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Kommerell Diverticulum and Agenesis of the Left Common Carotid Artery in a Patient with Dysphagia: A Case Report

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Abstract: Kommerell diverticulum is the aneurysmal dilatation of the proximal descending aorta at the origin of an aberrant subclavian artery and may occur in either the left-sided or right-sided aortic arch. We report an unusual case of right-sided aortic arch with Kommerell diverticulum and concurrent agenesis of the left common carotid artery in a patient with progressive dysphagia to solid food. A brief overview of embryology, relevant anatomy, radiographic findings, and treatment options of Kommerell diverticulum are presented.

Keywords: *Kommerell diverticulum, dysphagia, right aortic arch, aberrant left subclavian artery, agenesis of the common carotid artery*

Introduction

Congenital vascular anomalies of the aortic arch have been described extensively in the literature.[1] One such anomaly, right aortic arch (RAA) with Kommerell diverticulum (KD) and aberrant left subclavian artery (ALSA), has a reported average prevalence[2] between 0.016% and 0.16%. The agenesis of the left common carotid artery (CCA) is an even more rare entity with an unknown incidence.[3] To our knowledge, the co-occurrence of agenesis of the left CCA with RAA and ALSA was reported in the literature[4] only once, in an asymptomatic patient undergoing surgical evaluation for mitral valve repair. We report a case of similar anatomy in a patient who presented with the onset of insidious dysphagia.

Key Points

- The formation of a Kommerell diverticulum from a right-sided aorta in the absence (agenesis) of the left common carotid artery is an exceptionally rare congenital anomaly.
- Patients with Kommerell diverticulum may present with symptoms of tracheal and esophageal compression and/or complications of diverticulum rupture secondary to age-related degenerations of the diverticulum.
- The treatment options of patients with Kommerell diverticulum are largely dependent upon patient-specific anatomy and comorbidities.

Abbreviations
RAA: right aortic arch
KD: Kommerell diverticulum
ALSA: aberrant left subclavian artery
CCA: common carotid artery
CTA: computed tomography angiography
ICA: internal carotid artery
ECA: external carotid artery

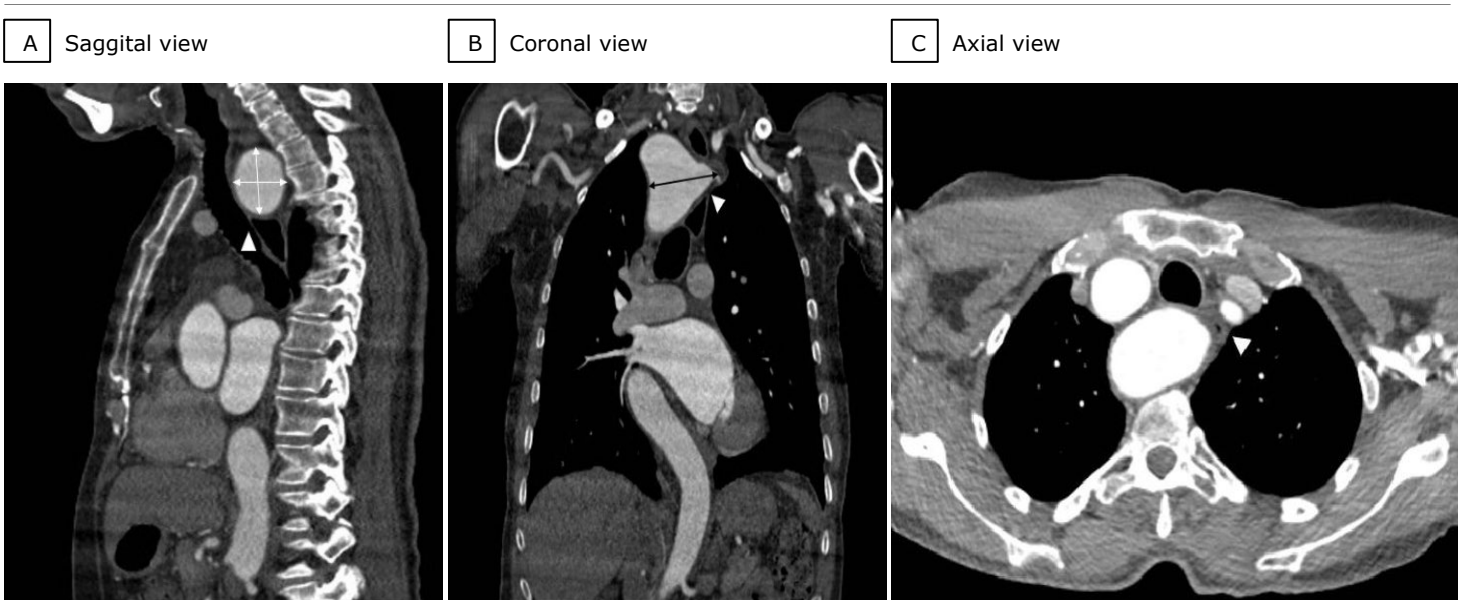
Case Presentation

The patient, an 81-year-old man with a history of seizures and hypertension, was found to have RAA with ALSA and an associated KD while undergoing evaluation for progressive intermittent dysphagia to solid food. Computed tomography angiography (CTA) of the chest showed the KD exerting significant mass effect on the upper esophagus near the thoracic inlet (Figure 1A-1C). Atheromatous changes and severe luminal narrowing of the proximal left subclavian artery were also present (Figure 2A, 2B). The left CCA was absent (Figure 2C, 2D), which was initially thought to be secondary to chronic atherosclerotic occlusion and not to agenesis. Subsequent CTA of the head and the neck showed absence of the left CCA, suggestive of its agenesis (Figure 3). In addition, there was hypoplasia of the left proximal internal carotid artery (ICA), diminutive caliber of the left distal ICA, and a small caliber of the left-sided cerebral vasculature with an intact circle of Willis. The patient denied any significant weight loss, dyspnea, or chest pain and did not report episodes of transient ischemic attacks or chronic left arm pain that may indicate subclavian steal syndrome. The patient's seizures were determined to be unrelated to his vascular anomalies. Subsequently, after being evaluated by a cardiac surgeon, the patient opted for conservative treatment.

Discussion

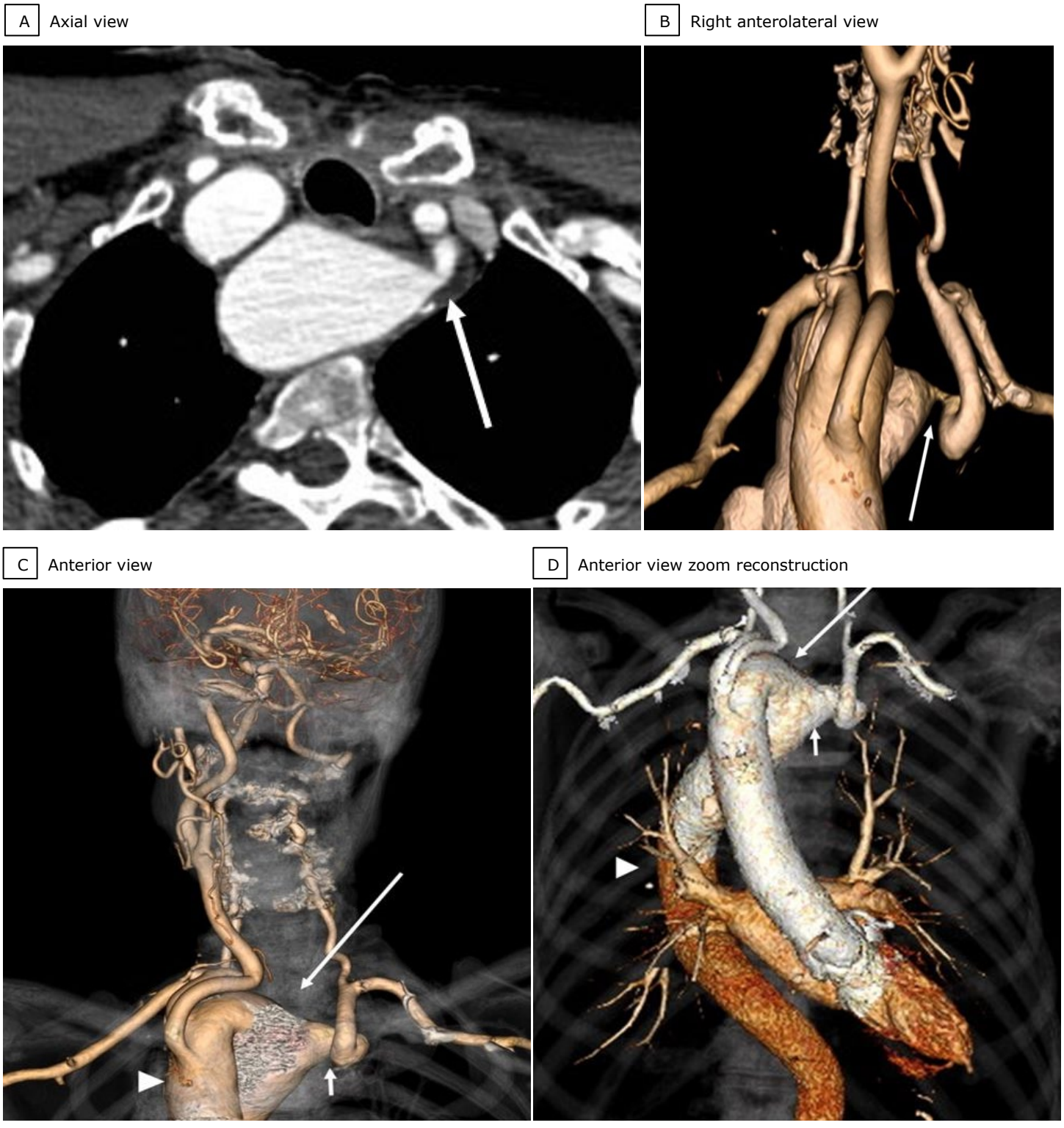
It is widely accepted[5] that the embryonic development of the ICAs starts from the cranial portions of the paired dorsal aortae and the third pair of the aortic arches (Figure 4). Research findings postulated that the CCAs also originate from the third aortic arches,[4-6] and the external carotid arteries (ECA) have separate origins from the ICAs, given reported cases of intact ECAs in the absence of ICAs.[6,7] It has been suggested that the ECAs originate from the second pair of the

Figure 1. Computed Tomography Angiography (CTA) of the Chest of an 81-Year-Old Man with Right Aortic Arch (RAA), Kommerell Diverticulum (KD), Aberrant Left Subclavian Artery (ALSA), and Agenesis of the Left Common Carotid Artery (CCA)



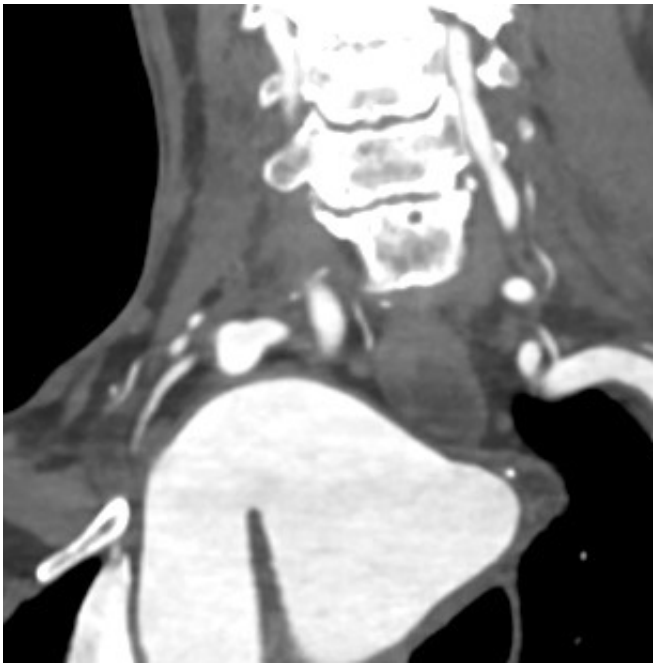
(A-C) CTA images show the size of KD. The diverticulum with a diameter of 42 x 32 millimeters measured at the orifice (A, double arrows) and a diameter of 58 millimeters measured from the opposite aortic wall to the tip of the diverticulum (B, double arrows). There is an associated mass effect with effacement of the upper esophagus by Kommerell diverticulum (A, B, and C, arrowheads).

Figure 2. Computed Tomography Angiography (CTA) of the Chest and Three-Dimensional Reconstruction of Right Aortic Arch, Kommerell Diverticulum, and Stenotic Proximal Left Subclavian Artery



(A-D) Two-dimensional (A) and three-dimensional (B) images show stenotic proximal left subclavian artery (A and B, arrow). Three-dimensional reconstructions illustrate right aortic arch (C and D, arrowhead), Kommerell diverticulum (C and D, short arrow), and the expected location of the congenitally absent left common carotid artery (C and D, long arrow).

Figure 3. Computed Tomography Angiography (CTA) of the Neck of an 81-Year-Old Man with Agenesis of the Left Common Carotid Artery (CCA) and Right Aortic Arch with Kommerell Diverticulum

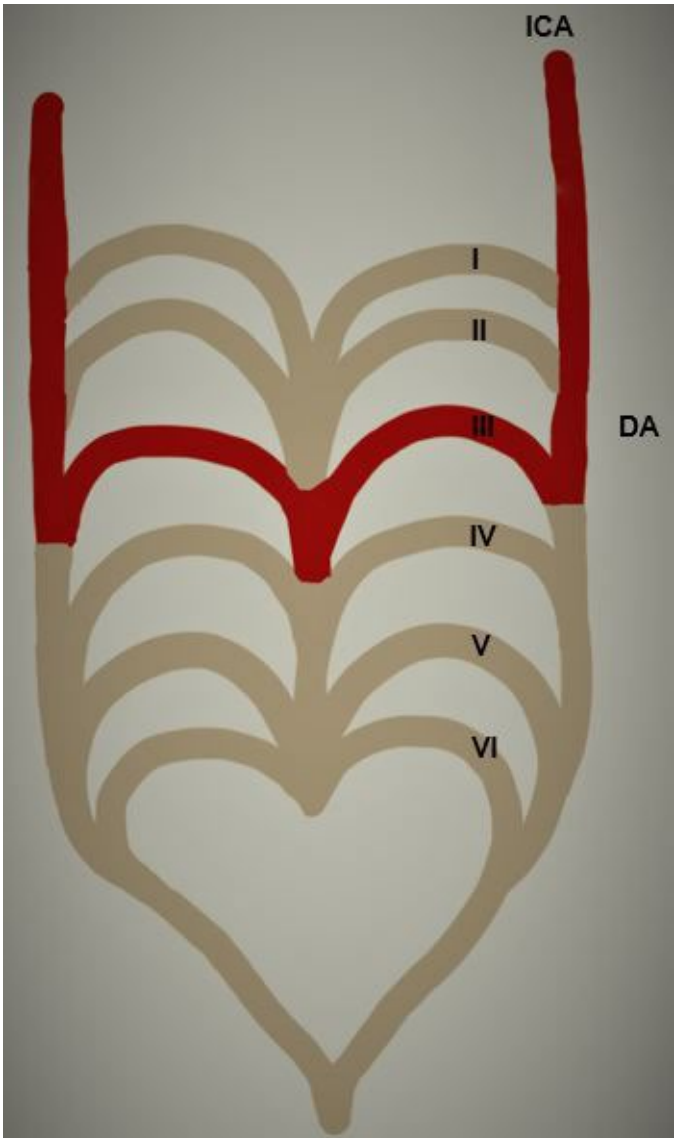


CTA image, coronal view, shows absence of the left CCA. There is no evidence of an atherosclerotic vessel to suggest atheromatous occlusion of the left CCA.

aortic arches during the later stages of their development.[5-7] Abnormal development of the left dorsal aorta as well as involution of the third aortic arch and a prolonged existence of ductus caroticus may lead to agenesis, aplasia, or hypoplasia of the left ICA and the left CCA, respectively.[3,7]

The natural history of KD is often uncertain, given its low incidence in the general population.[2,4] Based on observational studies,[8] an overall benign course with rare instances of progressive enlargement of KD seems to be the norm. While there is no consensus regarding treatment indications for asymptomatic patients, the most consistent indications for surgical repair of KD are its size and mass effect, aneurysmal degeneration, and acute rupture or dissection.[9,10] Currently, there are no guidelines on optimal strategies for surgical repair of KD because of the low incidence of this condition and heterogeneity of the patient population.[9] Options for treatment are determined by the patient-specific anatomy and comorbidities.[2,8,9] Large single center studies in the past decade have shown comparable outcomes for both open and hybrid endovascular repair approaches.[2,9,10] Open surgical repair of KD may be accomplished in the following ways: diverticulum resection, interposition grafts, patch repair, and elephant trunk procedure.[2,10] Endovascular procedures frequently entail thoracic endovascular aortic repair with stent grafting and embolization.[2,9,10] In most cases, both open and endovascular aortic repair include carotid-subclavian bypasses.[2,10] Overall, the choice of treatment is patient specific, and careful preoperative planning remains essential, given the complexity of the operations and potential serious complications.[2]

Figure 4. Diagram of the Embryogenic Development of the Aortic Arches and the Internal Carotid Artery (ICA)



DA: dorsal aorta. ICA: internal carotid artery. I: first aortic arch. II: second aortic arch. III: third aortic arch. IV: fourth aortic arch. V: fifth aortic arch. VI: sixth aortic arch.

Conclusion

The patient described in this report presented with an onset of insidious dysphagia secondary to the mass effect of the KD, as illustrated on CTA of the chest. Although the patient's existing comorbidities may not have precluded him from having a repair of KD, the complexity of the case lies in the patient's unique anatomy, specifically absence of the left CCA, which may have left limited options for bypass. The perioperative risk of cerebrovascular complications was also uncertain. The intact circle of Willis and no reported history of transient ischemic attacks in this case suggest that there must be extensive collaterals in place to maintain adequate cerebral perfusion. Nevertheless, the exact dynamic changes in arterial flow and stress response of abnormal cerebral vasculature during a major operation are factors that are difficult to predict preoperatively. Given these medical intricacies, it is possible to consider a conservative approach to symptom management. This case highlights the heterogeneity of the population of patients with KD and emphasizes the challenges associated with formulating a treatment plan that accounts for patient-specific anatomy and comorbidities.

Author Contributions

Conceptualization, S.H. and B.W.; Acquisition, analysis, and interpretation of data, K-L.N., A.P., and A.H.E-S.; Writing – original draft preparation, B.W.; Review and editing, S.H. and B.W.; Supervision, J.C. All authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All authors had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis

Disclosures

None to report.

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