Aberrant Right Vertebral Artery with a Diverticulum of Kommerell: Review of a Rare Aortic Arch Anomaly

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ABSTRACT

The normal aortic arch branching pattern is of a three-vessel arch with the vertebral arteries arising from the subclavian arteries. There are a variety of well-known symptomatic and asymptomatic aortic branching patterns widely reported in the literature. An anomalous right vertebral artery with a diverticulum of Kommerell is an extremely rare variant, with few other cases reported in the literature. Herein, we review the embryology of the aortic arch and vertebral artery, the various types of Kommerell's diverticula and the clinical significance of this anomaly.

CASE REPORT

CASE REPORT

A 57-year-old male with a past medical history of hypertension and remote gunshot wound to the chest presented to the emergency department with chest pain and shortness of breath. The patient underwent CT angiography (CTA) of the chest to rule out pulmonary embolism, which was negative for acute pathology. Incidentally noted on the CTA was anomalous origin of the right vertebral artery (VA) arising distal to the left subclavian artery (SCA) origin, with an associated diverticulum of Kommerell (Figure 1 and 2). The vessel originated distal and separate from the left SCA origin, and crossed posteriorly behind the esophagus and trachea and entered the right transverse foramen at C7 (Figure 2). Aside from common origin of the brachiocephalic and left common carotid arteries (a bovine arch configuration), no other vascular or cardiac anomalies were identified in the chest or of the great vessel origins.

DISCUSSION

Etiology & Demographics:

The normal aortic arch branching pattern is of a three-vessel arch with the vertebral arteries arising from the subclavian arteries. There are a variety of well-known symptomatic and asymptomatic aortic branching patterns widely reported in the literature. The most common vertebral artery variant is of the left vertebral artery arising directly from the aortic arch between the left common carotid artery and left subclavian artery, with a reported incidence rate of 2.4 to 5.8% in autopsy series in the general population [1,2]. In a large case series of 1286 patients evaluating all known vertebral artery origin variants, the left vertebral artery was significantly more involved (85%) than the right, and direct origin of the left vertebral artery between the left common carotid and left subclavian arteries represented the majority (60%) of all types of abnormal vertebral artery origins [3]. There are a variety of aberrant right VA reported in the literature with the most common right VA anomaly described being the right VA originating from the right subclavian root described in 80% of cases [3]. Origin of the right vertebral artery directly from the aortic arch distal to the left subclavian artery is a much more rare entity, with only 17 cases reported in the literature to date [1,2,4-10]. The reported incidence of a right vertebral artery originating distal to the left subclavian artery is as low as 1.5% among all vertebral artery origin variants (19 cases in a study

of 1231 cases of single aberrant vertebral artery origins including autopsy specimens) [3]. An anomalous right vertebral artery with a diverticulum of Kommerell is an extremely rare variant, with only 5 other cases reported in the literature, 2 of which had a bovine arch as presented here [4,6,10].

An understanding of aortic arch and vertebral artery embryology is crucial for delineating how this anomaly occurs. Originally proposed in 1948, Edwards hypothesis of a theoretical double aortic arch system describes an aortic arch and ductus arteriosus on each side, with the subclavian and carotid arteries arising from the respective ipsilateral right and left arches [11]. Based on this hypothetical model, normal aortic arch development and most congenital aortic arch anomalies may be explained by persistence of a segment that usually normally regresses, or regression of a segment that should normally persist [12]. Normally the vertebral artery arises from the first part of the ipsilateral SCA. Embryologically the right SCA is derived from the right 7th cervical intersegmental artery (CIA) and a portion of the adjacent right primitive dorsal aorta. The right vertebral artery develops from the right cervical intersegmental longitudinal anastomosis (which links the cervical intersegmental arteries) with some contribution from the right 7th CIA [13,14]. During normal fetal development, the right dorsal aorta caudal to the right 7th CIA regresses so that the right SCA and the right VA are on the ipsilateral side [Fig 3A (modified from Fig 6-A in Lemke)] [1]. It has been hypothesized that for the right VA alone to originate from the left arch distal to the left SCA (as in our case) requires (a), segmental regression of the right dorsal aorta between the 7th and the 8th right CIA and (b) persistence of the 8th right CIA as the future right VA. In this situation, the right SCA exhibits classic origin from the ipsilateral arch [1,15] [Fig 3B (modified from Fig 6-D in Lemke) [1]].

There are five different types of Kommerell's diverticulum described in the literature: (1) at the origin of an aberrant left subclavian artery in a right-sided aortic arch; (2) at the origin of an aberrant right subclavian artery in a left aortic arch; (3) at the aortoductal junction; (4) at the junction of a right aortic arch with the descending aorta in cases of mirror image branching, and (5) as described recently by Balani in 2015, involving a diverticulum at the origin of an aberrant right vertebral artery arising distal to the left subclavian artery [6]. While the exact embryological basis of the diverticulum of Kommerell is unclear, it is hypothesized that it is a remnant of the origin of the right dorsal aorta. During development of this anomaly, the right dorsal aorta regresses distally and will not connect to the right SCA (Figure 3B); with the residual right dorsal aorta origin remnant remaining as a diverticulum of Kommerell, and which embryologically also explains its retroesophageal course [4].

Clinical & Imaging findings:

Patients with an aberrant right vertebral artery are invariably asymptomatic and incidentally detected; if symptoms such as headache, dizziness, and/or dysphagia can be directly attributed to the presence of an aberrant right VA, this is referred to as dysphagia lusoria or arteria lusoria, though there is a paucity of literature describing patients with true vertebra lusoria. The clinical implications of an anomalous origin of a right VA are in pre-operatively detecting these anomalies prior to vascular surgery, esophageal surgery, and/or cerebral angiography and to avoid misinterpretation as vascular pathology such as an aneurysm. Additionally, some authors have even suggested that alterations in the aortic arch large vessel branching patterns can cause changes in cerebral hemodynamics that possibly may lead or predispose to vascular abnormalities, including an increased risk of dissection, aneurysms and arteriovenous fistulas [3]. A few studies have shown an increased risk of aortic dissection in patients with a direct aortic origin of the left VA when compared to classic subclavian origin of either the left or right VA [4,16]. Although the risk of dissection of an aberrant right vertebral artery is unclear, it is presumed that the hypothesized longer course of an anomalous left VA arising directly from the aortic arch with a reported corresponding associated increased risk of dissection may also be applied to an aberrant right VA, especially with a retroesophageal course [16]. It has been hypothesized that an aberrant right VA may be within a spectrum of other vascular abnormalities or more likely purely coincidental [3]. In a retrospective literature review of "aberrant vertebral artery" based on 214 articles by Yuan et al, with a cumulative total of 1286 cases, in 955 patients and 331 cadavers, 12% had one or more congenital cardiovascular anomalies, where an aberrant right subclavian artery was the most common [3]. Additionally, 9.3% of these patients with one or more congenital cardiovascular anomalies had one or more acquired vascular associations, including aneurysms, obstructions, stenosis, thrombus formation and dissection, of which 20.2% of which were VA-associated lesions and 0.8% having a VA dissection [3].

Treatment & Prognosis:

The case presented here is unique in light of the presence of a diverticulum of Kommerell associated with an aberrant right VA. In most cases previously describing an aberrant right vertebral artery the anomaly was detected in asymptomatic patients including those being evaluated for preoperative evaluation of aortic aneurysms, cerebral or spinal vascular malformations [2,4,17,18]. Of the cases published describing an aberrant right vertebral artery only 5 other case reports describe an associated diverticulum of Kommerell [4,6,10]. The hemodynamic consequences of this aberrant origin with an associated diverticulum and the potential changes in flow dynamics that could result from this diverticulum are unclear. However, complications described in the literature related to a diverticulum of Kommerell include an increased risk of atherosclerosis, embolism, rupture and dissection, all of which make detection and diagnosis even more crucial [4,6]. There is no consensus treatment guidelines for patients with complications related to a diverticulum of Kommerell, other than treat the respective complication whether it is a vascular dissection, rupture or embolic phenomena. Surgical intervention may be indicated in symptomatic patients with an aberrant right vertebral artery in a similar approach to those patients treated surgically for a symptomatic aberrant right subclavian artery. Although the aberrant right vertebral artery is significantly smaller in caliber than an aberrant right

subclavian artery, lack of awareness of this variant anatomy prior to esophageal or cardiac surgery with subsequent inadvertent injury of an unrecognized aberrant right vertebral artery may result in life-threatening complications including extensive hemomediastinum or neurological sequelae such as stroke. This case highlights the value of obtaining preprocedural MDCT to evaluate aberrant anatomy and avoiding potential complications related to them during angiography or head/neck/thoracic surgery.

Differential Diagnoses:

The differential diagnosis includes many aberrant aortic arch branching patterns and other lesions that can result in dilation at the origin of a vessel. Radiographic features in combination with clinical symptomatology are crucial to making the correct diagnosis.

Aberrant right subclavian artery with left aortic arch:

An aberrant right subclavian artery with left aortic arch is an aberrant branch arising from the distal left aortic arch and coursing as the right subclavian artery. This is the most common aortic arch anomaly and is typically asymptomatic, however it can present with dysphagia lusoria. As the aberrant artery passes posterior to the esophagus it may result in compression of the esophagus, giving the sensation of dysphagia.

Right sided aortic arch with aberrant left subclavian artery with or without a Diverticulum of Kommerell:

A right sided aortic arch results from persistence of the right fourth aortic arch and involution of the left aortic arch. Plain radiograph typically shows an absent left aortic contour with a right arch often projects as a mass in the right paratracheal region. There can be secondary tracheal bowing to the left. Findings on cross sectional imaging include a right-sided aortic arch with the descending thoracic aorta on the right side of the spine and aberrant left subclavian artery. There is an association of the right aortic arch with an aberrant left subclavian artery which may have a focal dilation at the origin consistent with a diverticulum of Kommerell.

Aortic ductus diverticulum:

An aortic ductus diverticulum is usually a smooth focal out pouching at the anteromedial aspect of the aorta at site of the ligamentum arteriosum. On cross sectional imaging this smooth focal bulge has obtuse angles within the aortic wall. It is important to distinguish the ductus diverticulum from an aortic pseudoaneurysm which has acute angles with the aortic wall.

Thoracic aortic aneurysm:

Thoracic aortic aneurysm is dilation of the thoracic aortic measuring greater than 5 cm. This dilation of the thoracic aorta can be identified on plain radiographs. The presence of atherosclerotic calcifications may also aid in the diagnosis. Cross sectional imaging will show luminal dilation of the thoracic aorta. Additional findings such as mural thrombus and atherosclerotic calcifications may also be present.

Aortic pseudoaneurysm:

Aortic pseudoaneurysm, results from disruption in the vessel wall in which blood leaks through the intima and muscularis propria but is contained by the adventitia. Ultrasound will show turbulent forward and backward flow resulting in the characteristic yin-yang sign. Cross sectional imaging will show a smooth walled sac adjacent to an artery. A communication to this adjacent vessel may also be visualized.

Right vertebral artery originating from the right subclavian root:

The right vertebral artery most commonly originates as the first branch from the right subclavian artery.

Direct origin of left vertebral artery:

Direct origin of left vertebral artery is the most common vertebral artery anomaly reported. Of the cases reported of anomalous left vertebral artery most originate from the aortic arch between the common carotid and left subclavian arteries.

In conclusion, though rare, the diagnosis of an aberrant right vertebral artery especially with a diverticulum of Kommerell is crucial prior to cerebral angiography, head, neck and thoracic surgical intervention to avoid unforeseen complications.

TEACHING POINT

The aberrant right vertebral artery is a rare aortic arch anomaly that is important to recognize on CT as a branch distal to the takeoff of the left subclavian artery and which takes a retroesophageal course. The aberrant right vertebral artery has important clinical implications in pre-operative planning and angiographic intervention.

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FIGURES

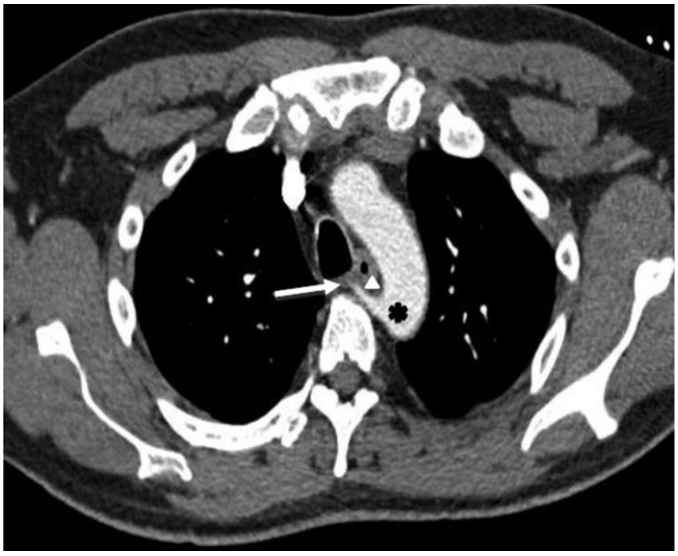

Figure 1: 57 year old male with an aberrant right vertebral artery with a diverticulum of Kommerell.
Findings: Direct origin of the right vertebral artery (arrow) as the last branch arising from the aortic arch (asterisk) and taking a retroesophageal course (triangle), with a diverticulum of Kommerell (lateral to asterisk) .
Technique: Axial contrast enhanced CT (Multislice, 64 detector) of the chest acquired in the arterial phase (3914 mAs, 120kV, 3.0mm slice thickness), Contrast: 100mL Omnipaque 350.

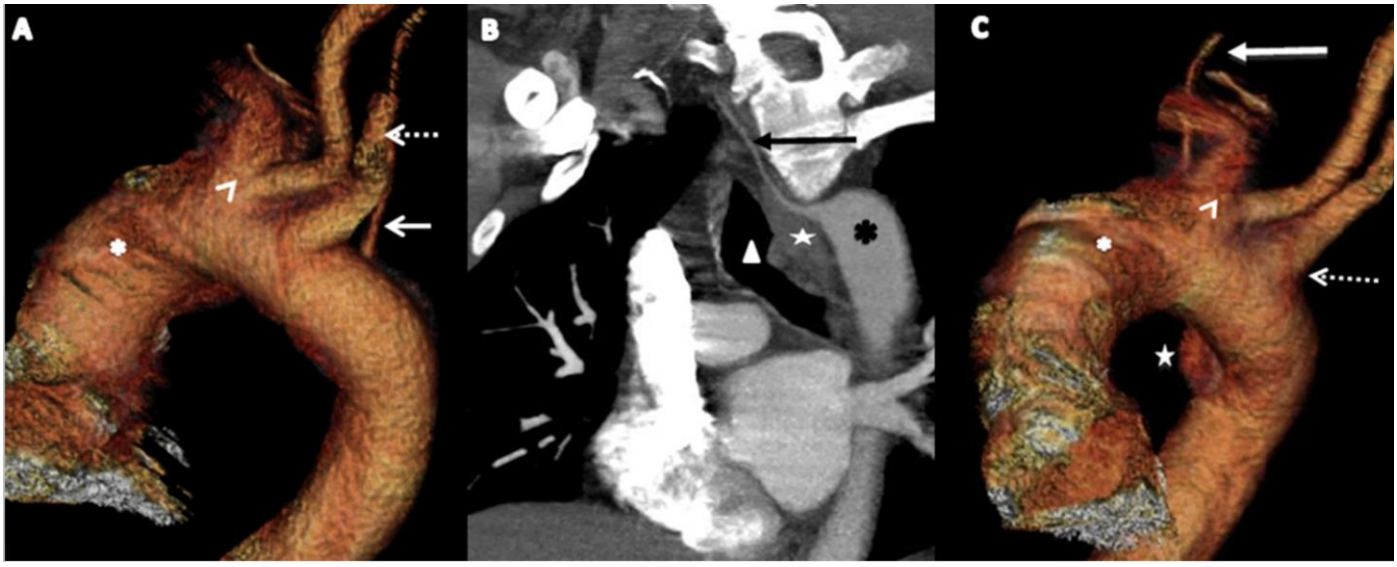

Figure 2: 57 year old male with an aberrant right vertebral artery with a diverticulum of Kommerell.
Findings: Coronal CT and maximum intensity projection (MIP) (Anterior and coronal) images showing diverticulum of Kommerell (star) arising directly from the aortic arch (asterisk) distal to the takeoff of the brachiocephalic trunk (arrow head) and right subclavian arteries (dashed arrow). The right vertebral artery crosses the midline from left to right (solid arrow) and has a retrotracheal course (triangle).
Technique: Oblique coronal contrast enhanced CT (Multislice, 64 detector) at the level of the aortic arch in the arterial phase MIP image (B) and anterior (A) and oblique coronal (C) 3D volume rendered image (3914 mAs, 120kV, 0.75mm slice thickness), Contrast: 100ml Omnipaque 350.

Figure 3A

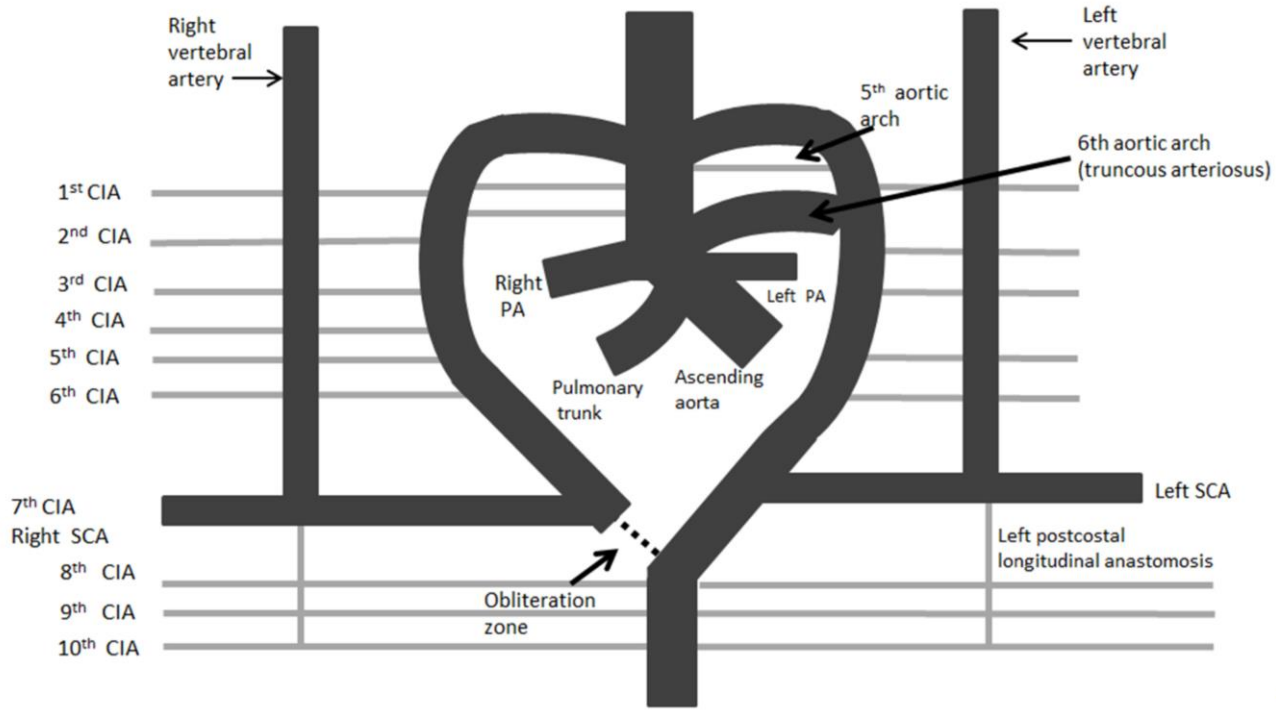

Figure 3B

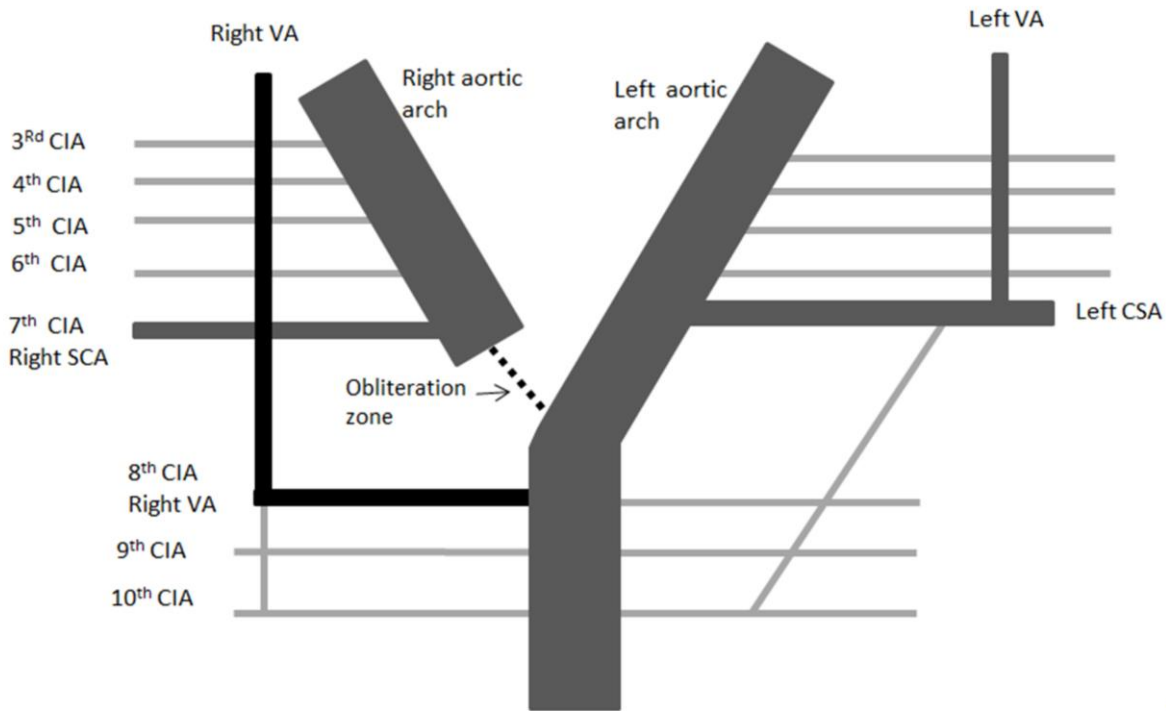

Figure 3: (A) Normal embryological development of the aortic arch and its major branches. (Adopted from Lemke et al 1999, figure 6A) [1]. (PA=pulmonary artery) (SA=subclavian) (CIA=cervical intersegmental artery).
(B) Aberrant embryological development of the right vertebral artery arising directly from the left side of the aortic arch. This may occur if the right VA arises from the 8th CIA with segmental regression (the obliteration zone) occurring distal to the right SA origin. This leads to the right SA originating from the right side of the aorta and the right VA arising from the left aorta distal to the left SA. (Adopted from Lemke et al 1999, Figure 6D) [1].

Etiology	Proposed segmental regression of the right dorsal aorta between the 7th and the 8th right CIA and persistence of the 8th right CIA as the future right VA
Incidence	Unknown, 5 cases published in the literature to date
Gender ratio	No gender predilection identified
Age predilection	Congenital anomaly Most cases reported as an incidental finding in adults
Risk factors	None reported
Treatment	Most often non-required. Surgical intervention may be indicated in symptomatic patients
Prognosis	Usually incidental, however, gone unrecognized can have life threatening consequences in thoracic surgery and angiography
Imaging Findings	Right vertebral artery originating distal to the left subclavian with a retroesophageal course and aneurysmal dilation at the origin consistent with a diverticulum of Kommerell

Table 1: Summary table for aberrant right vertebral artery with a Diverticulum of Kommerell.

Aberrant right vertebral with Diverticulum of Kommerell	CT/MRI • Right vertebral artery originating distal to the left subclavian with a retroesophageal course and focal dilation at the origin consistent with a diverticulum of Kommerell
Aberrant right subclavian artery with left aortic arch	CT/MRI • Aberrant branch arising from the distal left aortic arch and coursing as the right subclavian artery
Right sided aortic arch with Aberrant left subclavian artery +/- Diverticulum of Kommerell	Plain radiograph • Absent left aortic contour • Right arch often projects as a mass in the right paratracheal region +/- tracheal bowing to the left CT/MRI • Right-sided aortic arch with the descending thoracic aorta on the right side of the spine and aberrant left subclavian artery +/- arising focal dilation at the origin consistent with a diverticulum of Kommerell
Aortic ductus diverticulum	CT/MRI • Smooth focal bulge with obtuse angles within the aortic wall
Thoracic aortic aneurysm	Plain radiograph • Dilation of the thoracic aortic +/- atherosclerotic calcifications CT/MRI • Luminal dilation of the thoracic aorta +/- mural thrombus +/- atherosclerotic calcifications
Aortic pseudoaneurysm	Ultrasound • Turbulent forward and backward flow, characteristic yin-yang sign CT/MRI • Smooth walled sac adjacent to an artery +/- a communication
Right vertebral artery originating from the right subclavian root	CT/MRI • Right vertebral artery originating from the right subclavian artery
Direct origin of left vertebral artery	CT/MRI • Left vertebral artery originating from the aortic arch between the common carotid and left subclavian artery

Table 2: Differential diagnosis table for aberrant right vertebral artery with a Diverticulum of Kommerell.

ABBREVIATIONS

CIA = Cervical intersegmental artery
CTA = CT angiography
PA = Pulmonary artery
SCA = Subclavian artery
VA = Vertebral artery

KEYWORDS

Aberrant right vertebral artery; aortic arch anomalies; Diverticulum of Kommerell; aortic arch embryology; vertebral artery lusoria; CT angiography