

Letter to the Editor

Internal Carotid Artery Agenesis with an Intercavernous Anastomosis: A Rare Case

Background: Agenesis of the internal carotid artery (ICA) is a rare vascular anomaly that was first observed postmortem. Various anastomoses supply the distal vessels at the site of agenesis. Of these anastomoses, an intercavernous anastomosis is very rare. This paper presents a patient with ischemic stroke in whom we discovered left ICA agenesis and an ipsilateral intercavernous anastomosis. *Case:* A 58-year-old man with a history of myocardial infarction and diabetes mellitus presented with sudden-onset difficulty in speaking, numbness on the left side of the face, and weakness of the left arm and leg. Neurological examination revealed dysarthria, left facial paralysis, left hemiparesis, and bilateral absence of the plantar reflexes. Diffusion-weighted magnetic resonance imaging showed a right middle cerebral artery (MCA) infarction. On cranial and cervical magnetic resonance angiography, the left ICA could not be seen distal to the bifurcation; the left MCA was supplied through an intercavernous anastomosis between the right ICA and the left ICA. Cranial computed tomography (CT) revealed the absence of the left carotid canal. Digital subtraction angiography led to a diagnosis of left ICA agenesis with an intercavernous anastomosis. The patient was discharged on acetylsalicylic acid and warfarin. *Conclusion:* ICA agenesis with an intercavernous anastomosis is a rare vascular anomaly that should be differentiated from secondary causes of ICA stenosis and occlusions by showing agenesis of the carotid canal on cranial CT. **Key Words:** Agenesis of the internal carotid artery—intercavernous anastomosis—vascular anomaly embryogenesis.

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Dear Editor,

Agenesis, segmental aplasia, or hypoplasia of the internal carotid artery (ICA) is a rare vascular anomaly. It may be unilateral or bilateral, and it is seen in less than .01% of the population.¹ The distal ICA and intracranial arteries are supplied through various anastomoses when there is cervical ICA agenesis. Lie classified the collateral circulation into 6 different pathways. The most common collateral circulations (types A to C) develop via the circle of Willis. A type D anastomosis, a trans-sellar, or an intercavernous anastomosis is seen very rarely.² In this report, we present a patient who had an ischemic stroke and was found to have left ICA agenesis and an intercavernous anastomosis.

Case

A 58-year-old man presented with sudden-onset facial numbness, difficulty in speaking, and weakness of the left arm and leg. He had a history of myocardial infarction and diabetes mellitus and was taking acetylsalicylic acid irregularly. On neurological examination, he was alert, attentive, and followed commands. He had dysarthria and a flattened left nasolabial sulcus. Motor examination revealed mild paresis of the left arm and leg. The plantar reflex was absent bilaterally. He had hypoaesthesia on the left side of his face. His vital signs were normal. The physical examination was normal. His complete blood count, kidney and liver function tests, and infection markers were normal. There was no acute lesion on cranial computed tomography (CT). Diffusion-weighted magnetic resonance imaging showed acute infarction of the right middle cerebral artery (MCA). On further workup, we discovered left ICA occlusion on magnetic resonance angiography, which revealed an intercavernous anastomosis between the left ICA and the right ICA (Fig 1). A review of the cranial CT showed left carotid canal agenesis (Fig 2). To confirm the diagnosis,

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Figure 1. On magnetic resonance angiography, the left ICA is occluded proximally and is supplied through an intercavernous anastomosis arising from the right ICA. Abbreviation: ICA, internal carotid artery.



Figure 2. Axial cranial computed tomography showed left carotid canal agenesis.

digital subtraction angiography was performed and showed left-sided ICA agenesis and an intercavernous anastomosis (Figs 3 and 4). Electrocardiography showed normal sinus rhythm. The patient was discharged on acetylsalicylic acid and warfarin based on an ejection fraction of 45% on echocardiography.

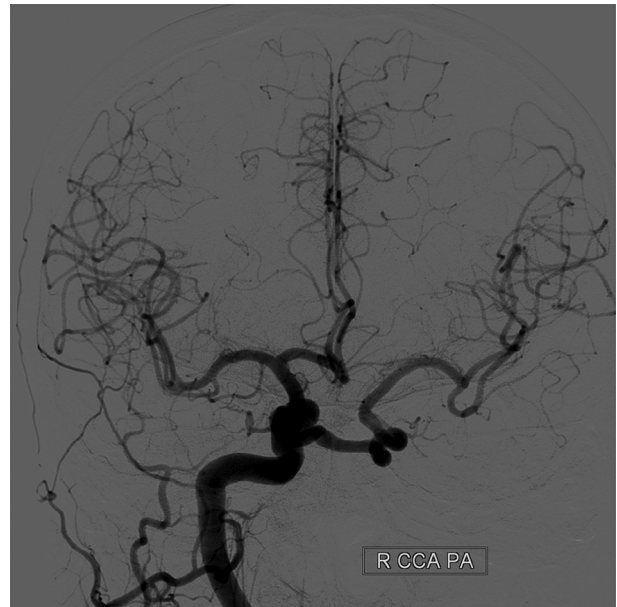


Figure 3. Intercavernous anastomosis is seen on the posteroanterior right CCA imaging in DSA. Abbreviations: CCA, common carotid artery; DSA, digital subtraction angiography; PA, posteroanterior; R, right.

Discussion

The formation of the ICA is first seen in the 3- to 5-mm embryonic stage, and it is completed in 6 weeks of gestation.³ Dorsal aorta, ventral aorta, and third aortic arch are the structures that give rise to the ICA.³ The formation of the circle of Willis takes place during the 7- to 24-mm embryonic stage.⁴ Mechanisms for failure of development of the ICA and the formation of various anastomoses are speculated. Keen suggested that excessive bending of the embryo to the one side or compression from amniotic bands may cause agenesis of the ICA on the affected side.⁵ Cali et al. postulated that the type of anastomoses is dependent upon the time at which the disruption to the embryo occurs. Disruptions before the 24-mm embryonic stage result in primitive anastomoses, whereas disruptions after this stage result in anastomoses via the circle of Willis.⁴

The origin of the anastomotic vessel is speculated. Lie stated that a failed connection between the basilar artery and primitive trigeminal arteries, and the fusion of the primitive trigeminal arteries on both sides were the origin of the anastomotic vessel.² Padget described plexiform channels around Rathke's pouch, which connects the ICAs, and thought that the origin of the anastomosis can be explained by these vessels.³ Sinha et al put forward that the contralateral ICA is filled via the primitive maxillary artery.⁶ Duan et al suggested that the anastomotic vessel is the same as the capsular artery because it originates from the horizontal segment of the ICA and courses medially in front of the cavernous sinus.⁷ In our case, the anastomotic vessel originated from the posterior of the cavernous segment of the right ICA and coursed anteriorly, and then medially

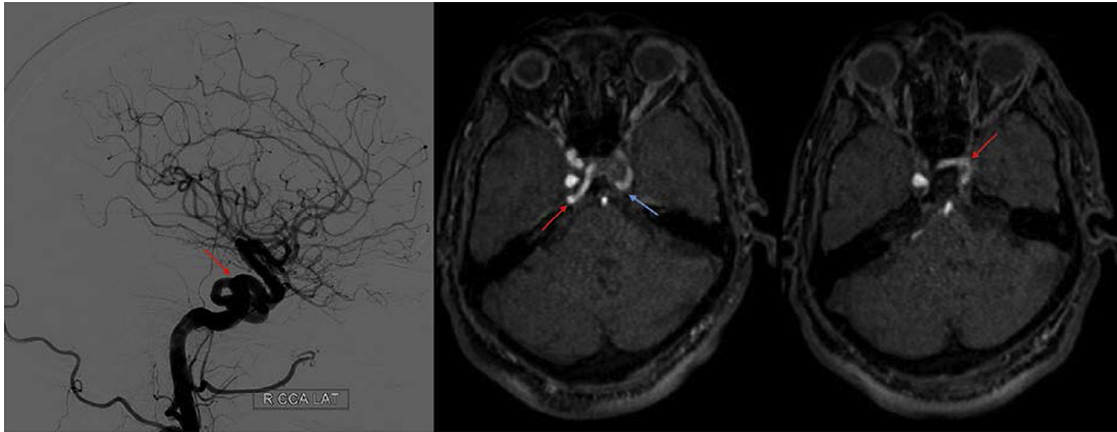


Figure 4. Posterior origin of the intercavernous anastomotic vessel is seen on the left side. After leaving the ICA, the anastomotic vessel courses medially and anteriorly (middle image) in front of the sella (on the right side). The anastomotic vessel then opens into the left ICA on its medial side (middle image). Abbreviations: CCA, common carotid artery; ICA, internal carotid artery; LAT, lateral; R, right.

in front of the pituitary gland, through the floor of the sella. The anastomotic vessel opened into the cavernous segment of the left ICA on the medial side. We believe that the anastomotic vessel does not originate from the primitive trigeminal artery or the primitive maxillary artery because it is related to the medial aspect of the ICA on the left side. The primitive trigeminal artery courses posteromedially, and the primitive maxillary artery courses anterolaterally, whereas our vessel courses anteriorly and medially. This finding also does not support the primitive trigeminal artery and primitive maxillary artery hypotheses. We think that plexiform vessels around the sella give rise to the intercavernous vessels.

Lie classified collaterals associated with ICA agenesis into 6 different groups.² The most common pattern is type A, in which the ipsilateral MCA is supplied via the posterior communicating artery (PCoM_A) arising from posterior cerebral artery, and agenesis of the ipsilateral A1 segment of the ACA is seen. In the type B pattern, the collateral circulation is achieved via the anterior communicating artery. Bilateral agenesis of the ICA is seen in the type C pattern, and the anterior circulation is supplied via the bilateral PCoM_As arising from the basilar artery. The type D pattern, which our patient had, is associated with an intercavernous anastomosis. The MCA is supplied via an intercavernous anastomosis between the intracavernous segments of the bilateral ICAs. In these patients, the anastomosis may be transsphenoidal, intrasellar, or supraclinoid.^{8,9} In type E anastomosis, both ICAs are hypoplastic, and they supply only ipsilateral ACAs. MCAs are supplied from the enlarged PCoM_As on both sides. Type F anastomosis is a condition in which distal flow is achieved via collaterals between ICAs and ECAs, and these collaterals are named as rete mirabile.²

In a literature review, Oz et al. summarized 36 intercavernous anastomosis cases involving 21 men and

15 women.¹⁰ The agenesis was right sided in 21 of these patients and left sided in 15. Zink et al. reported a 2:1 left predominance in ICA aplasia/hypoplasia regardless of the type of anastomosis, although they did not find any significant sex difference.¹¹ Another study reported male predominance in left-sided agenesis.¹² Oz et al. did not report a sex or side predominance in intercavernous anastomosis and stated that the small number of cases was insufficient to generalize the findings.¹⁰

Patients with ICA agenesis may be asymptomatic or present with different clinical situations, such as subarachnoid hemorrhage due to the rupture of an intracranial aneurysm or ischemic stroke.¹⁰ Intracranial aneurysms are seen 25%-43% of patients with ICA agenesis, which is much higher than that in the general population.¹¹ In type D anastomosis, an incidental diagnosis or nonspecific symptoms are frequent. Of the 36 cases summarized in Oz et al., 20 presented with nonspecific symptoms (headache, dizziness, tinnitus, and incidental symptoms), and 16 presented with cerebrovascular accidents (acute ischemia, subarachnoid hemorrhage, or vision loss).¹⁰

It is important to differentiate ICA agenesis with intercavernous anastomosis, a rare congenital anomaly, from ICA stenosis and occlusion related to secondary causes. Patients may be diagnosed after developing nonspecific symptoms or having cerebrovascular accidents. The absence of the carotid canal on cranial CT is important for diagnosing ICA agenesis. After diagnosis, patients need to be followed up regularly to check for aneurysms. Additionally, this anomaly may lead to serious complications in endarterectomy or transsphenoidal surgery.

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