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Case Report

Bilateral internal carotid artery hypoplasia presenting with watershed territory infarcts [☆]

Shikhil Uppal^a, Ankur Chandra^b, Bipin Chaurasia^{c,*}^a Department of Neurosurgery, Uppal Neuro Hospital, Amritsar, India^b Department of Radiology, EDIR, DICRI, Uppal Neuro Hospital, Amritsar, India^c Department of Neurosurgery, Neurosurgery Clinic, Birgunj, Nepal

ARTICLE INFO

Article history:

Received 9 May 2024

Revised 21 May 2024

Accepted 23 May 2024

Keywords:

Internal carotid artery

Hypoplasia

Infarcts

Imaging

Diagnosis

ABSTRACT

Internal carotid artery hypoplasia is a rare vascular anomaly that can lead to various neurological symptoms due to altered cerebral blood flow. We present a case of a 36 years old female who presented to us with forgetfulness and right sided weakness. She was ultimately diagnosed with bilateral internal carotid artery hypoplasia through imaging studies. This case highlights the importance of considering vascular anomalies in patients presenting with neurological symptoms and the significance of comprehensive diagnostic evaluation for appropriate management.

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Introduction

Internal carotid artery hypoplasia is a congenital vascular anomaly characterized by underdevelopment of one or both internal carotid arteries (ICAs) [1]. It is often asymptomatic but can manifest with various neurological symptoms such as transient ischemic attacks (TIAs), strokes, headaches, and visual disturbances. Here, we report a case of internal carotid artery hypoplasia presenting with forgetfulness and right sided weakness. We also discuss its diagnosis and subsequent management.

Case presentation

A 36-years-old female who was a known case of grade II hypertension and type 2 diabetes mellitus presented to our neurology clinic with complaints of forgetfulness and right sided weakness. The onset of patient's symptoms was sudden with mild slurring of speech since 2 days. It was not associated with heaviness of head, diplopia, blurring, or loss of vision. There was no prior history of cerebrovascular accidents (CVA) or seizure. Neurological examination revealed patient was conscious, coherent but there was forgetfulness to retain new in-

Abbreviations: CT, computed tomography; ICA, internal carotid artery; MRA, magnetic resonance angiography; MRI, magnetic resonance imaging; DSA, digital subtraction angiography; MMSE, Mini-mental state examination; CTA, Computed Tomography Angiography.

[☆] Competing Interests: None.

* Corresponding author.

E-mail address: trozexa@gmail.com (B. Chaurasia).

<https://doi.org/10.1016/j.radcr.2024.05.073>

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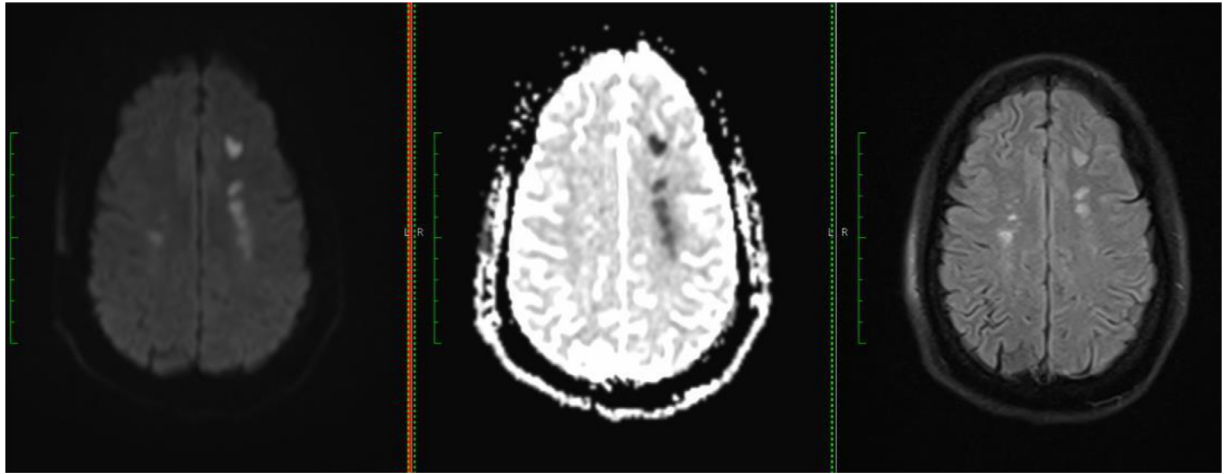


Fig. 1 – MRI DWI, ADC, FLAIR images showing acute watershed territory infarcts in left centrum semiovale and subacute infarct in right centrum semiovale.

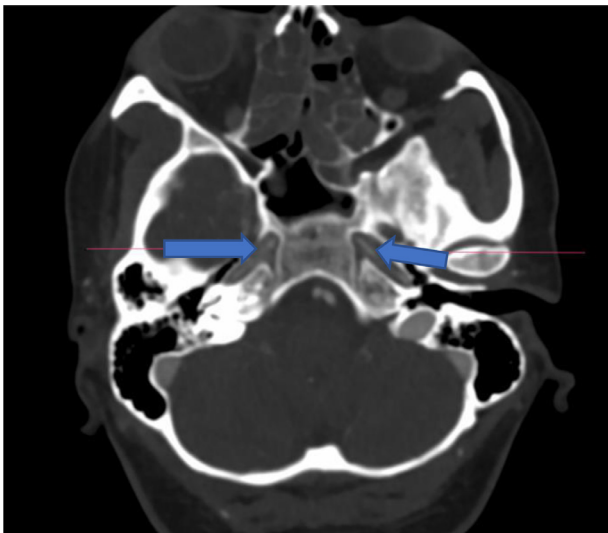


Fig. 2 – CT angiography: axial image showing reduced calibre of bilateral ICA.



Fig. 4 – VRT (Volume Rendering Technique) images showing bilateral hypoplasia of ICA.

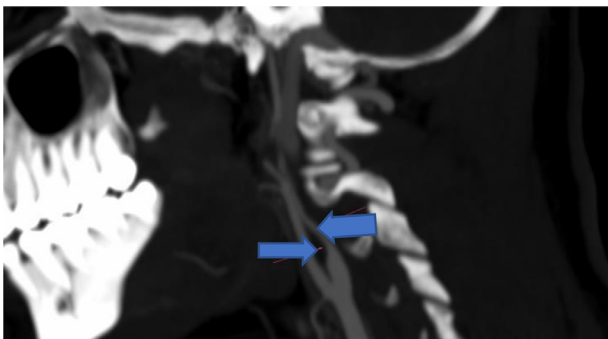


Fig. 3 – CT angiography Sagittal image showing narrow calibre of ICA.

formation. Her MMSE score was 26 with lost points in registration and recall. MRC (Medical Research Council scale) grade of power was 3/5 in right upper and 4/5 lower limbs. Further evaluation with MRI, CTA, and DSA demonstrated acute watershed territory infarcts in the regions of bilateral internal carotid artery.

Investigations: MRI was done which revealed acute watershed territory infarcts in left centrum semiovale and subacute infarcts in right centrum semiovale (Fig. 1). CTA revealed bilateral internal carotid artery hypoplasia (Figs. 2–4).

Additional investigations including digital subtraction angiography (DSA) confirmed the findings (Figs. 5 and 6). ACA and MCA on both sides was being filled by basilar artery via dilated Pcom (posterior communicating) artery.

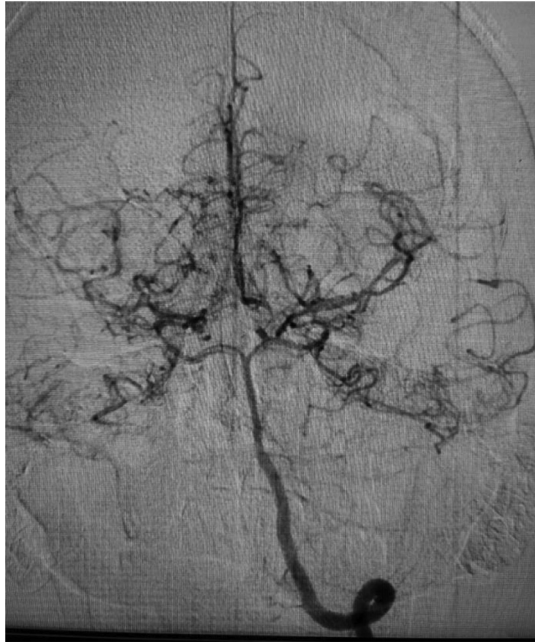


Fig. 5 – Digital subtraction angiograms show the ACA and MCA on both sides being filled by the basilar artery via dilated PCoAs.

Discussion

Bilateral hypoplasia of the internal carotid artery (ICA) is a rare vascular anomaly which is characterized by underdevelopment or reduced calibre of the internal carotid arteries on both sides [2–4]. It is often asymptomatic and incidentally discovered during imaging studies performed for unrelated conditions. However, in some cases, it can lead to cerebral ischemia, transient ischemic attacks (TIAs), or strokes due to compromised blood flow to the brain.

The aetiology of bilateral hypoplasia of the internal carotid artery is not entirely understood but it is believed to result from abnormal embryological development. The condition may occur in isolation or in association with other vascular anomalies or syndromes.

Hypoplasia of the ICA must be differentiated from acquired stenosis (especially when unilateral). Unilateral narrowing of the ICA is more likely to be due to chronic dissection, severe atherosclerosis, fibromuscular dysplasia or, sometimes, moyamoya disease.

The terms agenesis and hypoplasia are often used to describe the absence of ICA; however, there are some subtle differences among them. Agenesis refers to complete failure of the artery to develop while hypoplasia refers to incomplete embryonic development. Agenesis can be differentiated

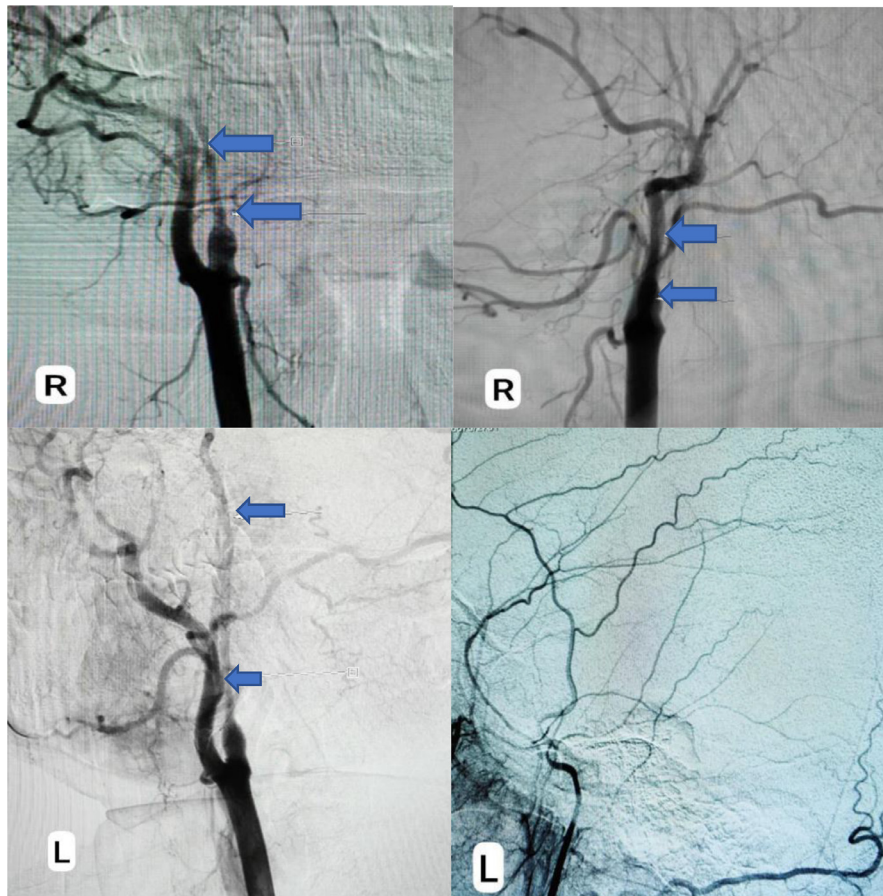


Fig. 6 – Frontal and lateral digital subtraction angiograms show bilateral ICA hypoplasia, with only the proximal ICA being normal (arrows).

from hypoplasia by establishing the complete absence of the carotid canal. This is because the carotid canal requires the presence of the ICA for its development. Therefore, agenesis and hypoplasia of the carotid artery can be distinguished by CT examination of the skull base. In case of congenital internal carotid artery hypoplasia (CICAH) small osseous carotid canal can be observed on skull base CT.

Treatment and Follow-up: The patient was managed conservatively with antiplatelet drugs, antihypertensive drugs and lifestyle modifications. Close follow-up was arranged to monitor for any progression of symptoms or development of complications.

Conclusion

Bilateral hypoplasia of the internal carotid artery is a rare vascular anomaly that can present with a range of symptoms or remain asymptomatic. Diagnosis is typically made through imaging studies, and management depends on the presence of symptoms and associated complications. Long-term monitoring is essential to prevent ischemic events and ensure optimal neurological outcomes.

Patient consent

Informed written consent was taken from patient prior to initiation of the project.

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