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Congenital Agenesis of the Unilateral Internal Carotid Artery Misdiagnosed as Occlusion or Stenosis on Time of Fight (TOF) MR Angiography & Importance of CT Scan Correlation which Make Difference in the Management Plan of Patient: A Case Report & Review of the Literature

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Abstract
A 58-year-old-male presented with episodes of numbness left sided face, arm & leg for few seconds from last 2-3 months & few episodes of transient ischaemic attacks (TIA's) in past. Physical examination showed high blood pressure (174/74). Case was misdiagnosed as occlusion or stenosis of internal carotid artry on magnetic resonance angiography utilizing the time-of-flight technique, but suspicion of ICA agenesis was raised by the absence of the typical flow void in the ICA region, on the spin-echo sequences (T2W). This finding was confirmed by magnetic resonance angiography of the cervical vessels, and axial computed tomography. CT scan showed agenesis of the left carotid canal. On magnetic resonance angiography of the cervical vessels, there was associated findings like hypoplastic right vertebral artery with accessory vertebral/collateral & complete occlusion or agenesis of right external carotid artery at its origin & atherosclerotic changes in right internal carotid artery.
Keywords: *Internal carotid artery; Agenesis; Transient ischaemic attack; Misdiagnosed; Occlusion or stenosis.*

Introduction

Unilateral agenesis of the internal carotid artery (ICA) is an extremely rare congenital anomaly with an incidence of 0.01% [(1–3)]. The absence of an ICA was first described in 1787 after being discovered during a post-mortem examination [(1, 4)]. In 1954, Verbiest reported a case with ICA agenesis recognised by cerebral angiography [(5)]. The cause of the carotid agenesis is not known, but may be secondary to an insult to the developing embryo [(19)]. When a unilateral ICA is absent collateral circulation is sufficient to maintain cerebral function with few or no neurologic symptoms[(3,6)]. Most patients with ICA agenesis present with focal neurologic signs, such as convulsions, headache, or transient ischemic attack, and ICA agenesis may be associated with aneurysm, hemorrhage, cerebral hypoplasia, hemangioma, or anomalous vascular anastomosis. We report a case of congenital absence of the left internal carotid artery associated with hypoplastic right vertebral artery along with accessory vertebral or collateral & complete occlusion or agenesis of right external carotid artery at its origin & atherosclerotic changes in the right internal carotid artery which misdiagnosed as

occlusion or stenosis on MR angiography using time-of-flight technique, later confirmed as agenesis on CT scan.

Case Report

A 58-year-old male came to outpatient department with history of episodes of numbness left sided face, arm & leg for few seconds from last 2-3 months. He had history of few episodes of transient ischaemic attack in past. The patient was referred to a neurologist. The patient was evaluated by neurologist & advised imaging studies .

Case was diagnosed as occlusion or stenosis on MR angiography (TOF). The suspicion of ICA agenesis was raised by the absence of the typical flow void in the ICA region, on the spin-echo sequences (T2W) (Figure 1).Considering the suspicion of agenesis or hypoplasia of the right ICA, the patient was submitted to non-contrast enhanced computed tomography (CT) and MRI angiography of cervical vessels, for diagnostic confirmation. The CT study was aimed at differentiating a congenital from an acquired agenesis of the bony carotid canal, but the absence of the left carotid canal was observed. The MRI angiography of brain along with cervical vessels was performed in an attempt to depict a global picture of the cerebral arterial network, and the absence of the left ICA from the its origin (Figures 2, 3 and 4).

Additionally, the patient was submitted to multislice CT, in an attempt to demonstrate some other skull bone abnormality, but only a confirmation of the other studies findings was observed by means of the 3D imaging reconstr-uction with volume rendering, demonstrating the absence of the left carotid canal (Figure 5).

MRI angiography & Doppler study of cervical vessels further reveals complete occlusion or agenesis of right External carotid artery at its origin along with atherosclerotic changes in the right internal carotid artery & hypoplastic right vertebral artery along with accessory vertebral or collateral.

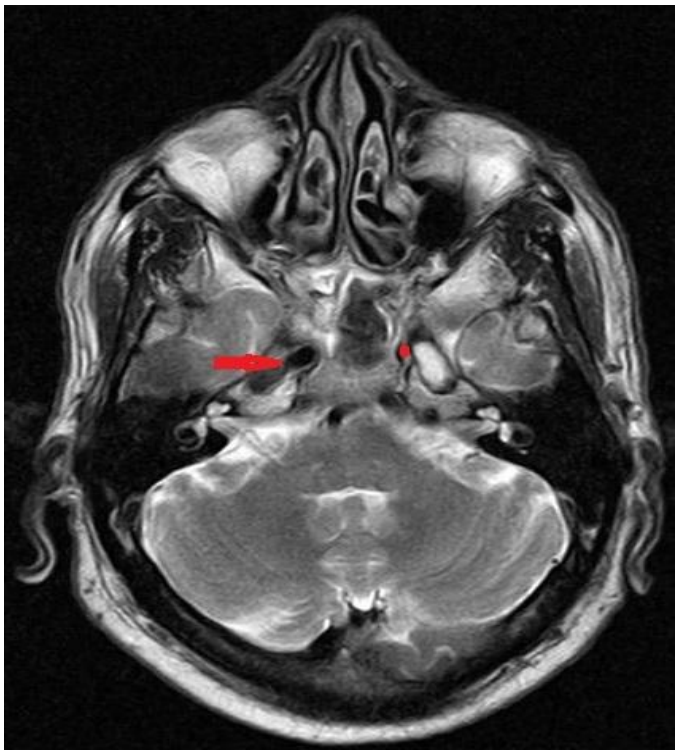

Figure 1 Axial MRI turbo spin- echo T2-weighted sequence showing absence of flow void in the left ICA (red dot), normal flow void in right ICA (red arrow).

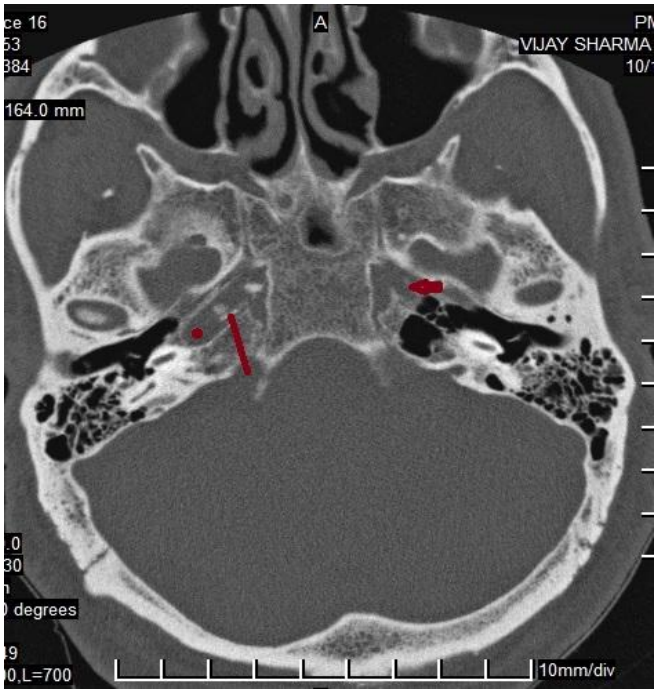

Figure 2 Non-contrast-enhanced, thin section axial CT, temporal bone window technique showing the absence of the left carotid canal (red arrow) with normal right carotid canal (red dot) atherosclerotic calcification in right ICA (red line).

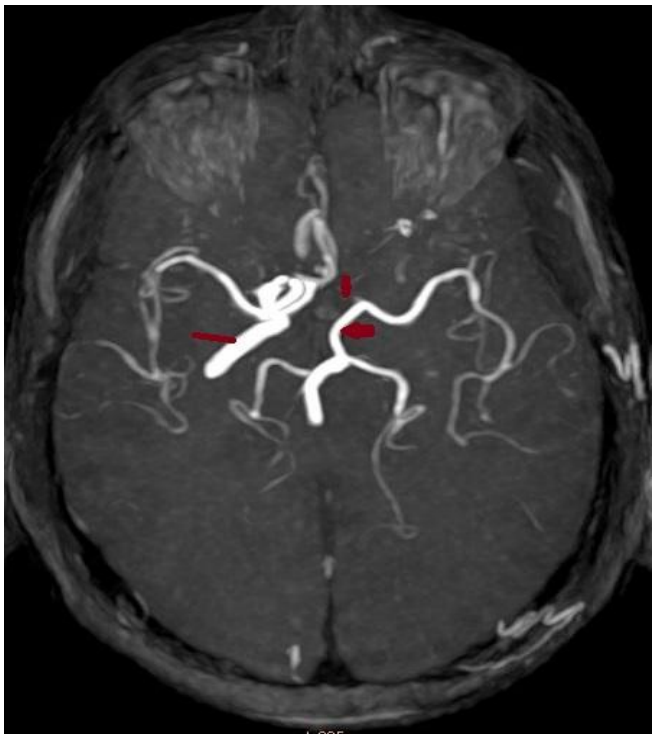

Figure 3 Cerebral MRI angiography (TOF) showing the left ICA absence with normal right ICA (large red line). It shows the basilar artery supplying the left middle cerebral artery via the left posterior communicating artery (red arrow). Absence of the A1 segment (small red line) is noted.

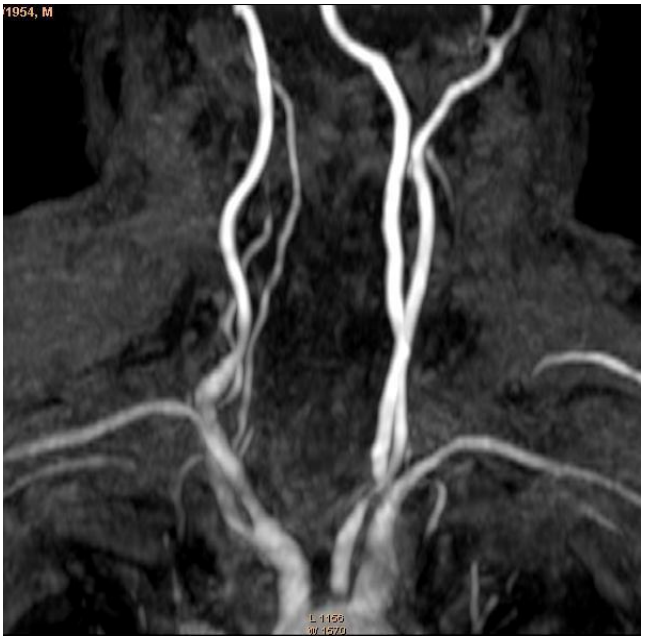

Figure 4 Cervical vessels MRI angiography (TOF) confirming the left ICA absence & right external carotid artery occlusion or agenesis with hypoplastic right vertebral artery along with accessory vertebral or collateral.

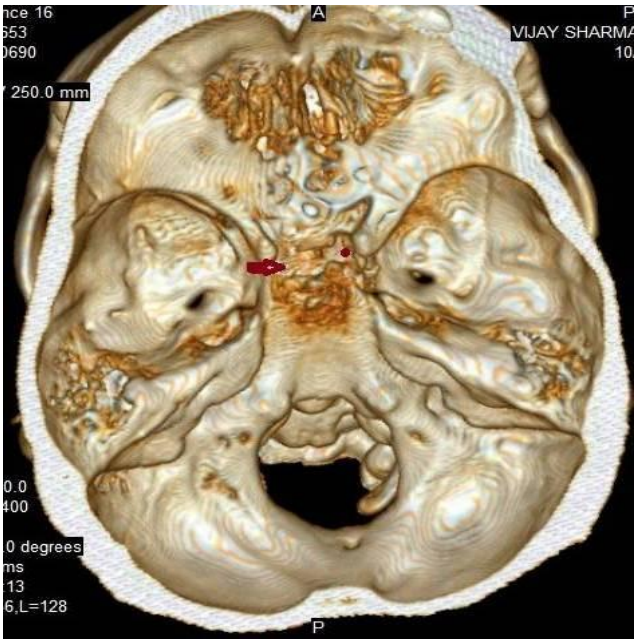

Figure 5 Multislice CT with 3D imaging reconstruction with volume rendering – internal view of the skull base showing left carotid canal agenesis (red arrow) & normal right carotid canal (red dot) .

Discussion

An extremely rare incidence of ICA agenesis has been reported, with few scarce tens of cases being worldwide reported, may be, to some extent, because the dysgenesis is generally asymptomatic in the greatest majority of patients. This occurs because there is a sufficient cerebral circulation supplied by anastomosis in the circle of Willis, intracavernous and external carotid artery anastomosis, besides persistent embryonal arteries. In these cases, the patients are referred for medical assistance because of complications resulting from abnormalities associated with carotid artery agenesis [7, 9].

The ICA agenesis is usually unilateral, although there are reports of asymptomatic bilateral agenesis [10]. If the agenesis is detected by MRI angiography, it must be confirmed by CT, in an attempt to find hypoplasia or absence of the carotid canal. Generally, the main secondary source of blood supply is the vertebrobasilar system (in cases of bilateral agenesis) or the dominant ICA (in cases of unilateral agenesis or hypoplasia) [9].

The main findings associated with these anomalies are transsphenoidal encephaloceles, circle of Willis aneurysms, and an extensive rete mirabilis in the cranial base. Intracranial aneurysms are found in about 25% of cases of symptomatic internal carotid artery agenesis with intracranial hemorrhagic manifestations [7,8, 11,12]. Also, other less frequent findings are reported. There are few cases where agenesis is associated with neuropsychomotor development delay, agenesis of the corpus callosum and persistent cavum vergae, in patients with bilateral agenesis. Few cases of unilateral agenesis in association with arachnoid cyst are reported [13,14]. Also, there is a report about association of megadolichobasilar anomaly and olivopontocerebellar atrophy with unilateral ICA agenesis [15], besides rare cases of hypopituitarism associated with unilateral artery agenesis, although intracavernous anastomosis seem to be efficient [16,17].

To confirm the patency or dysgenesis of the carotid canal, corresponding to the vessel agenesis or hypoplasia, multidetector CT with 3D reconstruction also is useful in cases of dubious diagnosis. In the literature no such case has been reported previously presenting with recurrent TIA along with agenesis of the internal carotid artery with atherosclerotic calcification in the intracranial segment of contralateral ICA & hypoplastic right vertebral artery along with accessory vertebral or collateral & occlusion or agenesis of right ECA.

Three types of collateral circulation have been described as occurring in patients with unilateral or bilateral agenesis of the ICA [1, 12]. The first is called the fetal type. With the fetal type, the anterior cerebral artery of the affected side is supplied by the normal contralateral ICA via the anterior communicating artery. The middle cerebral artery arises from the basilar artery through an enlarged posterior communicating artery. This type of collateral circulation is the most commonly encountered [1,12,18]. In the present case, the anterior cerebral artery on the left side was also mainly supplied by the right anterior cerebral artery via the anterior communicating artery and the middle cerebral artery was supplied by the basilar artery via the posterior communicating artery. The second type is called the adult type. With the adult type, both the anterior cerebral and middle cerebral arteries are supplied via the anterior communicating artery [1]. With the third type, which is the rarest, transcranial anostomosis may develop from the external carotid system, from the contralateral ICA, or from the primitive vessels [1,2,12,18].

In the present case, left ICA was absent & left MCA is supplied by basilar artery through posterior communicating artery (fetal type) (Figure3).

Conclusion

In this brief case report, there is no intention to affirm that agenesis of the ICA should be always suspected. However, it may be concluded that a detailed and methodological evaluation of the carotid canals and ICA flow void in the investigation of primary or acquired stenosis (a usual finding in patients with neurological complaints) may lead to the finding of this anomaly. The carotid canals should always be evaluated during the reading of routine cranial CT studies with axial slices and bone window images, considering that this is the confirmatory sign of the ICA absence, and does not imply an increase neither in the acquisition time nor in the cost of the study but it make difference in the management plan of patient . In this case cause of recurrent TIA was agenesis of left ICA with atherosclerotic changes in contralateral ICA & hypoplastic right vertebral artery.

References

1. Midkiff RB, Boykin MW, McFarland DR, Bauman JA. Agenesis of the internal carotid artery with intercavernous anastomosis. AJNR Am J Neuroradiol 1995; 16: 1356–1359.
2. Damry N, Hanquinet S, Christophe C, Janssen F, Delatte P, Perlmutter N.

Bilateral congenital absence of the internal carotid artery with a primitive transmaxillary arterial anastomosis. Pediatr Radiol 1994; 24: 200–203.
3. Turnbull I. Agenesis of the internal carotid artery. Neurology 1962; 12: 588–590.
4. Lie TA. Congenital Anomalies of the Carotid Arteries. Amsterdam: Excerpta Medica Foundation; 1968:30–51.
5. Verbiest H. Radiologic findings in a case with left the internal carotid artery agenesis & compression of several cranial nerve roots in the posterior fossa by basilar artery. Med Contemp 1954; 71;601-9.
6. Florio F, Balzano S, Nardella M, et al. Congenital absence of the internal carotid artery. Cardiovasc Intervent Radiol 1999;22:74–78.
7. Blustajn J, Netchine I, Frédy D, et al. Dysgenesis of the internal carotid artery associated with transsphenoidal encephalocele: a neural crestsyndrome? AJNR Am J Neuroradiol. 1999;20: 1154–7.
8. Quint DJ, Silbergleit R, Young WC. Absence of the carotid canals at skull base CT. Radiology. 1992;182:477–81.
9. Tasar M, Yetiser S, Tasar A, et al. Congenital absence or hypoplasia of the carotid artery: radioclinical issues. Am J Otolaryngol. 2004; 25: 339–49.
10. Rumboldt Z, Castillo M, Solander S. Bilateral congenital absence of the internal carotid artery. Eur Radiol. 2003; 13 Suppl 6:L130–2.
11. Gailloud P, Clatterbuck RE, Fasel JHD, et al. Segmental agenesis of the internal carotid artery distal to the posterior communicating artery leading to the definition of a new embryologic segment. AJNR Am J Neuroradiol.2004;25:1189–93
12. Cali RL, Berg R, Rama K. Bilateral internal carotid artery agenesis: a case study and review of the literature. Surgery. 1993; 113:227–33.
13. Kidooka M, Okada T, Handa J. Agenesis of the internal carotid artery – report of a case combinedwith arachnoid cyst in a child. No To Shinkei. 1992; 44:371–5.
14. Owada Y, Sakuta Y. Bilateral carotid artery agenesis with corpus callosum hypogenesis – a case report. No To Shinkei. 1995; 47:589–94.
15. Okabe S, Oda N, Ishii M. Agenesis of left internal carotid artery associated with megadolicho-basilar anomaly and olivopontocerebellar atrophy. Rinsho Shinkeigaku. 1989; 29:1395–400.
16. Moon WJ, Porto L, Lanfermann H, et al. Agenesis of internal carotid artery associated with con-genital anterior hypopituitarism. Neuroradiology.2002; 44:138–42.
17. Baysefer A, Akay KM, Tasar M, et al. Congenital absence of internal carotid artery associated with hypogonadotropic hypogonadism – a case report. Vasc Endovascular Surg. 2002; 36:457–60.
18. Smith RR, Kees CJ, Hogg ID. Agenesis of the internal carotid artery with an unusual primitive collateral: case report. J Neurosurg 1972; 37:460–462,
19. Lie TA. Congenital anomalies of the carotid arteries. Amsterdom: Ecerpta Medica; 1968. p. 44-9.
20. Neves WS, Kakudate MY, Cêntola CP, Garzon RG, Poça d'Água A, Sanches R. Ageness of internal carotid artery : a case report. Radiol Bras. 2008; 41(1):63–66.