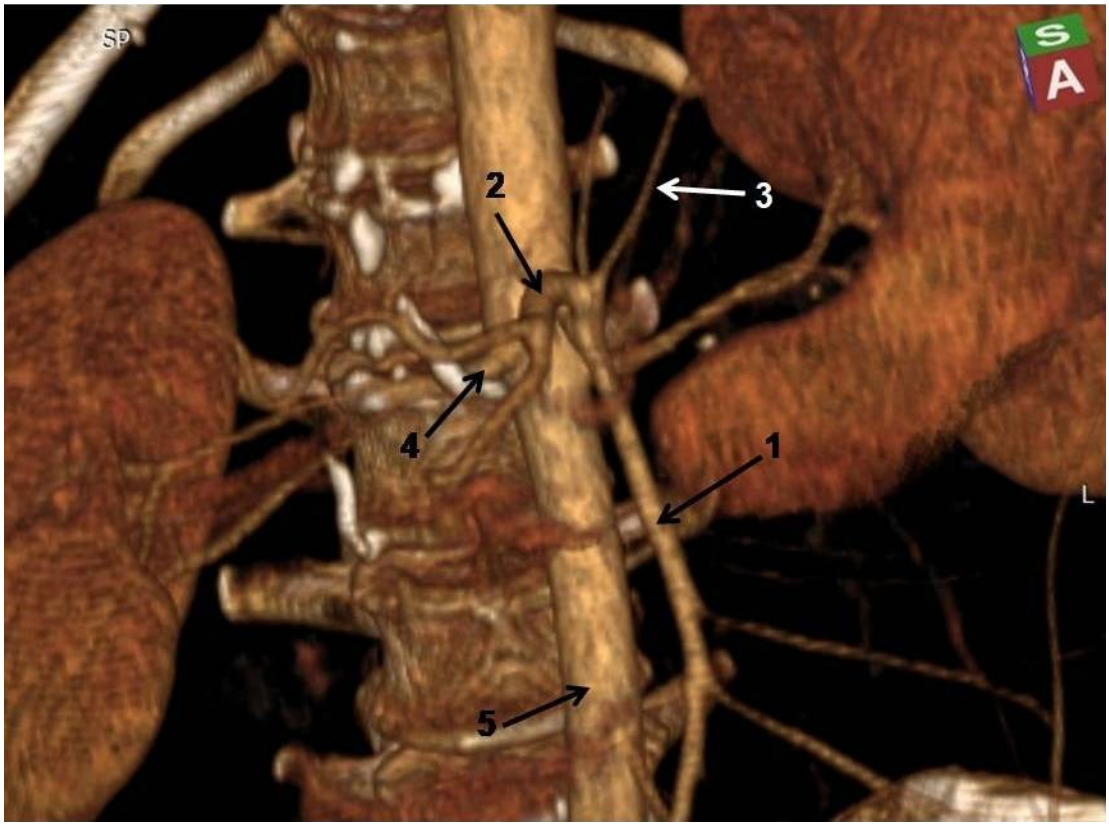


Figure 2A



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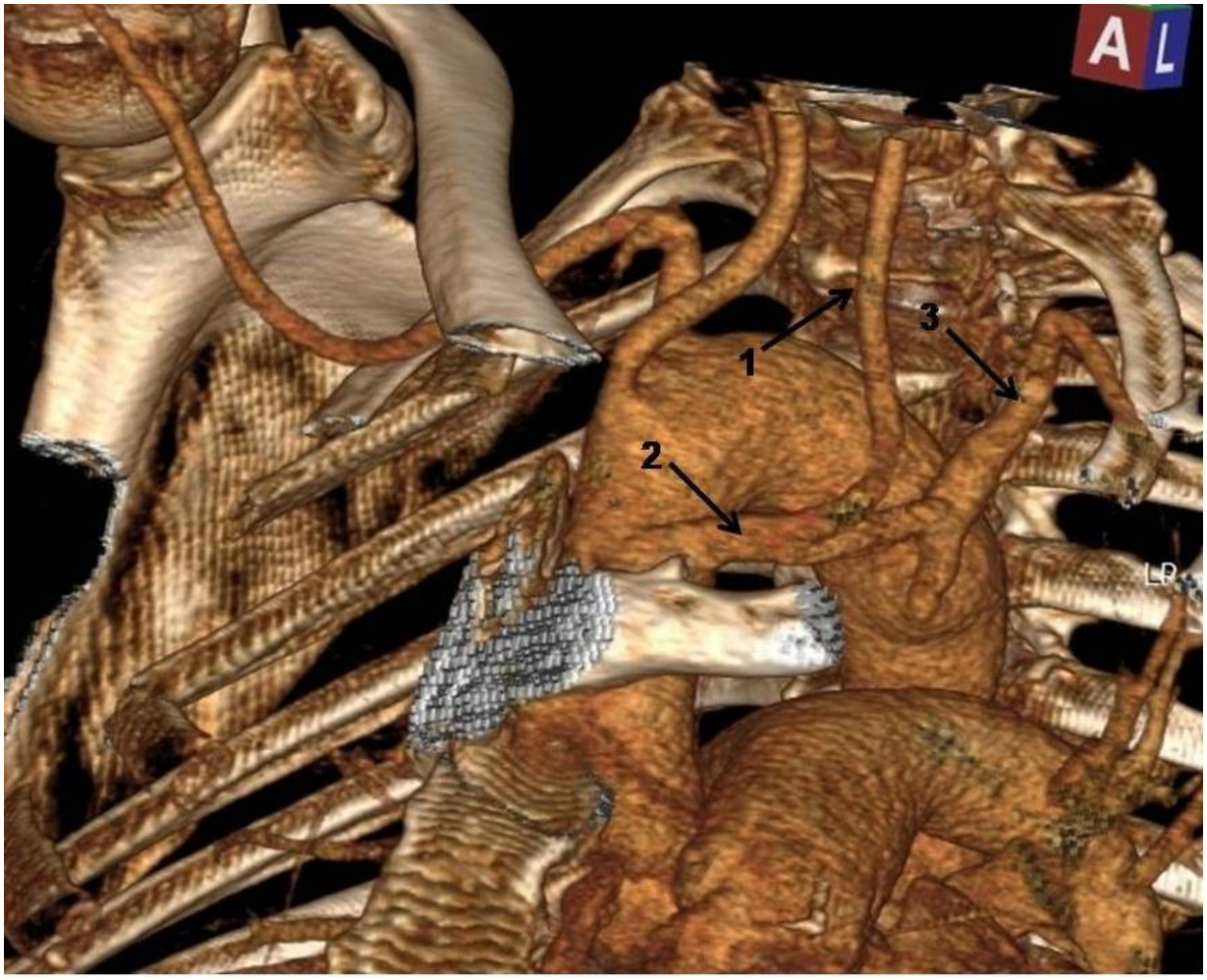

Figure 3A

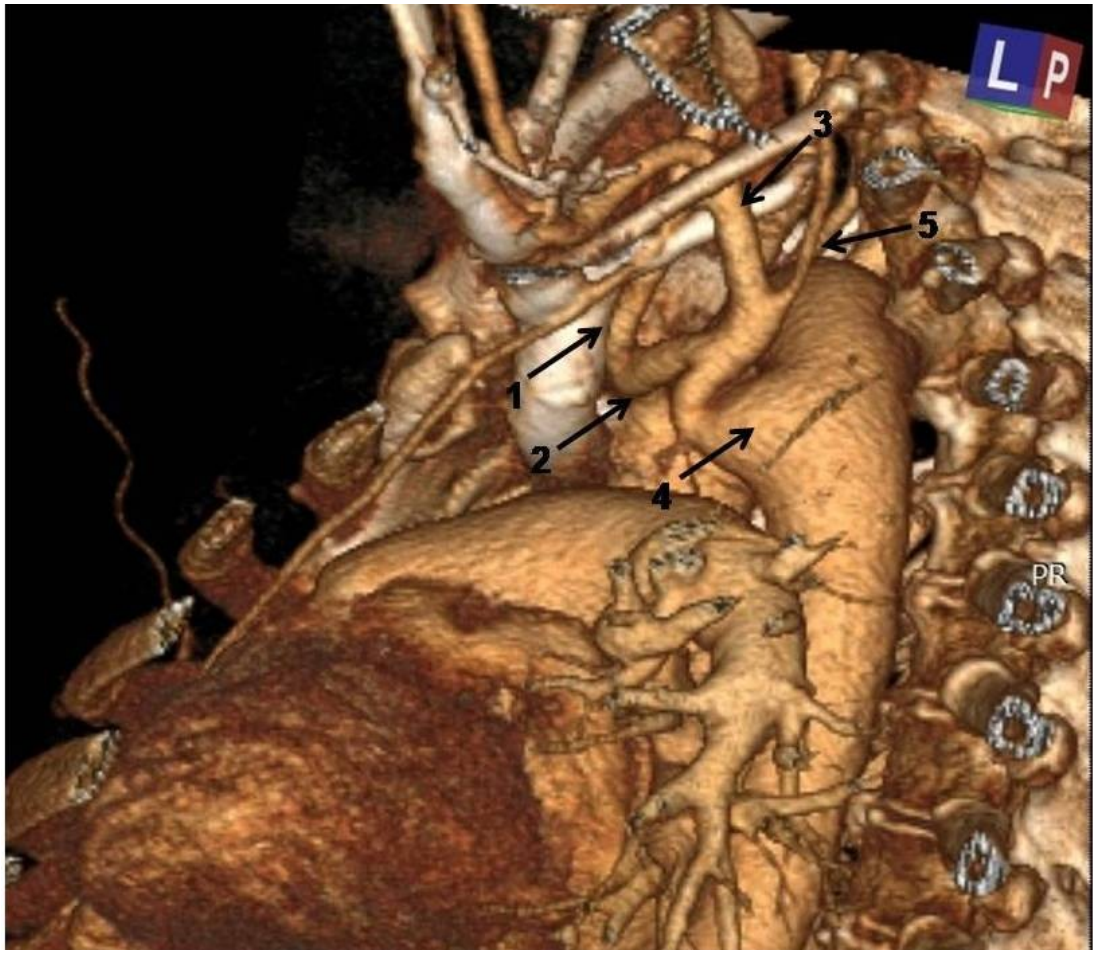

Figure 3B

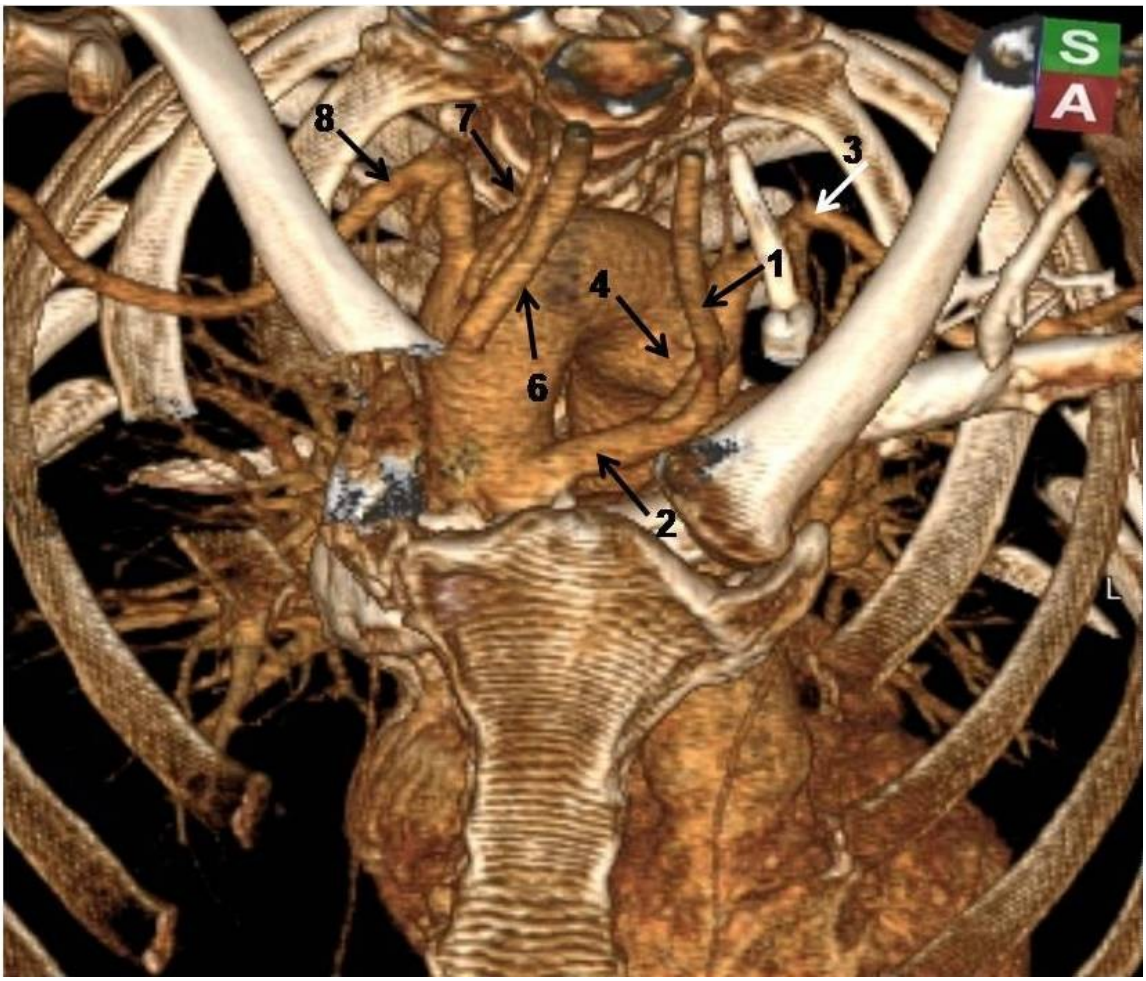

Figure 3C