



## Case Report

# Right-sided aortic arch with complex anomalies presented with transient ischemic attack

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Received : 05 October 2022  
Accepted : 08 October 2022  
Published : 28 October 2022

DOI  
10.25259/SNI\_913\_2022

### Quick Response Code:



## ABSTRACT

**Background:** The right-sided aortic arch (RAA) is an uncommon anatomical anomaly found in <0.1% of the adult population. In this article, we report a case of RAA anomaly with an aberrant left subclavian artery (ALSA) and Kommerell's diverticulum associated with aneurysmal dilation of the ascending aorta, left carotid artery (CCA) stenosis, and pancake kidney presented with a transient ischemic attack (TIA). To the best of our knowledge, this is the first case in the literature that discusses such associations, especially in a symptomatic patient with neurological rather than tracheoesophageal symptoms and in the absence of the steal phenomenon.

**Case Description:** A 52-year-old male, with a history of recurrent multiple TIAs, presented immediately after the onset of blurred vision and left-sided weakness. The initial diagnostic cerebral angiogram revealed a left CCA stenosis of <30%, with normal posterior circulation vasculature. The diagnosis of RAA was made with computed tomography angiography (CTA) of the thoracic and abdominal aorta, which revealed Type 2 RAA, with ALSA, which had a bulbous dilatation at its origin that suggests Kommerell's diverticulum. Another two findings on CTA were a persistent left-sided superior vena cava that ended in the coronary sinus and a single pelvic fused renal mass (Pancake kidney).

**Conclusion:** We presented an extremely rare case of RAA with ALSA associated with a group of extra rare anomalies. Understanding the anatomical variants of RAA and its characteristics is critical to improving the management and follow-up of patients with such anomalies.

**Keywords:** Aberrant left subclavian artery, Kommerell's diverticulum, Pancake kidney, Right-sided aortic arch, Vascular ring

## INTRODUCTION

The right-sided aortic arch (RAA) is an uncommon anatomical anomaly found in <0.1% of the adult population due to the continuation of the right fourth embryologic aortic arch and in-folding of the left aortic arch, with only half of the cases (0.05%) being associated with an aberrant left subclavian artery (ALSA).<sup>[13]</sup>

RAA often exhibits no symptoms and most adulthood diagnoses are unintentional. Dyspnea, coughing, and difficulty swallowing are the major symptoms that appear once Kommerell's diverticulum compresses the surrounding structures.<sup>[22]</sup> The likelihood of cerebral infarction will increase as a result, as cardiac abnormalities constitute at least 20% of the causes of ischemic strokes.<sup>[9,10]</sup>

However, in this article, we report a case of RAA anomaly with an ALSA and Kommerell's diverticulum associated with aneurysmal dilation of the ascending aorta, left common carotid artery (CCA) stenosis of approximately 30–40%, and pancake kidney presented with a transient ischemic attack (TIA). This is the first case in the literature that discusses such associations, especially in a symptomatic patient with neurological rather than tracheoesophageal symptoms with the absence of the steal phenomenon. Furthermore, our experience will provide a helpful report on the neuroendovascular aspect of diagnosing this condition.

## CASE PRESENTATION

A 52-year-old male, with a history of recurrent multiple TIAs, presented immediately after the onset of blurred vision and right-sided weakness, which we confirmed with the physical examination. This patient has no medical history of TIA risk factors. Based on the clinical history and examination, a left internal carotid artery (ICA) stenosis was suspected and the patient was admitted for immediate therapeutic cerebral catheter angiography.

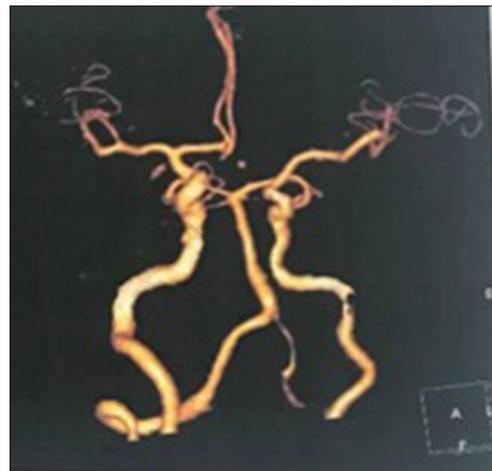
The initial diagnostic cerebral angiogram revealed a left CCA stenosis of <30%, with normal posterior circulation vasculature. However, we could not examine the anterior circulation of the brain due to technical difficulties, which we will discuss later. As a result, the therapeutic catheterization was discontinued and medical treatment was warranted. A computed tomography angiography (CTA) of the head, neck, chest, and abdomen was recommended for a more thorough examination.

The native CT study of the brain after 45 days of catheterization showed signs of watershed infarction involving the right posterior frontal and parieto-occipital cortex. The CTA confirmed diffuse stenosis of nearly 30–50% in the left CCA and ICA, involving the entire vessel length, with diffuse circumferential mural thickening. However, a segment of mild narrowing (nearly 30%) was seen involving the right CCA measuring 30 mm in length, with a normal diameter of the right ICA. The circle of Willis was normal with a hypoplastic left A1 segment of the anterior cerebral artery with no vessel wall irregularities [Figure 1].

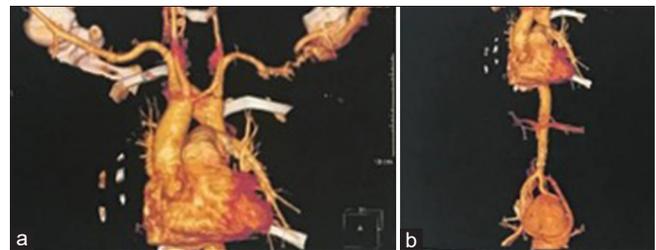
The diagnosis of RAA was made with CTA of the thoracic and abdominal aorta, which revealed Type 2 RAA, with ALSA, which had a bullous dilatation at its origin that

suggests Kommerell's diverticulum [Figure 2]. The RAA was lying on the right side of the trachea and esophagus, with its branches being in the following order: the left CCA first, followed by the right CCA, the right SCA, and finally, the ALSA. The course of the ALSA was posterior to the trachea and esophagus. However, no definite evidence of tracheal or esophageal compression was noted in the CTA. There were no cardiac abnormalities. The ascending aorta was normal in diameter with no wall irregularities. However, the root was ectatic with a diameter of 32 mm at the annulus and 5 cm at the aortic sinus, suggesting an aortic root aneurysm. The descending aorta was mildly tortuous with a near midline position and positioned to the right mildly. However, atherosclerotic thickenings were seen involving the descending thoracic and abdominal aorta and iliac arteries.

Another two unexpected findings on CTA were a persistent left-sided superior vena cava (PLSVC) that ended in the



**Figure 1:** Computed tomographic angiography of the head-and-neck arteries (Lateral oblique view) shows diffuse stenosis of nearly 30–50% in the left common carotid artery and internal carotid artery.



**Figure 2:** (a) Computed tomography angiography (CTA) of the thoracic aorta (anteroposterior view) revealing the branches of the aortic arch in the following order: left common carotid artery, right common carotid artery, right subclavian artery, and finally, the aberrant left subclavian artery with aneurysmal origin (Kommerell's diverticulum). (b) CTA of the abdominal aorta (anteroposterior view) shows the pancake kidney.

coronary sinus, then to the right atrium. A single pelvic fused renal mass (Pancake kidney) was discovered at the lower end plate of the L4 to S3 vertebrae, with a single ureter terminating into the left vesicoureteral junction. Moreover, a single renal artery arises from the right common iliac artery and divides later into two major arteries.

### Technical difficulties during diagnostic cerebral catheterization

Highlighting the endovascular difficulties is critical since the atypical anatomy will affect the entire procedure, from increased operation time to failing to complete the examination. This case formulates in a retrograde fashion as we did not recognize the difficulty until after the catheterization had already begun.

The cerebral angiography was performed using a 5-F (Vert; Merit) diagnostic catheter through a right trans-femoral route. Because the anatomical characteristics of a RAA limit catheter navigation, the first attempts to reach any of the main arteries of the Aortic Arch were unsuccessful. As a result, we replaced the (Vert; Merit) catheter with a (SIM1; Merit) catheter, which failed to reach them despite several maneuvers performed by the examiner. Finally, we shifted to the (SIM2; Merit) catheter, which yielded some results.

The (SIM2; Merit) catheter was placed into the aortic arch. First, we navigated the catheter at the left SCA origin; injection of the contrast material showed a small diameter left SCA and hypoplastic left VA. The origin of the left SCA was dilated, known as Kommerell's diverticulum [Figure 3]. At the same time, injection of the contrast material at the brachiocephalic trunk origin showed a large diameter right SCA with a dominant right VA. However, the brachiocephalic trunk was absent and the four major branches arose independently. At this time, we speculated that it was a type of congenital anomaly; otherwise, we did not yet know about it.

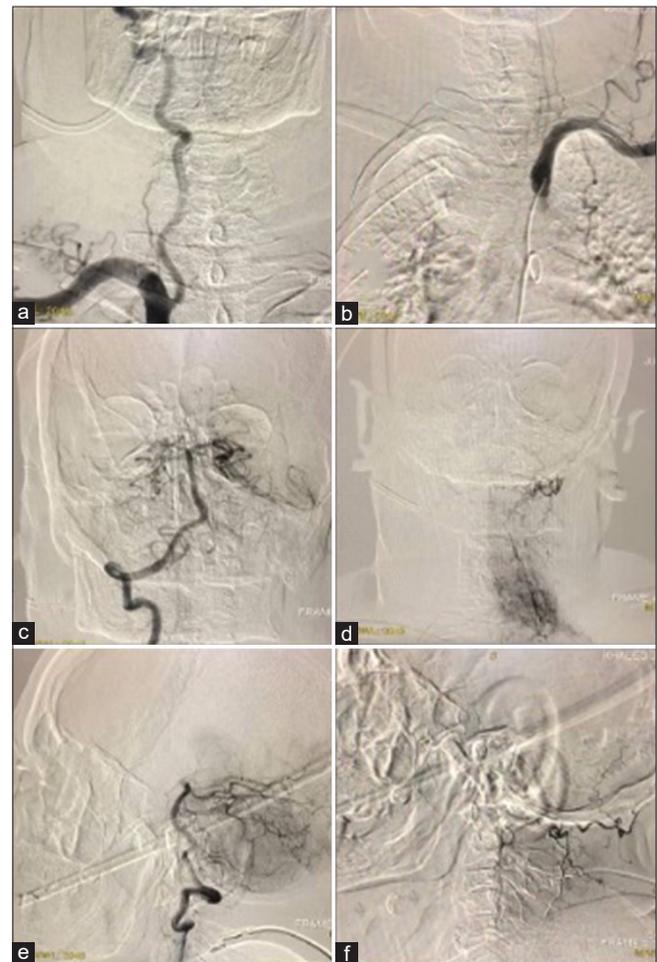
The carotids were the major challenge. We could not visualize their shadows by injecting contrast material near their origins or accessing them even with varying angles and maneuvers; all these efforts were unsuccessful in visualizing them. After several attempts of catheter navigation, the injection of the contrast material at the ascending aorta successfully visualized them only [Figure 4]. Nevertheless, we could not access them, so we could not examine the brain's anterior circulation.

### DISCUSSION

RAA is a rare anatomical abnormality established in <0.1% of the adult population when the aortic arch crosses the right main stem bronchus to the right of the trachea and

esophagus and descends along the right side of the vertebral bodies.<sup>[7]</sup> Fioratti and Aglietti originally described the RAA in 1763, and Edward later divided it into three categories in 1948. Type 1 RAA with mirror image branching occurs when the left CCA and LSCA originate from the left innominate artery and are followed by the right CCA and right SCA, mirroring the branches of the normal LAA. This type is typically associated with cyanotic congenital heart disease such as Tetralogy of Fallot or truncus arteriosus. Type 2 RAA with ALSA is seldom related to congenital cardiac disease and can cause compression symptoms, as in our case.

In addition, in Type 3 RAA, the left SCA is isolated and not connected to the aorta. However, because it is related to the pulmonary artery through the ductus arteriosus, it may also be associated with vertebrobasilar inefficiency and subclavian steal syndrome. The most frequent variety is Type 2, which



**Figure 3:** Catheter Angiography of the aortic arch branches shows a right-sided arch, aberrant left subclavian artery with kommerell's diverticulum (a,b). c-f: showing the right (dominant) and left (hypoplastic) vertebral arteries injection in PA and Lateral view respectively.



**Figure 4:** Catheter angiography of the aortic arch shows the left and right carotid arteries being barely visible after injection of the contrast material in the ascending aorta.

accounts for approximately 40% of all RAA cases, while Type 3 is extremely uncommon.<sup>[20,21]</sup>

The RAA with ALSA is formed during embryonic development as a result of the continuation of the right fourth embryologic aortic arch and in-folding of the left aortic arch, with regression of the segment between the left CCA and LSCA so that the left fourth arch becomes the proximal SCA rather than the definitive aortic arch.<sup>[12]</sup> In this type, and as in our case, the left CCA is the first branch from the arch, followed by the right CCA, right SCA, and left SCA. According to the literature, the RAA with ALSA must always have a left ductus arteriosus. The existence of the left ductus arteriosus creates a vascular ring by connecting the left pulmonary artery to the root of the ASLA. This is the most often encountered kind of vascular ring. In most cases, the vascular ring is often loose, so tracheal or esophageal compression symptoms are often mild-to-absent.<sup>[13]</sup>

Patients with a right aortic arch with a left ligamentum arteriosum are more likely to develop Kommerell's diverticulum, which is a remnant part of the left fourth aortic arch arising as a left SCA aneurysm.<sup>[14]</sup> An ALSA's proximal segment that is conically dilated at its aortic origin is referred to as a diverticulum. However, it is also referred to as "lusoria root," "remnant diverticulum," and "lusoria diverticulum." Kommerell's diverticulum can be seen behind the trachea in 5% of cases, between the trachea and esophagus in 15% of cases, and in the retroesophageal region in 80% of cases.<sup>[3]</sup> The absence or existence of retroesophageal diverticulum is essential to be reported due to its prognostic significance.<sup>[16]</sup> Furthermore, the size of the diverticulum has importance in deciding whether to make a surgical intervention or not. Complications, including esophageal distention,

tracheomalacia, or even aneurysmal tearing, may occur in patients having a diverticulum diameter of 5 cm or more.<sup>[11]</sup>

In general, patients with Type II RAA are primarily asymptomatic. Unless the atherosclerotic changes of the anomalous vessels, dissection, and aneurysms develop, 5% of adult patients with aberrant SCA experience symptoms due to atherosclerotic changes. The morbidity caused by compression of adjacent structures results in dysphagia (dysphagia lusoria), dyspnea, stridor, wheezing, cough, recurrent pneumonia, obstructive emphysema, or chest pain by structure compression.<sup>[4,8]</sup> Moreover, the mortality associated with dissection and rupture of an aberrant artery and Kommerell's diverticulum has been reported in 20–50% of patients. Stenosis at the origin of an ASLA arising from a Kommerell's diverticulum with a RAA is extremely rare.<sup>[6]</sup>

We report the first case of Type 2 RAA to present with the left side anterior circulation TIA and to be accompanied by five different anomalies as the following:

1. Kommerell's diverticulum
2. Aneurysmal dilation of the ascending aorta
3. Bilateral CCA and Left ICA stenosis
4. PLSVC
5. Pancake kidney

RAA is a quite rare variant of the aortic arch, not to mention that the association with ALSA is scarcer (0.05%), particularly when associated with Kommerell's diverticulum.<sup>[7]</sup> Moreover, to be combined with all those anomalies are an exceptionally rare congenital abnormality with no exact prevalence ratios in the literature, and to have all those anomalies considered have never been reported, especially in a symptomatic patient and especially when the symptoms are neurological rather than tracheoesophageal compression with no steal phenomena as well.

Based on a literature review of adult cases with Type 2 RAA, done between 2011 and 2019, two out of 31 cases presented anterior circulation abnormalities of nonatherosclerotic etiologies.<sup>[22]</sup> One with cerebrovascular insufficiency is caused by severe right CCA stenosis and steal phenomena.<sup>[19]</sup> The other case was pseudo-occlusion of the left ICA.<sup>[14]</sup> We report a case of anterior circulation insufficiency due to symptomatic left CCA and ICA stenosis (30–50%) causing the patient's symptoms. Furthermore, asymptomatic right CCA stenosis (30%) was traced only on the native brain CT scan as a watershed infarction of the left posterior, frontal, and parieto-occipital cortex but without any neurological deficit. Hence, this presentation has never been reported in patients with Type 2 RAA.

Various factors can influence atherosclerotic changes. Aging might also play a role in influencing a variety of functional and structural alterations of the large arteries by a compound

interaction with related agents, including genetics, disease, and environmental agents. Those functional and structural alterations of the aortic wall are among the variations of the proximal aorta associated with age, which will likely lead to functional and configurational alterations of the aortic arch itself.<sup>[17]</sup>

On the other hand, PLSVC and pancake kidney were exceptionally unexpected associations with RAA. Furthermore, it is considered to be one of the rarest instances. PLSVC is an uncommon variance among other vascular malformations and it mostly connects to the right atrium within the coronary sinus. PLSVC is accountable for venous drainage of the left part of the head, neck, and nearly 20% of the left arm. It is usually asymptomatic and incidental findings with a prevalence range between (0.2% and 3%). However, it is considered the most frequent inborn abnormality of the venous system in the chest.<sup>[2]</sup> In the literature, PLSVC was associated with RAA in nearly 16% of cases, according to an investigation by Ari *et al.*<sup>[1]</sup>

Pancake kidney is an exceptionally rare congenital abnormality with no exact prevalence ratios in the literature. This anomaly involves a full union of the medial parenchyma of both kidneys, no interposing septum, a specific collecting system for each kidney, and anteriorly positioned ureters that enter the bladder normally. However, our report described a pancake kidney case with only a single ureter drained into the bladder's left vesicoureteral junction.<sup>[15]</sup> To the best of our knowledge, there was no relation between the RAA and pancake kidney, so this may be the first study to report such an association.

Preoperative diagnosis of RAA will be crucial in avoiding significant neuroendovascular difficulties. Noninvasive echocardiography can provide an overall image of the aortic arch. Furthermore, magnetic resonance angiography and CTA afford accurate evaluation of the aortic arch, which can also aid in surgical or interventional planning. Doppler examination of the vessel must also be made to confirm the aberrant vessel.<sup>[5]</sup> However, in some cases of diagnostic catheterization or endovascular emergencies, the preoperative step is not something that would be routinely done. The catheter motion can aid in the diagnosis of RAA intraoperatively; if the catheter moves between the midline and right side of the chest during the catheter angiography performed through the transfemoral route, this will raise the possibility of RAA. The endovascular difficulties associated with RAA are very limited in the literature. However, difficulty due to the transfemoral route was reported in a carotid artery stenting (CAS) case for treating left ICA pseudo-occlusion associated with RAA. In this case, the examiner could not advance the catheter into the left CCA through the transfemoral route; they attributed it to the more proximal origin of the left CCA from the ascending

aorta. They did overcome this difficulty by shifting to the transbrachial route when the catheter passes through only one steep turn at the low origin of the left CCA.<sup>[14]</sup> In another case of CAS of the left ICA stenosis in a patient with RAA, the examiner successfully completed the procedure though a transfemoral approach, attributing this success to the preoperative recognition of the aortic arch anatomy.<sup>[18]</sup> From their experiences, we believe that the preoperative diagnosis with the knowledge of the anatomical characteristic of RAA can limit the technical difficulties during endovascular procedures. Still, we found that the literature lack more technical points on intraoperative diagnosis and on how to overcome upcoming difficulties in cases like these.

## CONCLUSION

We presented an extremely rare case of RAA with ALSA associated with a group of extra rare anomalies. Taking into consideration the incidental diagnosis of these anomalies and their uncommon occurrence, it is tremendously important to understand the anatomical variants of RAA and its characteristics to improve the intraoperative recognition of these vascular anomalies, which may aid in better management and follow-up of these anomalies.

## Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

## Financial support and sponsorship

Nil.

## Conflicts of interest

There are no conflicts of interest.

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**How to cite this article:** Al-Khazaali YM, Hummadi NA, Ismail M, Al-Waely NK, Ahmed FO, Hoz SS, *et al.* Right-sided aortic arch with complex anomalies presented with transient ischemic attack. *Surg Neurol Int* 2022;13:493.

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