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CASE REPORT

Unilateral Agenesis of Internal Carotid Artery with Intercavernous Anastomosis: A Rare Case Report

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ABSTRACT

Unilateral agenesis of internal carotid artery (ICA) with intercavernous anastomosis is a rare congenital anomaly. We present a case of a 25-year-old female with 2-month history of holocranial headache. Neurological examination was unremarkable. Magnetic resonance imaging (MRI) with magnetic resonance angiography (MRA) showed absence of left ICA with an abnormal intercavernous vessel in the sella. Computed tomography (CT) showed absence of the left carotid canal. Doppler ultrasonography (USG) showed high resistance flow in the left common carotid artery (CCA). Since no hemorrhage or aneurysm was seen, patient was managed conservatively and is on regular follow-up. Based on our knowledge, this is the first case to demonstrate the features of unilateral agenesis of ICA with intercavernous anastomosis in X-ray, Doppler USG, CT, and MRI scans of the brain.

Key words: Aplasia, internal carotid artery, unilateral agenesis

INTRODUCTION

Unilateral agenesis of internal carotid artery (ICA) is a rare congenital anomaly. In such cases, the anterior cerebral artery (ACA) and middle cerebral artery (MCA) on the side of agenesis are usually supplied by the basilar artery and the contralateral ICA through the circle of Willis.[1] But an abnormal intercavernous anastomosis is extremely rare. Patients are usually asymptomatic but a few patients may present with headache, subarachnoid hemorrhage (SAH) and transient ischemic attack (TIA).

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CASE REPORT

A 25-year-old female patient visited the neurology outpatient clinic with severe holocranial headache that had persisted for 2 months. The pain was intermittent and relieved by analgesics. Clinical examination was unremarkable.

Computed tomography (CT) scan of the brain showed absent bony carotid canal on the left side [Figure 1a and b]. Magnetic resonance angiography (MRA) [Figure 2a and b] showed complete absence of the cervical and petrous part of the left ICA. An abnormal vessel was seen arising from the cavernous segment of the right ICA. This vessel

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was passing across the floor of sella, reconstituting the cavernous and supraclinoid segments of the left ICA, and continuing as the left MCA. Aplasia of A1 segment of left ACA was also observed.

Magnetic resonance imaging (MRI) showed absent left ICA flow void with an abnormal flow void in the sella [Figure 3a and b]. No evidence of any SAH or aneurysm was seen. The vertebrobasilar system was normal.

Doppler imaging [Figure 4a-d] showed complete absence of the left ICA. The left common carotid artery (CCA) was seen continuous with the external carotid artery (ECA). The left CCA and ECA showed high resistance flow with narrow systolic peak and low diastole. Since the right CCA has to compensate for the absent left ICA, it showed low resistance flow with broad systolic peak and high diastolic flow.

Lateral radiograph of the skull [Figure 5a] and the corresponding sagittal reformatted CT scan [Figure 5b] showed a well-defined lytic area in the anteroinferior aspect of the sella due to the abnormal intercavernous vessel traversing across it.

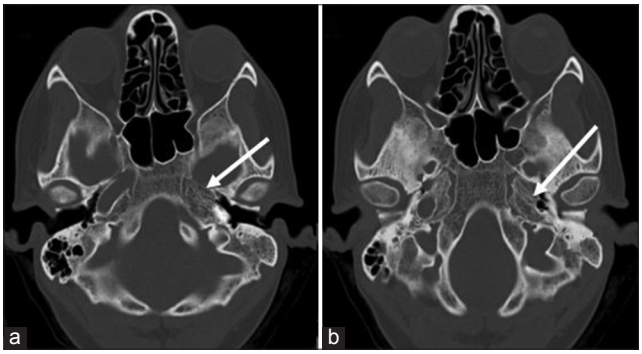

Figure 1: 25-year-old female presented with holocranial headache and was diagnosed with unilateral agenesis of internal carotid artery with intercavernous anastomosis. Axial CT scan (a and b) Two consecutive sections at the base of skull show absent bony carotid canal on the left side (arrows).

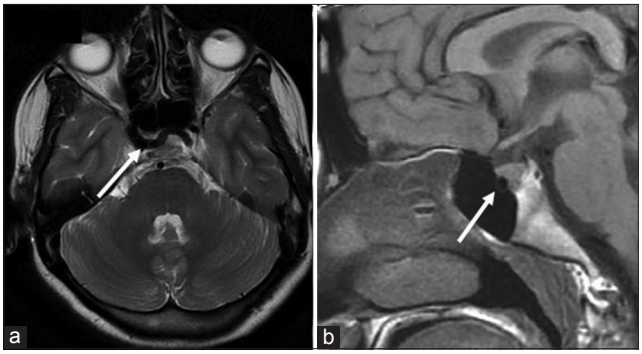

Figure 3: 25-year-old female presented with holocranial headache and was diagnosed with unilateral agenesis of internal carotid artery with intercavernous anastomosis. (a) Axial T2-weighted sequence shows an abnormal vessel running across the sella (White arrow). (b) Sagittal T1-weighted sequence shows abnormal flow void (arrow) in the floor of sella and mildly bulging into the sphenoid sinus, representing the intercavernous anastomotic vessel, which can cause devastating complication during a transphenoidal pituitary surgery.

DISCUSSION

Agenesis of ICA is a rare anomaly. Male predominance has been seen with slightly increased incidence on the left side. Patients with agenesis of ICA are usually asymptomatic due to the collateral circulation from the contralateral ICA and the vertebrobasilar system via the circle of Willis.[1]

In 1787, Tode documented the first case of carotid agenesis, identifying it on postmortem examination. In 1954, the first case of ICA agenesis at cerebral angiography was reported by Verbiest.

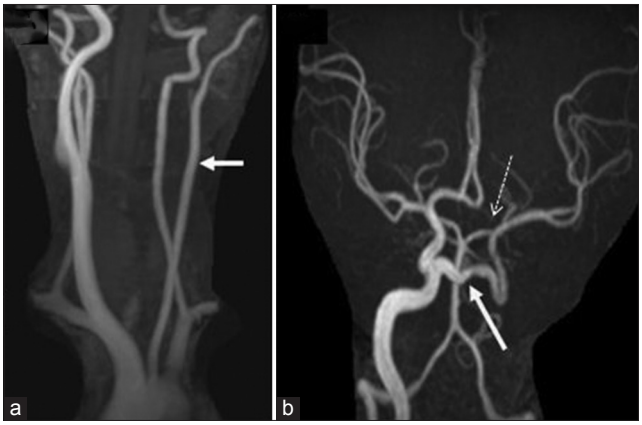

Figure 2: 25-year-old female presented with holocranial headache and was diagnosed with unilateral agenesis of internal carotid artery with intercavernous anastomosis (a and b) Time of flight (TOF) MRA with maximum intensity projection (MIP) of the neck vessels and the intracranial arteries show complete absence of the cervical and petrous part of the left ICA with left common carotid artery continuing as external carotid artery (arrow in a). MRA with MIP of intracranial arteries shows an abnormal vessel (arrow in b) seen arising from the cavernous segment of the right ICA and reconstituting the cavernous and supraclinoid segments of the left ICA, and continuing as the left MCA. A1 segment of left ACA is not seen (Dotted arrow) suggesting aplasia.

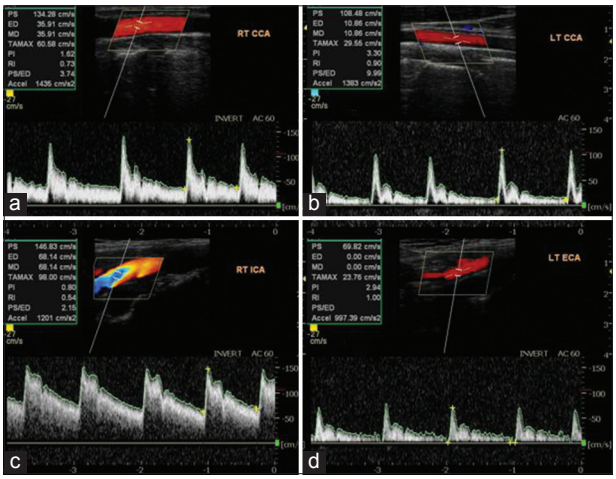

Figure 4: 25-year-old female presented with holocranial headache and was diagnosed with unilateral agenesis of internal carotid artery with intercavernous anastomosis (a-d) Duplex Doppler scans with spectral waveform of the (a) right CCA, (b) left CCA, (c) right ICA, and (d) left ECA show high velocity and low resistance flow in the right ICA because it has to compensate for the absent left ICA. Low resistance flow is also seen in the right CCA. High resistance flow with externalization is seen in the left CCA.

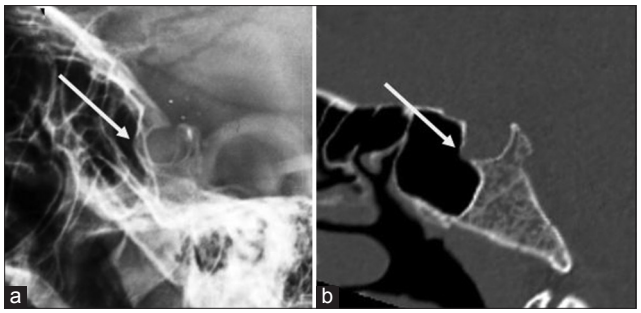

Figure 5: (a) 25-year-old female presented with holocranial headache and was diagnosed with unilateral agenesis of internal carotid artery with intercavernous anastomosis. (a) Lateral radiograph of the skull (zoomed image) and (b) sagittal reformated CT scan (bone window) show a well defined lytic area (arrows) in the anteroinferior aspect of the sella due to the abnormal intercavernous vessel traversing across it.

Certain mechanical and hemodynamic stresses over the embryo like exaggerated folding of embryo toward one side and constriction by amniotic band have been postulated as hypothesis for agenesis.[2,3]

A few patients occasionally present with headache, SAH, and TIA. Associated abnormalities include vascular aneurysm. Rarely, transsphenoidal encephalocele and rete mirabilis in the cranial base are seen.[4]

Agenesis, aplasia, and hypoplasia of ICA form a spectrum, with the agenesis showing complete absence of ICA and the carotid bony canal, whereas hypoplasia shows a small bony carotid canal. Absence of bony carotid canal on CT scan is a characteristic feature of agenesis of ICA.

The circle of Willis is formed during the 7- to 24-mm stage of embryonic development. The pattern of collateral blood supply and intracranial vasculature is dependent on time at which insult happened. The ICA is developed from seven distinct segments and agenesis of any one segment will lead to regression of all the segments proximal to it.[5]

According to Lie, there are six pathways of collateral circulation in association with absence of the ICA.[5] These are:

Type A – Unilateral absence of the ICA with collateral circulation to the ipsilateral ACA through a anterior communicating artery (ACOM) and to the ipsilateral MCA from the hypertrophic posterior communicating artery (PCOM)
Type B – Unilateral absence of ICA with collateral flow to the ipsilateral ACA and MCA across a patent ACOM artery
Type C – Bilateral ICA agenesis with supply to the anterior circulation via carotid-vertebrobasilar anastomoses and through hypertrophic PCOM
Type D – Unilateral agenesis of the cervical portions of the ICA with collateral from an intercavernous communication from the cavernous segment of contralateral ICA
Type E – Bilateral hypoplastic ICAs supplying ACA and the MCAs are supplied by enlarged PCOMs
Type F – Flow to ICA across transcranial anastomoses from the branches of the ECA system, the so-called rete mirabile.

The simplified form of Lie's original six collateral pathways are: Collateral flow through the circle of Willis (most frequent), Collateral flow via persistent fetal circulation, and Reconstitution of the ICA through skull base collaterals from the ECA.

Hypertrophy of normally present embryologic vessels or their persistence can explain the collateral circulation in types A, B, C, and E. However, in Lie's type D intercavernous communication between the ICAs could not be explained as there is no *in utero* communication observed between these arteries.[6]

Various theories have been put forward regarding the development of abnormal intercavernous vessel. According to Lie, the collateral vessel is thought to arise from a union between two primitive trigeminal arteries that have lost their communication with the basilar artery.[5]

Another theory states that this may be due to hypertrophy of few embryologically developing vessels, like plexiform network surrounding Rathke's pouch or remnants of the primitive maxillary artery.[5,7,8] These differing theories may answer the variable course of intercommunincating artery in relation to the sella turcica. The intercavernous channel can pass along the floor of sella, above the sella or posterior to clivus.

Also according to Lie in type D collateral pathway, supply to the ipsilateral ACA and MCA is derived from both the intercavernous communication and a patent ACOM.[5,6] This concept differ from the few reported cases of this intercavernous vessel.[3,7] The reported cases demonstrate that when an intercavernous communication exists, there is associated hypoplasia or aplasia (Aplasia in our case) of the ipsilateral A1 segment of the ACA with ACOM supplying the ipsilateral distal ACA territory. Thus the intercavernous communication supplies the majority of the ipsilateral MCA supply making the hemisphere isolated from alternate routes of collateral flow.

Nearly, 18 cases of unilateral agenesis of ICA with intercavernous anastomosis are reported in the literature. In adults presenting with thromboembolic disease, diagnosis of this anomaly is important as emboli in one

cerebral hemisphere can be explained by atherosclerotic disease in the contralateral carotid artery or vertebrobasilar system. It is also important when planning for carotid endarterectomy, as both cerebral hemispheres may be dependent upon the atheromatous carotid artery. On imaging, the transsellar course of this vessel has been known to mimic a pituitary microadenoma.

There are other different anatomic variations involving the ICA. They can be aberrant ICA, persistent carotid vertebrobasilar anastomosis, or retropharyngeal tortuous ICA.

In aberrant ICA, due to the agenesis of the cervical segment of ICA the inferior tympanic branch of the ECA and the carotico-tympanic branch of petrous ICA are enlarged and the ICA is reformatted from the petrous segment. Multidetector computed tomography (MDCT) show hypoplasia/aplasia of the vertical segment of carotid canal as well as reduced caliber and lateralization of the ICA with protrusion of the artery into the middle ear. Clinical examination of the ear can mimic glomus tumor and other vascular lesions like dehiscent jugular bulb, cholesterol granuloma, petrous carotid aneurysms, pseudoaneurysms, and hemangiomas.[9]

Persistent carotid vertebrobasilar anastomosis include persistent trigeminal artery arising from the proximal cavernous ICA, persistent otic artery arising from the petrous ICA, persistent hypoglossal artery arising from the cervical ICA, and the persistent proatlantal artery.

More importantly, the ICA can be tortuous and course medially bulging into the retropharyngeal space. On clinical examination it appears as a pulsating submucosal mass along the posterior pharyngeal wall. Potential life-threatening complications can occur during intubation, endoscopy, and invasive procedures in the oropharynx like tonsillectomy or retropharyngeal abscess drainage.[10]

CONCLUSION

Agenesis of ICA is a rare anomaly. The finding of absent ICA flow void on routine MRI and absence of bony carotid canal on routine CT should suggest the diagnosis. It is important to identify the transsellar intercavernous anastomosis on imaging to avoid devastating complications during transsphenoidal pituitary surgery.[3] It is also important in the management of cerebrovascular accidents as the single ICA supply both the cerebral hemispheres. Other associated intracranial lesions like aneurysms should be looked for and managed appropriately.

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