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Ischemic Stroke Revealing Internal Carotid Artery Hypoplasia: A Case Report

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Abstract **Case Report**

Congenital hypoplasia of the internal carotid artery (ICA) is a rare condition, often asymptomatic due to collateral circulation. We report a 22-year-old male with a history of ischemic stroke and chronic headaches, where brain MRI and CT scans revealed right ICA hypoplasia. Imaging, particularly MR angiography and CT, is crucial for diagnosing ICA hypoplasia, evaluating collateral pathways, and detecting potential complications, ensuring accurate assessment and management.

Keywords: ICA hypoplasia, Congenital vascular anomaly, MR angiography, Brain CT scan.

INTRODUCTION

Congenital anomalies of the internal carotid artery (ICA) include agenesis, aplasia, and vascular hypoplasia. Hypoplasia of the ICA results in a diffuse narrowing of the arterial lumen. It is a very rare anomaly of the embryonic development with an incidence of less than 0.01 % [1].

This congenital anomaly is usually asymptomatic due to effective collateralization from the circle of Willis. However, it is crucial to acknowledge its potential implications in ischemic strokes, epilepsy, headaches, and its potential association with intracranial aneurysms [2].

We report the case of a 22-year-old young man with a history of old ischemic stroke, in whom brain CT scan and MRI revealed hypoplasia of the internal carotid artery (ICA).

CASE REPORT

We present the case of a 22-year-old young male patient with a history of an ischemic stroke involving the deep right sylvian territory at the age of 17, and a traumatic brain injury at the age of 19, resulting in bilateral frontal contusion lesions. The patient was currently seeking evaluation for chronic headaches. Clinical examination revealed signs of pyramidal irritation.

To investigate the patient condition, a brain CT scan and MRI were performed.

The MRI revealed a right capsulo-lenticular porencephalic cavity, characterized by hypointensity on T1-weighted images and pronounced hyperintensity on T2-weighted images that diminished on FLAIR sequence. This cavity resulted in the displacement of the ipsilateral lateral ventricle and dilation of cortical sulci adjacent to it indicative of post ischemic sequelae. In addition, bilateral frontal gliosis lesions were observed, showing hyperintensity on FLAIR images. These lesions exhibited hemorrhagic stigmata on the gradient-echo T2 sequence, indicative of post-traumatic sequelae (Figure 1). On the MR angiography sequences, an extensive amputation of the right internal carotid artery signal (ICA) was observed (Figure 2). No anomalies were observed at the cerebral level in the circle of Willis. The brain CT scan passing through the base of the skull confirmed the presence of stenosis of the carotid foramen with a significant reduction in the diameter of the right carotid canal, measuring 3 mm in diameter compared to 10 mm on the left side (Figure 3). Thus, allowing the diagnosis of constitutional hypoplasia of the right carotid artery have been established.

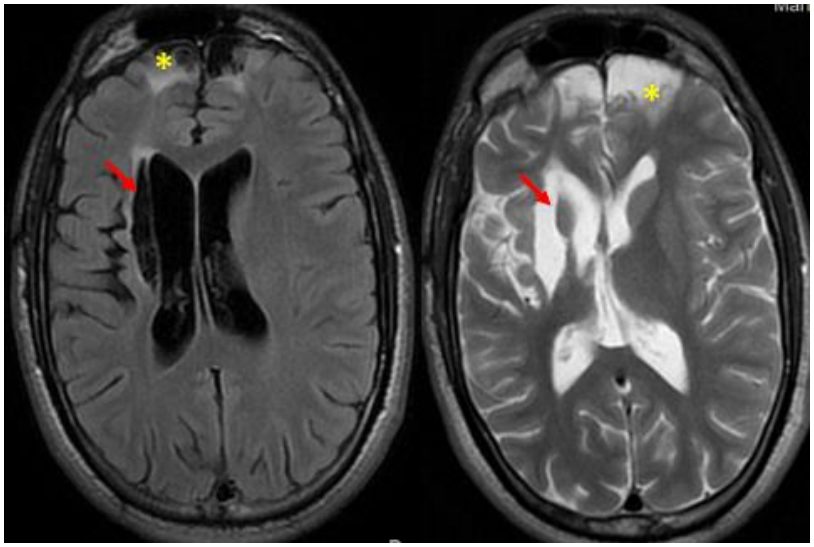

Figure 1: Brain MRI in axial T2 and Flair séquences revealing a right capsulo-lenticular porencephalic cavity, characterized by hypointensity on T1-weighted images and pronounced hyperintensity on T2-weighted images that diminished on FLAIR sequence (red arrows). This cavity resulted in the displacement of the ipsilateral lateral ventricle and dilation of cortical sulci adjacent to it indicative of post ischemic sequelae. In addition, bilateral frontal gliosis lesions were observed, showing hyperintensity on FLAIR images. These lesions exhibited hemorrhagic stigmata on the gradient-echo T2 sequence, indicative of post-traumatic sequelae (yellow asterix)

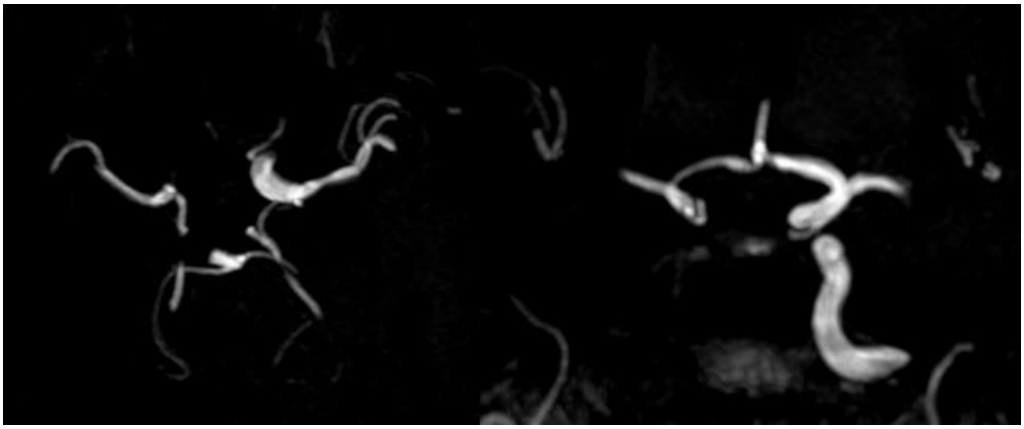

Figure 2: TOF MR angiography sequences, displays a significant loss of signal in the right internal carotid artery (ICA)

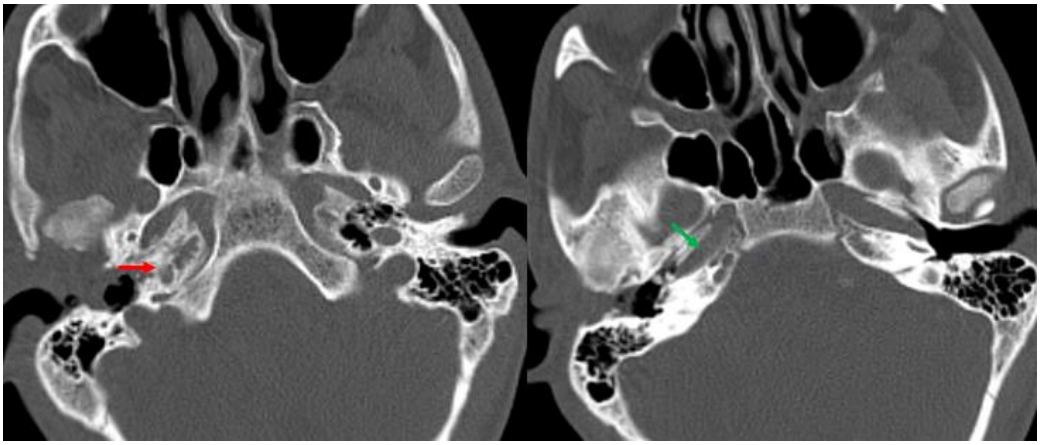

Figure 3: Brain CT scan passing through the base of the skull illustrates a stenosis of the right carotid foramen (indicated by a red arrow), along with a notable reduction in the diameter of the right carotid canal(green arrow), measuring 3 mm compared to 10 mm on the left side. These findings suggest constitutional hypoplasia of the right carotid artery

DISCUSSION

The hypoplasia of the internal carotid artery (ICA) described by Tode in 1787 is an extremely rare congenital malformation, observed in less than 0.01% of the population. This type of arterial malformation occurs before the 24th day of embryonic life, this leads to a subsequent lack of development of the bony carotid canal at the 6th week. This anomaly can be bilateral or unilateral. The circumstances of discovery vary widely and can include ischemic stroke, cerebro-meningeal hemorrhage, headaches, and seizures. However, it is often asymptomatic discovered incidentally through color Doppler ultrasound of the carotid arteries or cerebral MR angiography conducted for other reasons [3].

Vascular compensation varies depending on the type of dysgenesis of the internal carotid artery (ICA), whether it is unilateral or bilateral. Six types have been described in the literature:

- **Type A:** Unilateral ICA hypoplasia is compensated by hypertrophied ipsilateral anterior communicating and posterior communicating arteries.
- **Type B:** Compensation is provided by the anterior communicating artery for both the anterior cerebral artery and middle cerebral artery territories.
- **Type C:** In the case of bilateral ICA hypoplasia, compensation for the reduced blood supply from both ICAs is achieved through the two posterior communicating arteries.
- **Type D:** Unilateral ICA hypoplasia is compensated by anastomoses from the cavernous sinus that drain into the carotid siphon.
- **Type E:** Compensation for hypoplasia of both ICAs is provided by the two posterior communicating arteries for the territories of the middle cerebral arteries, while the anterior cerebral arteries continue to be supplied by the hypoplastic ICAs.
- **Type F:** Distal collateral circulation is ensured by the external carotid artery, internal maxillary artery, and skull base anastomoses [4].

The diagnosis of internal carotid artery (ICA) hypoplasia is made through imaging techniques, Doppler ultrasound. Digital angiography has long been the gold standard for diagnosing this anomaly. However, the development of non-invasive angiography now allows for diagnosis.

Normally, the internal carotid arteries have an average diameter of 4 mm with a variation of 0.8 mm. Therefore, congenital ICA hypoplasia is suspected when the ICA diameter is less than 3 mm, accompanied by reduced blood flow (less than 100 mL/min) without visualizing any morphological abnormalities in the arterial wall [4].

Magnetic resonance imaging (MRI) with angiographic sequences can assess the various collateral pathways and their courses. Three-dimensional reconstructions using CT angiography can confirm the small caliber of the hypoplastic internal carotid artery, the absence of visualization of its intracavernous segment, and collateral circulation. It is particularly useful in identifying any associated intracranial aneurysms [5].

CT scan of the the skull base demonstrates a reduction in the caliber of the carotid canal, allows for the differentiation between congenital carotid hypoplasia and acquired conditions that lead to a narrowing of the internal carotid artery (ICA) such as arterial dissection, fibromuscular dysplasia, atherosclerosis, or moya moya desease, each has its own therapeutic implications [5].

Up to this point, no therapeutic consensus has been established for managing dysgenesis of the internal carotid artery (ICA) as most cases are asymptomatic. However, due to the high risk of aneurysm formation, prevention of cerebral vascular insufficiency should be established [6].

CONCLUSION

Congenital hypoplasia of the internal carotid artery is a rare anomaly. Its recognition is important to avoid inappropriate management. The development of non-invasive imaging techniques enables the identification of this anomaly as well as the detection of potential complications. Doppler ultrasound coupled with MR angiography and/or CT angiography can suggest the diagnosis of congenital ICA hypoplasia. A skull base CT scan can confirm the diagnosis by demonstrating a small carotid canal.

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