Case 16795

Eurorad

Congenital Absence of the left Internal Carotid Artery.

Published on 22.06.2020

DOI: 10.35100/eurorad/case.16795
ISSN: 1563-4086
Section: Head & neck imaging
Area of Interest: Anatomy Arteries / Aorta
Neuroradiology brain
Procedure: Normal variants
Imaging Technique: MR
Imaging Technique: MR-Angiography
Special Focus: Congenital Case Type: Clinical Cases
Authors: Dr. Simon Gatt1, Dr. Adrian Mizzi2 , Dr. Francis Zarb3
Patient: 22 years, female

Clinical History:

22-year-old female complaining of almost daily migrainous headaches for a number of months. No associated neurological signs of note. Family history of subarachnoid haemorrhage secondary to aneurysm rupture.

Imaging Findings:

On MRA imaging, the left internal carotid artery is absent at the cervical, skull base and cavernous sinus levels (Figure 1). T2-axial imaging shows hypoplasia of the left carotid canal with a normally developed right carotid canal.

At the level of the circle of Willis, the left middle and anterior cerebral arteries are being supplied via a prominent and tortuous left posterior communicating artery as well as from the anterior communicating artery (Figure 2).

The brain parenchyma appears preserved with no evidence of any previous infarction being seen. Normal appearances of the rest of the posterior circulation of the brain.

Discussion:

Background: Absence of the internal carotid artery (ICA) is a rare congenital disorder that affects less than 0.01% of the population [1]. It is often used as an ‘umbrella’ term that encompasses a spectrum of varying degrees of ICA under-development, ranging from hypoplasia to agenesis [1,4,5]. Aplasia can be found at the midpoint of this spectrum, implying the existence of an early structure which eventually failed to form later during development.

Clinical Perspective: The majority of patients tend to have no symptoms related to this anatomical variant, in view of good collateral flow via the rest of the vessels of the Circle of Willis. However, a number of different symptoms have been reported, ranging from headaches [1] to hearing loss [2]. This condition is also reported to have associations with intracerebral arterial aneurysm formation [1-2] which could themselves cause the patient to be symptomatic.

Imaging Perspective: Imaging plays a key role in diagnosis. Sonographic assessment of the carotids might show a markedly stenotic ICA with/without complete obstruction of flow. CT is excellent for assessment of the bony passage the ICA normally takes throughout the base of skull and cavernous sinus. An absent carotid canal can help confirm congenital aetiology of this condition as opposed to an acquired one (such as post-traumatic ICA stenosis/occlusion) [5]. MR imaging (especially time-of-flight protocols) can again confirm the extent of ICA under-development, and demonstrate the collateral flow that would have consequently developed [1-3].

An important reason to obtain this diagnosis is when there is contralateral arterial disease that might need therapeutic intervention. Having knowledge of the existence of this anatomical variant is therefore essential as management plans will naturally move to favour the preservation of the solitary patent carotid supply (which might not have been considered had both ICAs been patent) [1-3].

Outcome and Teaching Points: Treatment options do not currently exist for this congenital condition and are often not required due to satisfactory collateral compensatory flow to the affected brain segments. However, part of the management plan should include the performance of angiographic imaging to exclude the presence of any concurrent aneurysm formation which, as discussed previously, tends to have a higher prevalence in these patients [3].

Written patient consent for this case was waived by the Editorial Board. Patient data may have been modified to ensure patient anonymity.

Differential Diagnosis List: Congenital Absence of the left Internal Carotid Artery.,

Final Diagnosis: Congenital Absence of the left Internal Carotid Artery.

References:

Given CA II., Huang-Hellinger F, Baker MD, et al. Congenital absence of the internal carotid artery: case reports and review of the collateral circulation. AJNR Am J Neuroradiol 2001;22:1953–9.
Costa NR, Reis AM, Duarte D, et al. Congenital absence of internal carotid artery: an unsuspected diagnosis Case Reports 2016;2016:bcr2016218289.
Li S, Hooda K, Gupta N, Kumar Y. Internal carotid artery agenesis: A case report and review of literature. Neuroradiol J. 2017 Apr;30(2):186-191. doi: 10.1177/1971400917692162. Epub 2017 Jan 1. PMID: 28424012; PMCID: PMC5433594.
Lie TA. Congenital Anomalies of the Carotid Arteries. Amsterdam: Excerpta Medica; 1968:35–51.
Teal JS, Naheedy MH, Hasso AN. Total agenesis of the internal carotid artery. AJNR Am J Neuroradiol 1980;1:435-442

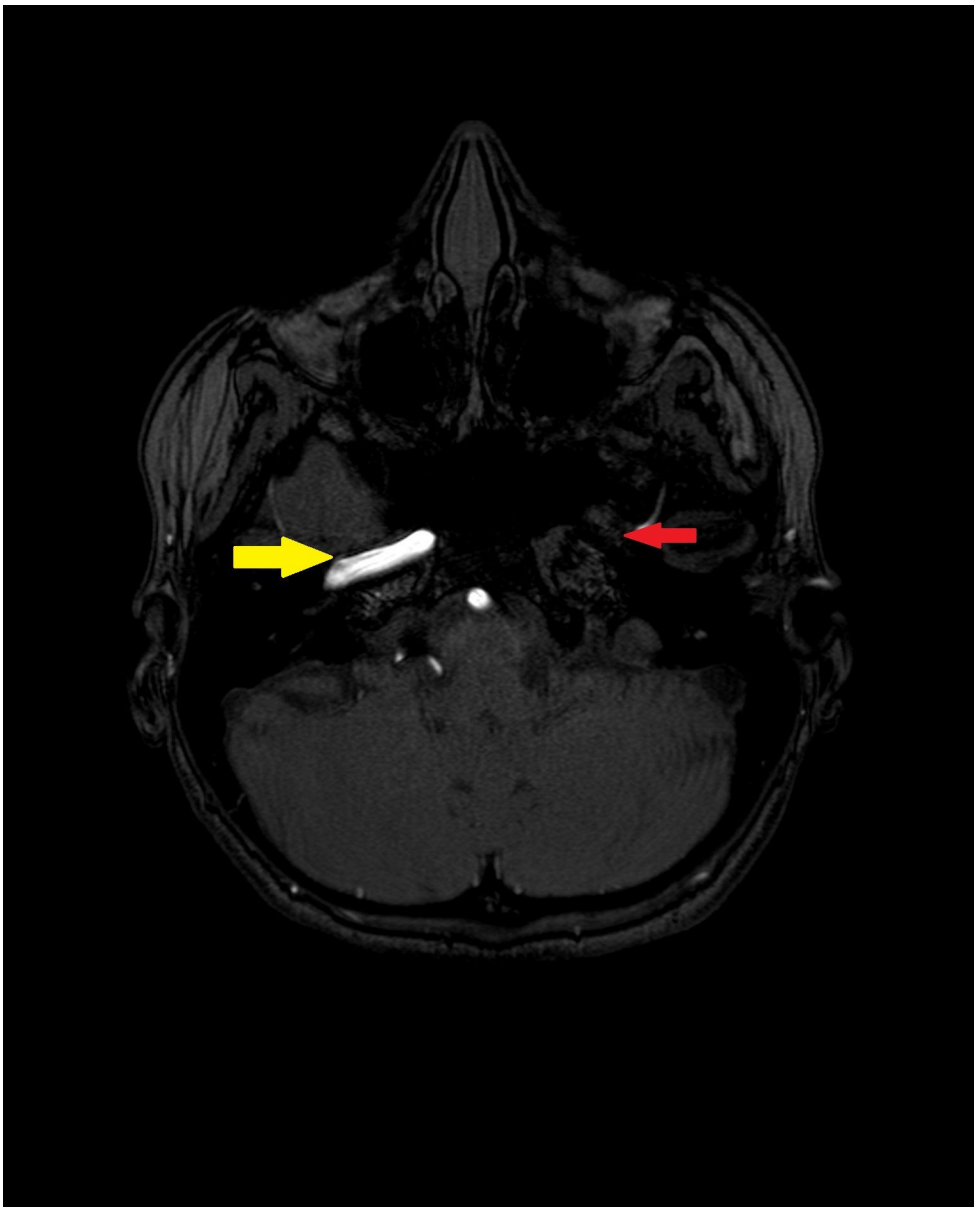
Figure 1

a

Description: Spot image from an axial time-of-flight MR angiogram showing a normal right ICA (yellow arrow) within the right carotid canal, and an absent contralateral ICA (red arrow) within a hypoplastic carotid canal. **Origin:** © Medical Imaging Department, Mater Dei Hospital, Malta, 2019

Figure 2

a



Description: Spot image from a reconstructed 3D MR angiogram (axial view at the level of the Circle of Willis) showing a normal right ICA (blue arrow) but an absent contralateral ICA (red arrow). A hypertrophied left posterior communicating artery (white arrow) is noted to partially supply the left middle cerebral artery. **Origin:** © Medical Imaging Department, Mater Dei Hospital, Malta, 2019