Azygous Right Internal Carotid Artery with Congenital Absence of the Cervical Left Internal Carotid Artery Associated with Bilateral Aberrant Subclavian Arteries: A Case Study

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ABSTRACT

Congenital absence of the internal carotid artery is a rare vascular anomaly. Incidence associated with an anomalous aortic arch with aberrant both right and left subclavian arteries is unique. Although often asymptomatic, developmental anomalies of the cervicocerebral vasculature have become increasingly common incidental findings due to the widespread use of cross-sectional imaging. We present the unique case of a 60-year-old female admitted for bacterial meningitis, with imaging incidentally diagnosing congenital absence of the left internal carotid artery associated with a midline, "azygous", right internal carotid artery, as well as an anomalous aortic arch with bilateral aberrant subclavian arteries.

CASE REPORT

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A 60-year-old female presented to the emergency department with a two-week history of worsening headache. She also reported photophobia and increasing confusion. Upon presentation, she was found to be febrile and tachycardic with a significant leukocytosis. She was admitted, and subsequent blood cultures and lumbar puncture confirmed a diagnosis of bacterial meningitis. Computed tomography (CT), Magnetic resonance imaging (MRI) of the brain, and computed tomography angiography (CTA) of the neck and head were performed.

Imaging Findings

MRI brain without contrast was unremarkable in terms of evaluation for CNS cerebritis or meningitis. Imaging, however, showed curious asymmetric widening of the left Meckel's cave. The left carotid canal was suspected to be absent, subsequently confirmed by CT brain imaging (Figure 1).

CTA of the head and neck was subsequently performed demonstrating a dominant right common carotid artery coursing superiorly to a midline, retropharyngeal location (Figure 2). The right internal carotid artery (ICA) then further courses superiorly in a midline, retropharyngeal location to the level of C2, then courses laterally to a normally located right carotid canal. The left common carotid artery was identified as the second branch of the aortic arch but was notably diminutive in caliber. The left common carotid terminated in branches of the left external carotid artery without an identifiable ICA. Therefore the right ICA was termed azygous as it represented the sole cervical internal carotid artery (Figures 3a,3b). The vertebral arteries arose from their respective normal anatomic positions from their respective subclavian arteries. There was reconstitution and patency of the left supraclinoid ICA as well as continuous opacification of the anterior and middle cerebral arteries supplied by collaterals (Figure 4).

CTA of the head and neck also demonstrated an anomalous aortic arch with aberrant right and left subclavian arteries (Figure 5). The right common carotid artery arises normally as the first branch off the brachiocephalic artery with continuity to the right subclavian artery. However, in this case, the right subclavian arises as the fourth branch of the aortic arch, arising from the medial wall of the descending aorta. It courses in a retroesophageal, retrotracheal position to its normal right subclavicular position. The left subclavian artery arises as the third branch of the aorta, but from an anomalous low position, then courses retrograde, medially, and parallel to the aorta to its normal anatomic position prior to coursing laterally to its normal subclavicular position. Therefore, both subclavian arteries were termed aberrant, with both arising from anomalous positions from the aortic arch. Further, the aberrant left and right subclavian arteries arise from opposing positions from the descending aorta.

Management and Follow-up

After starting antibiotics, the patient's symptoms and lab work improved. She was discharged home with outpatient antibiotics. It was determined that the absence of her left ICA had not caused her any symptoms, nor was she symptomatic from her aortic arch anomalies. Follow-up for her vascular abnormalities was not indicated at the time.

DISCUSSION

Etiology and Demographics

Congenital absence of the internal carotid artery is a rare vascular anomaly that is estimated to occur in less than 0.01% of the population [1]. Furthermore, it has been reported that a unilateral left ICA absence is three times more likely than the absence of the right ICA [2]. A review of the literature has not supported any difference in the prevalence of this anomaly between males and females.

Embryologically, the cervical great vessels, aortic arch, and pulmonary trunk develop from six pharyngeal arches [3]. The ICA arises from the involution of the dorsal aorta and the third aortic arch and is usually entirely developed by six weeks [4]. Furthermore, it has been suggested that the carotid canal develops in association with the ICA, and the skull base does not begin to form until weeks five to six [5]. Thus, the failure of the third aortic arch and the distal aorta to involute by approximately week five will result in agenesis of the ICA with resultant agenesis of the carotid canal. Moreover, the embryologic development of the ICA has several complex steps, and any disruption in these steps will result in an abnormal artery.

Aberrant right subclavian artery (ARSA) is another infrequent anomaly that occurs in 0.4-1.8% of the population [3]. It is associated with trisomy 21, trisomy 18, and other chromosomal defects [6]. Previous studies have suggested that the anomaly may be more prevalent in females [7]. The development of the left aortic arch occurs early in gestation and, following a series of complex steps, normally results in the formation of a left aortic arch from which the brachiocephalic trunk, the left common carotid, and the left subclavian arteries arise. Embryologically, a double aortic arch first develops with a left and right arch connecting the ascending portions. Each aortic arch gives rise to a common carotid artery and a subclavian artery. Normally, the right arch distal to the origin of the right subclavian artery regresses while the right common carotid and subclavian arteries merge to form the brachiocephalic artery [8]. An aberrant right subclavian artery forms when the right aortic arch regresses between, rather than distal to, the common carotid and right subclavian arteries [9]. This impedes their fusion to form the brachiocephalic artery. The result is a left aortic arch that gives right to four arteries: the right common carotid artery, the left common carotid artery, the left subclavian artery, and the aberrant right subclavian artery.

An anomalous left subclavian artery arising from the distal descending aorta is an even rarer anomaly that has been reported very seldom in the literature. An aberrant left subclavian artery is most commonly associated with a right-sided aortic arch, which occurs in 0.1% of the population. About half of patients with a right sided aortic arch also have an aberrant left subclavian artery [10]. These findings are also associated with congenital heart diseases such as tetralogy of Fallot or coarctation of the aorta. In the literature, there has only been one case of an aberrant left subclavian artery not associated with a right sided aorta or a congenital heart abnormality [11]. These authors hypothesized that an anomalous left subclavian artery is due to interrupted movement of the seventh intersegmental artery cranially during embryogenesis rather than anomalous aortic arch development.

Clinical and Imaging Findings

Clinically, most patients with congenital absence of the left internal carotid artery are asymptomatic. When symptomatic, the most common presentations are cerebrovascular events, including transient ischemic attacks, subarachnoid hemorrhages, and parenchymal hemorrhages [12]. The fact that most patients remain asymptomatic is due to compensatory collateral circulation.

A CT head or MRI brain is the preferred imaging modality for diagnosis. Initial findings will show a missing internal carotid artery. CT or MRI can be used to evaluate an absent bony carotid canal in the case of agenesis. MR angiography of the cerebral vasculature can then adequately visualize the circle of Willis and other congenital variations that may be present.

Similar to absence of the ICA, patients with an aberrant right subclavian artery are most often asymptomatic. However, it can be associated with dysphagia, termed dysphagia lusoria. In these cases, the aberrant right subclavian artery courses behind the esophagus and compresses it [13]. The compression can present with cough or dysphagia.

Imaging findings of ARSA depend on the modality used. Plain radiographs of the chest may show obliteration of the retrotracheal space. An upper GI contrast study can demonstrate displacement of the contrast-filled esophagus. The displacement of the esophagus by the aberrant vessels produces the classic bayonet deformity of the aberrant right subclavian artery. CT and MRI are the preferred imaging modalities for diagnosing ARSA. They both demonstrate the aberrant branch arising from the distal left aortic arch with the artery then coursing medially. Visualization of an aberrant left subclavian artery is best achieved using MRI or multislice CT. Findings show a left subclavian artery arising more distally from the descending aorta than its normal origin.

Treatment and Prognosis

For each of the anomalies, no treatment is recommended in asymptomatic cases. Patients with an absent internal carotid

artery have an increased incidence of intracranial aneurysms [10]. Thus, it is important to follow these patients clinically and radiographically for the development of complications from existing aneurysms or the formation of new ones. Congenital absence of the ICA is also significant in the evaluation of cerebrovascular disease, especially when considering revascularization procedures such as carotid artery stenting or endarterectomy, as both cerebral hemispheres may be supplied by the atherosclerotic carotid. Additionally, failure to identify intercavernous collaterals can have grave implications in the setting of transsphenoidal hypophyseal surgery [2].

There is currently no primary treatment for ICA agenesis, aside from surgical bypass procedures or endovascular augmentation procedures should collaterals become compromised by acquired traumatic or atherosclerotic stenosis.

Awareness of potential aberrant bilateral subclavian arteries is vital in the treatment of cardiovascular and vertebrobasilar diseases, where access to the subclavian arteries may be essential for vertebral or internal thoracic artery access. Treatment of aberrant subclavian arteries can be considered when there are persistent symptoms of dysphagia lusoria. The preferred treatment is endovascular stenting is stenosis is symptomatic. There are three main surgical options: open repair, thoracic endovascular aortic repair (TEVAR), and hybrid repair [14]. Identifying an aberrant right subclavian artery is important when considering any surgical procedure involving the esophagus. Without sufficient caution, injury to the aberrant artery can cause mediastinal hemorrhage or an arterioesophageal fistula [3]. Due to the extreme rarity of an anomalous left subclavian artery not associated with a right aortic arch or other congenital cardiac abnormalities, treatment protocols have not been established.

Differential Diagnosis

A differential diagnosis when considering absence of the internal carotid artery includes severe stenosis, which may be secondary to dissection, atherosclerotic stenosis, or occlusion. In these cases, a carotid artery is present but decreases in size secondary to acquired disease. Contrary to congenital absence of the ICA, the carotid canal would remain present in these diagnoses.

There is no differential for aberrant subclavian arteries. However, if one sees bulbous enlargement of the proximal subclavian artery at its origin from the aortic arch, a Kommerell diverticulum must be considered. This was not present in our case.

TEACHING POINT

Congenital absence of an internal carotid artery, combined with both aberrant left and right subclavian arteries, is truly unusual. The synchronous occurrence of all three of these anomalies is truly rare and unique. Congenital absence of the ICA can be diagnosed with lack of visualization of the cervical ICA and absence of the bony carotid canal at the skull base. An aberrant right subclavian artery is diagnosed when the aberrant branch of the subclavian artery is seen arising independently as the fourth branch of the aortic arch, and an aberrant left subclavian artery is diagnosed when its origin is identified below the aortic arch, with the vessel subsequently coursing retrograde.

Authors' contributions

Robert Koenigsberg conceived, supervised, and supported the study.

Luke Ross wrote, reviewed, and edited the manuscript.

QUESTIONS

1. What is the name of the clinical condition that may be caused by an aberrant right subclavian artery?

Achalasia
Esophageal spasm
Dysphagia lusoria (applies)
Boerhaave syndrome
Mallory-Weiss tear

Explanation: Dysphagia lusoria occurs when an aberrant right subclavian artery courses posterior to the esophagus and compresses it. Symptoms include cough or difficulty swallowing (dysphagia) which can be intermittent or persistent depending on the severity of the compression.

2. Which of the following is the preferred imaging modality for diagnosis of congenital absence of the internal carotid artery?

X-ray
Ultrasound
CT angiography (applies)
PET scan
Echocardiography

Explanation: CT angiography is the most appropriate choice due to its effective ability to evaluate vascular abnormalities. The other answer choices are not effective.

3. Which of the following is false concerning congenital absence of the internal carotid artery?

It is due to failure of the third aortic arch and distal aorta to involute during embryogenesis.
It is most commonly unilateral.
These patients have an increased incidence of intracranial aneurysms.
It may present with a transient ischemic attack (TIA).
It always requires treatment. (applies)

Explanation: It is true that congenital absence of the ICA is most likely due to errors during embryogenesis, specifically involving the failure of the third aortic arch and distal aorta to involute. It is also true that this anomaly most often occurs

unilaterally although bilateral absence can occur. Patients with this condition may present with a TIA and are reported to have higher rates of intracranial aneurysms. It is false to say that these patients always require treatment because they are typically asymptomatic. Furthermore, no treatment is available to correct the absence.

4. What is the embryologic basis for the development of an aberrant right subclavian artery?

Persistent of the right dorsal arch
Developmental anomaly of the pharyngeal arches
Involution of the left aortic arch
Regression of the right aortic arch (applies)
Failure of the third aortic arch and distal aorta to involute

Explanation: In ARSA, there is an anomaly in the regression of the right aortic arch. Instead of regressing distally to allow the right subclavian to arise from the brachiocephalic trunk, it regresses between the origins of the right common carotid and right subclavian arteries leading to a right subclavian artery arising as the most distal branch off the aortic arch.

5. The bilateral common carotid arteries are derived from which aortic arch?

First
Second
Third (applies)
Fourth
Fifth

Explanation: The third aortic arch contributes to the formation of the common carotid arteries bilaterally. It also gives rise to the proximal internal carotid arteries bilaterally.

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FIGURES

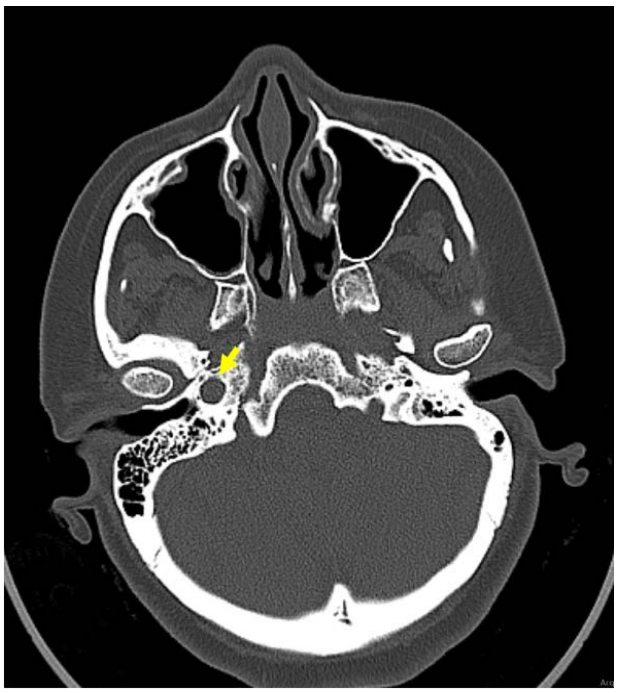


Figure 1: 60-year-old female with incidental findings of congenital absence of the left internal carotid artery and aberrant left and right subclavian arteries.
Findings: Axial CT brain showing the right carotid canal (arrow) and developmental absence of the left carotid canal.

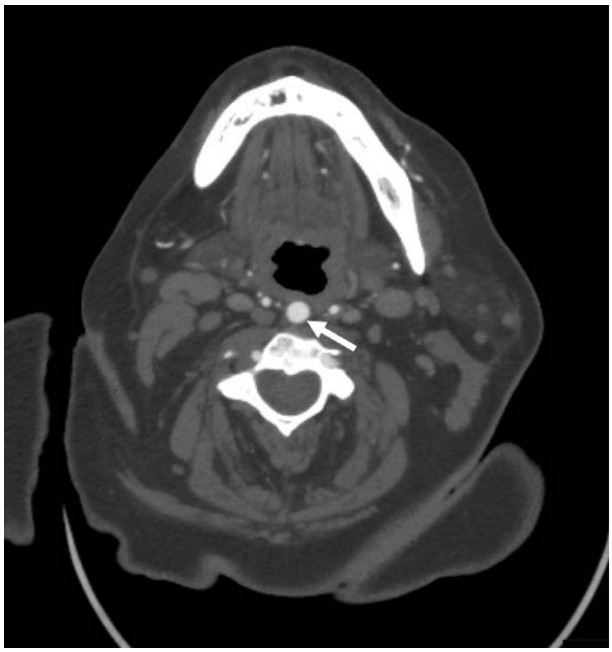
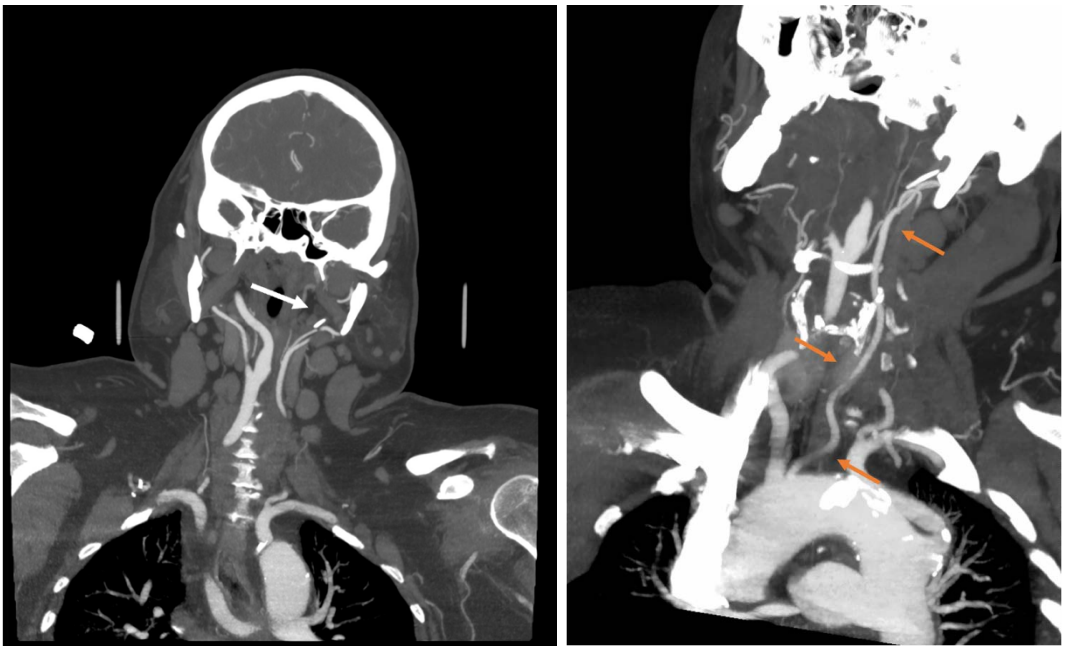


Figure 2:
Findings: Axial CTA neck and head showing the right internal carotid coursing superiorly in a midline, retropharyngeal position (arrow).



a b

Figure 3:
Findings: a. Coronal CT demonstrating a hypoplastic left common carotid artery terminating in branches of the left external carotid artery, and b. oblique coronal MIP image confirming the hypoplastic left carotid artery terminating in external carotid branches.

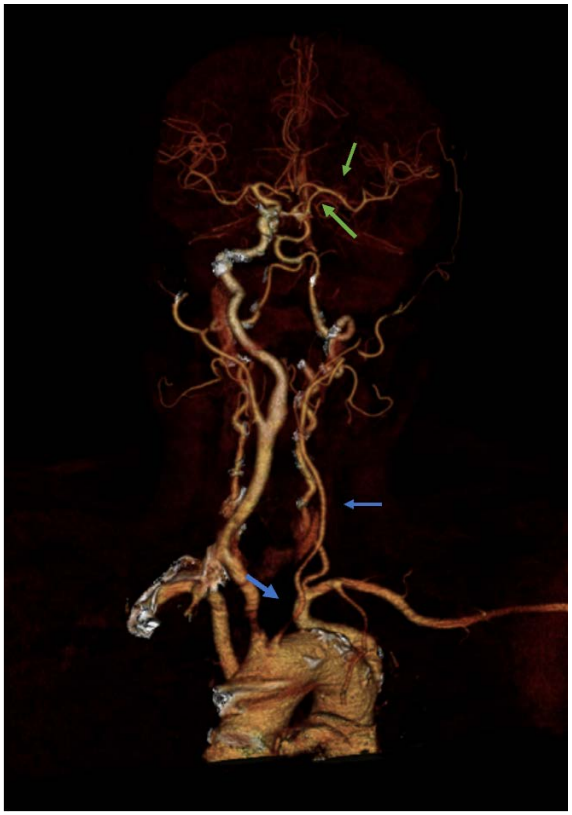


Figure 4:
Findings: Volume rendering AP image redemonstrating the midline right carotid bifurcation and proximal ICA, the hypoplastic left ICA (blue arrows), the absence of the left petrous and cavernous left ICA segments, and the intact vertebrobasilar system. Note the patency of the left supraclinoid ICA, as well as continuous opacification of the anterior and middle cerebral arteries supplied by collaterals (green arrows).

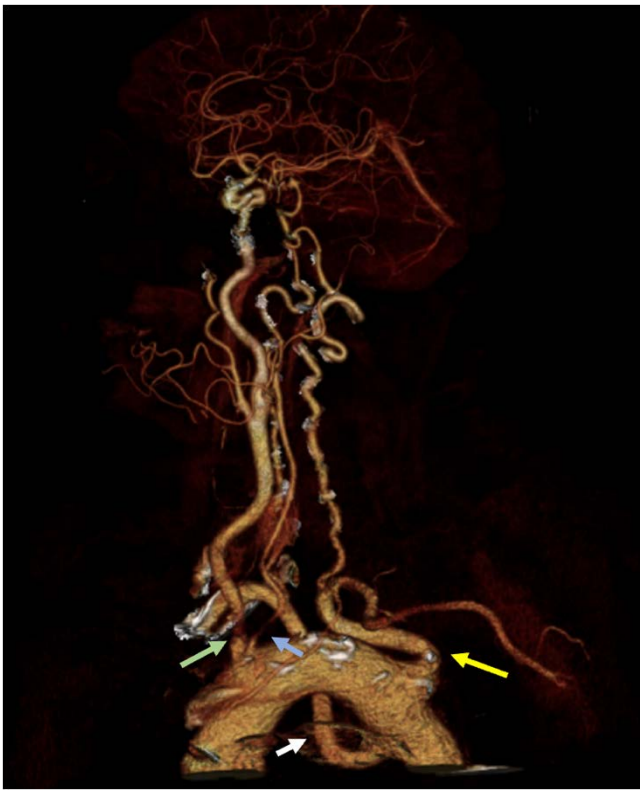


Figure 5:
Findings: Volume rendering image showing the right common carotid artery (green arrow) arising normally as the first branch off the aortic arch. The left common carotid artery (blue arrow) arises normally as the second aortic branch but is noted to be a diminutive vessel, terminating in left external carotid artery branches. The left subclavian artery (yellow arrow) arises from the lateral wall of the descending aorta in an anomalous low position, coursing retrograde, medially, and parallel to the aorta to its normal anatomic position prior to coursing laterally to its normal subclavicular position. The aberrant right subclavian artery (white arrow) arises from the medial wall of the descending aorta, coursing posterior to the aorta to its normal subclavicular position.

SUMMARY TABLE

	Etiology	**Incidence**	**Gender Ratio**	**Age Predilection**	**Risk Factors**	**Treatment**	**Prognosis**	**Findings on Imaging**
Congenital Absence of ICA	Failure of the third aortic arch and the distal aorta to involute	<0.01% of population	No known gender ratio	Congenital anomaly most often discovered incidentally	No known risk factors	Most often asymptomatic	Increased frequency of aneurysm formation in these patients. Implications during carotid endarterectomy and transsphenoidal surgery.	Absence of ICA and bony carotid canal
Aberrant Right Subclavian Artery	Right aortic arch regresses between, rather than distal to, the common carotid and right subclavian arteries	0.4 – 1.8% of population	Possible predominance in females	Congenital anomaly most often discovered incidentally	No known risk factors	Most often asymptomatic, surgical intervention is an option if symptomatic	Poses a substantial risk of hemorrhage in patients undergoing surgery in the region such as esophagectomy. Also important when considering cardiovascular surgery.	Aberrant right subclavian artery arises on its own as the fourth branch distal to the left subclavian artery. Aberrant artery then courses to the right side most often posteriorly to the esophagus
Aberrant Left Subclavian Artery	Interrupted movement of the seventh intersegmental artery cranially during embryogenesis	<0.05% of population	No known gender ratio	Congenital anomaly most often discovered incidentally	No known risk factors	Most often asymptomatic unless associated with other cardiac abnormalities	Asymptomatic unless associated with other cardiac abnormalities	Left subclavian artery arises more distally from the descending aorta than its normal origin

KEYWORDS

Congenital absence of ICA; Aberrant right subclavian artery; vascular anomaly; Arteria lusoria; Aortic arch anomaly; Agenesis of ICA

ABBREVIATIONS

ICA = Internal Carotid Artery

ARSA = Aberrant Right Subclavian Artery

MRI = Magnetic Resonance Imaging

CTA = Computed Tomography Angiography

MCA = Middle Cerebral Artery

TEVAR = Thoracic Endovascular Aortic Repair

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