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Case Report

Congenital absence of the internal carotid artery with intercavernous anastomosis

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ABSTRACT

Congenitally absent internal carotid artery with intercavernous anastomosis is an exceedingly rare vascular anomaly. We report such a case in a 65-year-old man with chronic symptoms resembling sporadic transient ischemic attacks. While these patients are usually asymptomatic due to compensatory collateralization, they are at increased risk of aneurysm formation, and thus proper identification is important. This report serves as both a case of a very rare anomaly and as a lesson on how to avoid this misdiagnosis by carotid duplex sonography. Additionally, we review the limited number of published cases of congenitally absent internal carotid artery with intercavernous anastomosis.

Introduction

Congenital absence of the internal carotid artery (ICA) occurs in less than 0.01% of the population [1]. These anomalies are most often asymptomatic due to the level of compensatory collateralization [2]. Lie first categorized 6 pathways of collateral circulation, with most cases involving collateral blood supply from the Circle of Willis (Types A-C) [3]. Specifically, Type D involves unilateral agenesis of the cervical ICA with collateral supply from an intercavernous anastomosis from the cavernous segment of the intact contralateral ICA, and this type is extremely rare. Types E and F involve collateralization from the posterior circulation and external carotid arteries, respectively. The purpose of this paper is not only to provide a brief literature review of this very rare Type D ICA agenesis, but also to address the pitfalls of carotid duplex exemplified by this patient's work up.

Case report

A 65-year-old male presented with several years of sporadic episodes of collapse when getting out of bed in the morning. During these episodes, the patient would maintain consciousness however, he would experience several minutes of self-resolving quadriplegia. To work up these TIA-like symptoms, the patient underwent an initial carotid duplex which was interpreted as right ICA occlusion, and left ICA 60%-79% stenosis. The symptomatic patient was planned for left ICA endarterectomy, however preoperative CTA of

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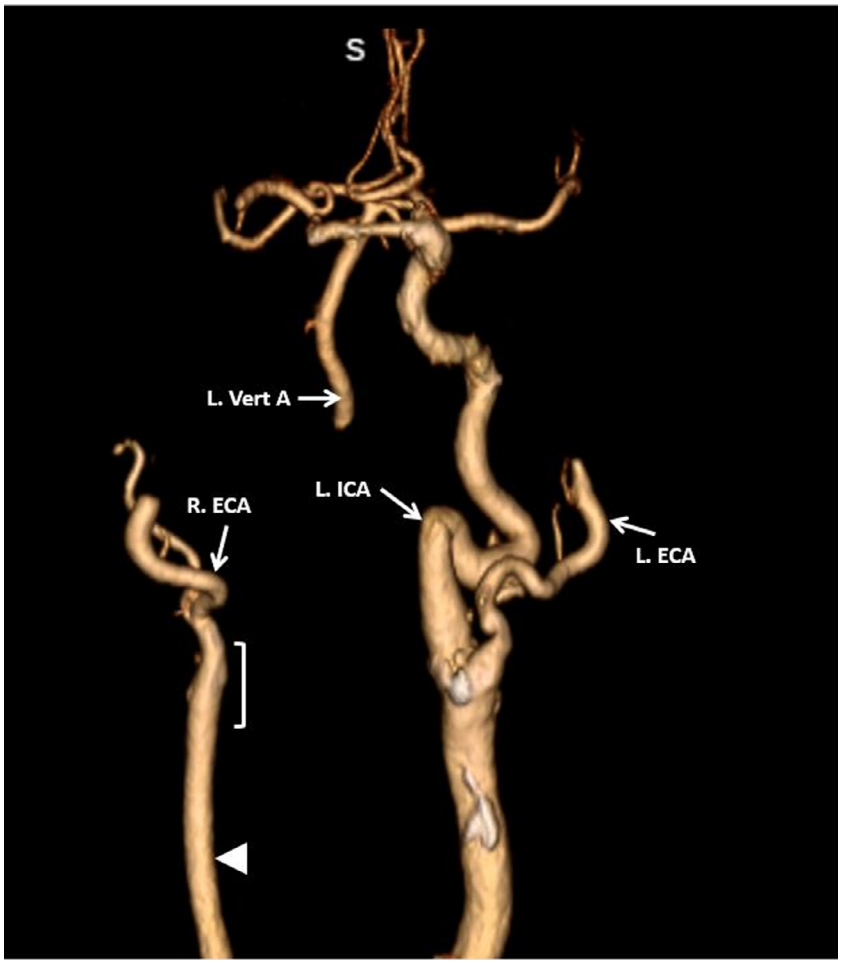

Fig. 1 – 65-year-old male with congenital absence of the right ICA. AP view of 3D rendering from CTA of the neck and brain demonstrating relatively small right CCA (arrowhead), absence of carotid bifurcation (bracket), and continuation as ECA (labeled). The left ICA is widely patent although tortuous (labeled). CCA, common carotid artery; ECA, external carotid artery; ICA, internal carotid artery; Vert, vertebral artery.

the neck demonstrated absence of the R ICA, and a widely patent L ICA (Fig. 1). The patient's right middle cerebral artery received its dominant supply from an intercavernous anastomosis, and the right anterior cerebral artery was supplied by the anterior communicating artery (Fig. 2). A smaller right posterior communicating artery was also identified (Fig. 2). Examination of the skull base on CT demonstrated absence of the right petrous carotid canal, confirming agenesis of the right ICA (Fig. 3).

The preoperative CTA thus revealed several errors in the initial carotid duplex interpretation. The right ICA was erroneously interpreted as occluded based on nonvisualization. However, pertinent findings (discussed in detail below) which should have argued against ICA occlusion included (1) a low resistance ipsilateral CCA waveform (Fig. 4) and (2) comparable peak systolic velocity (PSV) and end diastolic velocity (EDV) within the right and left CCA (Figs. 4, and 5a). The left ICA was also misinterpreted as having significant stenosis based on an elevated PSV (258 cm/s) (Fig. 5b) however nonvisualization of plaque or stenosis should have suggested an artifactually elevated PSV.

The patient was identified as having a COPD exacerbation which likely contributed to his most recent episode of cerebral hypoxia. Informed consent was obtained for the publication of this report. IRB approval was waived in the reporting of this case.

Discussion

ICA agenesis

The mechanism of ICA agenesis is hypothesized, with multiple papers attributing the anomaly to a mechanical stress (eg amniotic bands) or excessive bending of the cephalic portion of the embryo during development [4,5]. The ICA is first formed in the 3-5 mm embryonic stage and is complete by 6 weeks gestation (roughly 13 mm) while the circle of Willis is formed between the 7- and 24-mm embryonic stages [4]. Thus, disruptions prior to the 24 mm embryonic stage will result in primitive pathways of collateral circulation such as the

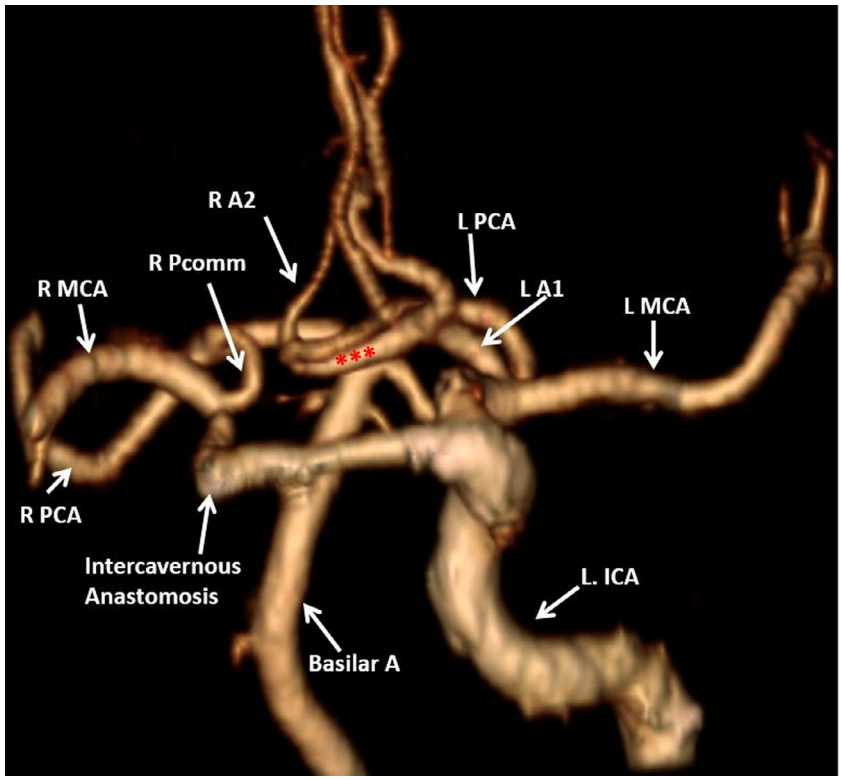

Fig. 2 – AP view of 3D rendering of intracranial vasculature from CTA of brain demonstrating an intercavernous anastomosis from the cavernous portion of the left ICA (labeled), supplying the right middle cerebral artery (labeled). The right anterior cerebral artery is supplied by the anterior communicating artery (asterix). A1/A2: A1 and A2 segments of the anterior cerebral artery. ICA, internal carotid artery; MCA, middle cerebral artery; PCA, posterior cerebral artery; Pcomm, posterior communicating artery.

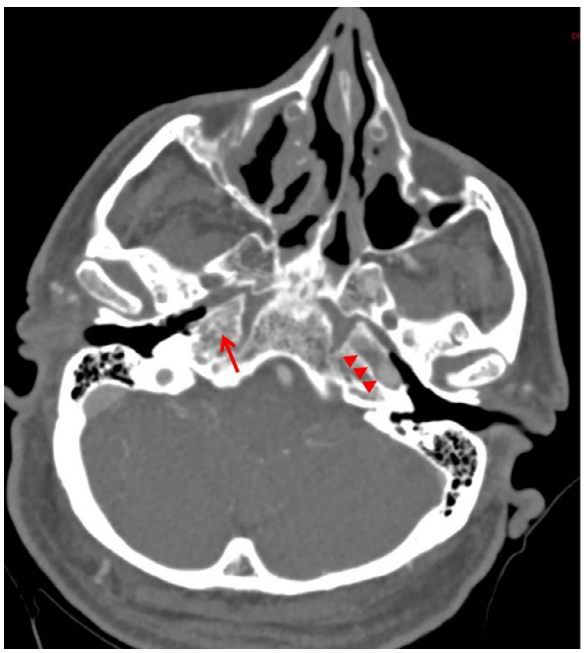

Fig. 3 – Axial CT through the skull base demonstrates absence of the right petrous carotid canal (arrow) in contrast to the patent left carotid canal (arrowheads).

intercavernous anastomosis seen here while later disruptions lead to collateralization from the circle of Willis [4]. The skull base forms at 5-6 weeks and thus in true congenital absence of the ICA, the carotid canal should be diminutive, serving as an essential component of the diagnosis [4]. Skull base CT is thus necessary to confirm ICA agenesis (vs aplasia, hypoplasia, or occlusion) [6].

In cases of ICA agenesis, collateral circulation is generally sufficient to maintain cerebral perfusion, however these patients are at risk for transient ischemic attacks as experienced by the patient described here [7]. These patients also have a higher incidence of circle of Willis aneurysm (25%-43%) and thus proper diagnosis of ICA agenesis is vital [8,9]. Specifically, knowledge of a Type D anastomosis has important surgical implications during transphenoidal surgery, in thromboembolic disease, and during carotid endarterectomy procedures [1]. Upon identification of ICA agenesis, patients should be followed to monitor existing aneurysms and to screen for the development of new ones [1].

In the very rare type of ICA agenesis (Lie's "Type D"), the MCA is supplied by an intercavernous anastomosis, with this anomalous vessel being transphenoidal, intrasellar, or supraclinoid [4]. Reports of this conformation with collateralization via an intercavernous anastomosis were reviewed by Oz et al in 2016, including a total of 36 patients [1]. In this cohort, 21 were male, and 14 were female; 21 patients had

Table 1 – Newly reported cases of unilateral internal carotid artery (ICA) absence with intercavernous anastomosis.

Case	First author	Year	Age	Gender	Absence	Symptoms
1	Alurkar	2016	39	F	Right	Severe headache after aneurysm rupture
2	Duan	2016	59	M	Right	Fatigue and dysphasia
3	Erdogan	2017	58	M	Left	Facial numbness, difficulty in speaking, and weakness of the left
4	Babichev	2018	30	F	Left	Periodic headache
5	Park	2018	32	F	Left	Right side weakness, pulsatile headache
6	Chen	2018	65	M	Right	Transient quadriplegia

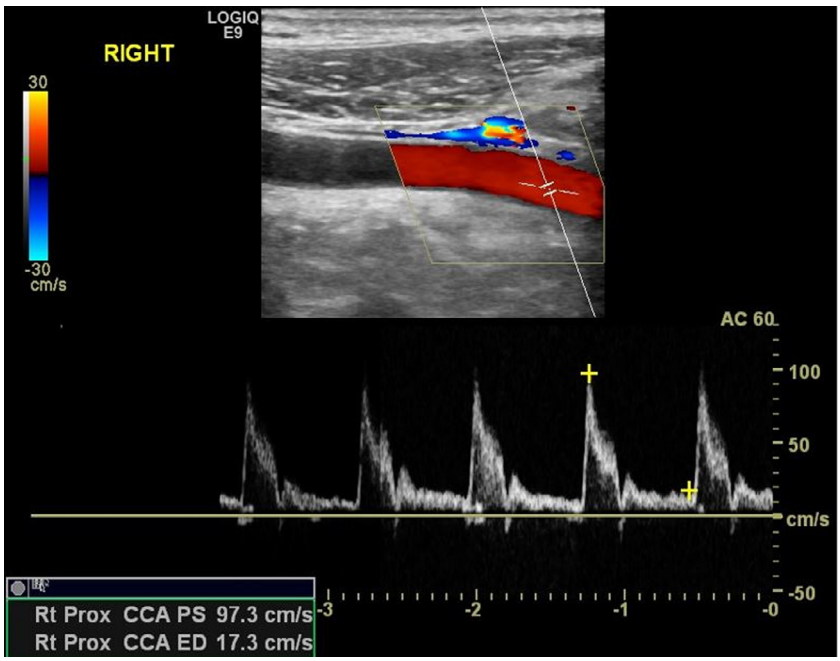

Fig. 4 – Spectral Doppler image of the R CCA, demonstrating low resistance waveform. The right CCA PSV and EDV match those values seen on the patient's left side, shown in Fig. 5.

right-sided, and 15 patients had left-sided congenital absence. The authors thus concluded that there was preponderance for gender or side of abnormality in cases of ICA agenesis with intercavernous anastomosis.

In the last 2 years, at least 5 additional cases have been reported apart from the case described here [4,5,10,11,7]. To add to the 36 cases presented by Oz, combined information on documented sex, laterality of the lesion, age at diagnosis, and presenting clinical signs/symptoms from these cases are collated in Table 1. The 6 new cases are split evenly among gender and laterality.

In the total 42 cases of congenital absence of ICA with Type D configuration, 24 (57%) are male and 17 (43%) are female; 24 (57%) cases have right sided congenital absence and 18 (43%) cases are left sided. With these additional cases, there remains no significant difference for gender or side preference with a 2-tailed test ($P > .05$; $z = 0.93$ and 0.77, respectively). The rate of right-sided ICA agenesis was 62.5% in males and 52.9% in females. There remains no difference between males and females for side of congenital absence ($P = .54$).

Sonographic work up of ICA

In the context of stroke or TIA symptoms, patients will often undergo a work-up including a carotid duplex, and misinterpretation should be avoided. As an example, this patient was scheduled for an unnecessary surgery due to misinterpretation of a carotid duplex.

The right ICA was interpreted as occluded based on nonvisualization, however a closer inspection of the Doppler waveform would provide more information. It has been reported previously that when an ICA is not visualized on carotid duplex, a low resistance CCA waveform, as seen in this patient, argues against ICA occlusion (Fig. 4) [2]. This low resistance ipsilateral CCA waveform suggests an ECA collateralization though one was not definitely seen on follow-up CTA. Not surprisingly, the right ECA waveform was also low resistance in this patient. In addition, in ICA occlusion, there is a characteristic "to-and-fro" flow pattern at the point of occlusion, not seen in this patient [12]. Also, nonvisualization of the carotid bifurcation in this patient should have raised suspicion for ICA

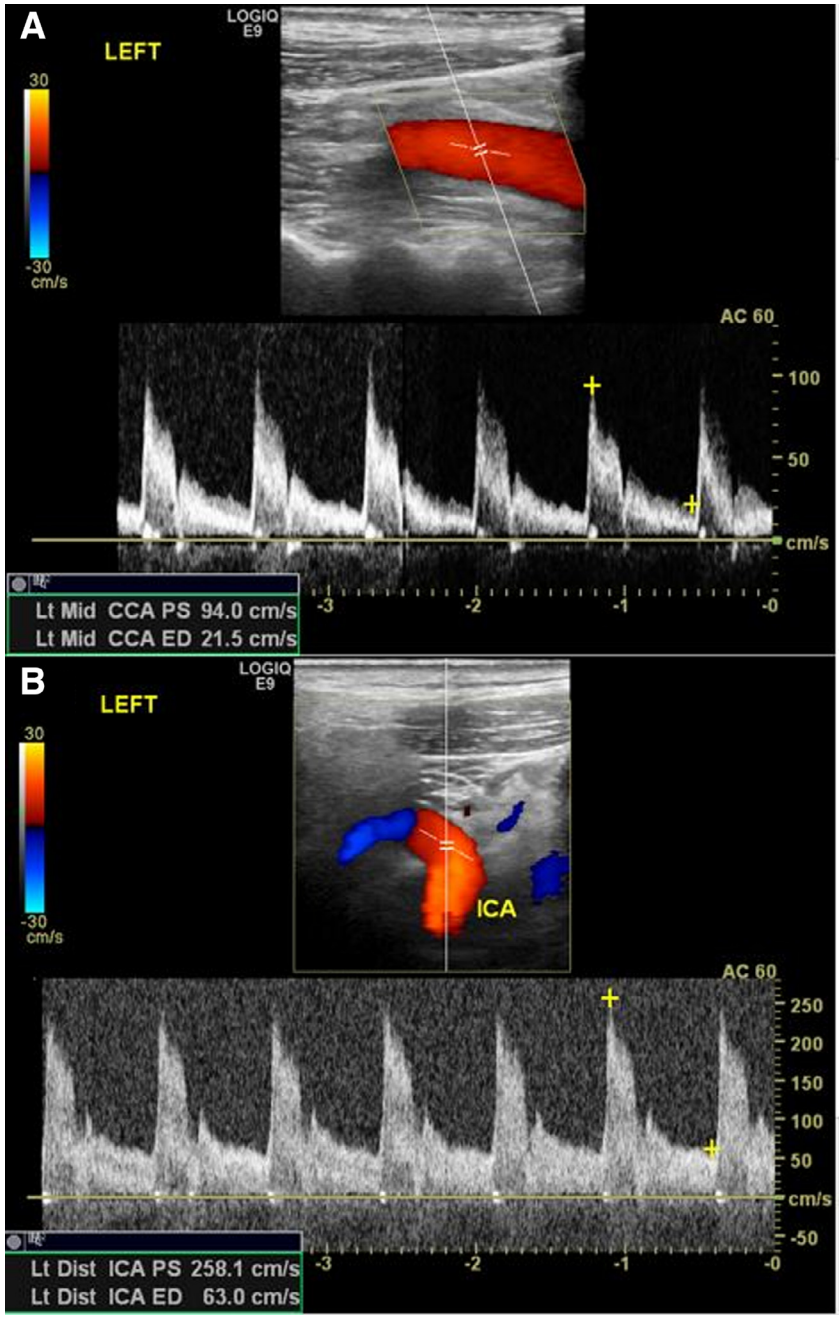

Fig. 5 – Spectral Doppler image of the L CCA and L ICA demonstrating peak systolic velocities of 94 cm/s and 258 cm/s, respectively (ICA/CCA ratio of 2.66). No evidence of stenosis seen in the tortuous ICA, with velocity measured at an appropriate Doppler angle of 60°.

agenesis [13]. Moreover, in ICA occlusion, there is usually significantly decreased or even reversal of EDV with reduced PSV of the ipsilateral CCA [14]. Our patient's R CCA PSV and EDV closely match those values measured on the left, again arguing against R ICA occlusion (Figs. 4, and 5a). Lastly, Kaya et al described a hypoplastic ipsilateral CCA as a sonographic predictor of ICA agenesis, and in our patient, the right CCA was 5 mm and the left CCA was 9 mm when measured at the middle third.

The left ICA was interpreted as having significant stenosis (60%-79%, with symptoms) as the PSV and EDV were both elevated (258 cm/s and 63 cm/s, respectively) (Fig. 5b). The ICA/CCA velocity ratio was also elevated (2.66). However, this case serves as a reminder that both the Society of Radiologist in Ultrasound consensus criteria and NASCET Index indicate that velocity measurements alone are insufficient to determine the degree of stenosis [15,16]. Visible plaque/narrowing should be identified to make the diagnosis, and our patient's

left ICA was widely patent, even on ultrasound imaging. We believe the increased velocity to be attributable to the tortuosity of the measured ICA. Indeed, vessel tortuosity producing falsely elevated velocities increases the false-positive rate for stenosis on Doppler ultrasound [12,17]. The color Doppler presented here (ICA) is devoid of color aliasing and color bruit artifact in surrounding tissue which are further signs of high grade stenosis [12].

Conclusion

ICA agenesis with intercavernous anastomosis is very rare, and proper diagnosis and differentiation from ICA stenosis or occlusion is important. Although there remains a small number of reported case of Lie's Type D conformation, reevaluation of the literature again demonstrates no predominance of Lie's Type D conformation for gender or side of agenesis. Although examination of the carotid canal is essential for the diagnosis of ICA agenesis, misdiagnosis by carotid duplex can be avoided with proper evaluation.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2019.05.030.

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