Case Report ISSN 2639-944X

Journal of Medical - Clinical Research & Reviews

Bilateral Hypoplasia of the Cervical Internal Carotid Arteries in a Pregnant Patient with Patent Foramen Ovale – A Case Report and Discussion

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Received: 22 Jan 2023; **Accepted:** 27 Feb 2023; **Published:** 03 Mar 2023

Citation: Nicol AT, Huang SQ, Patel S, et.al. Bilateral Hypoplasia of the Cervical Internal Carotid Arteries in a Pregnant Patient with Patent Foramen Ovale – A Case Report and Discussion. J Med - Clin Res & Rev. 2023; 7(3): 1-5.

ABSTRACT

Introduction: *Congenital hypoplasia or aplasia of the internal carotid artery (ICA) is believed to affect less than 0.01% of the population, with fewer than 200 unilateral and 30 bilateral cases reported in the literature. While often asymptomatic, they have been associated with stroke, transient ischemic attack (TIA), and other neurovascular pathologies.*

Case Report: *We describe a 33-year-old 28 weeks pregnant female with a history of patent foramen ovale (PFO) and ischemic stroke in the right middle cerebral artery (MCA) distribution three years prior who experienced new onset transient right upper extremity weakness and numbness. Duplex sonography revealed bilateral hypoplasia of the internal carotid arteries (HICA), confirmed by CT and MR angiography of the head and neck.*

Discussion: *To our knowledge, this is the first reported case of bilateral HICA with concurrent PFO and pregnancy presenting with TIA. All three conditions are associated with stroke and TIA. HICA should be included in the differential diagnosis of neurologic deficit despite its rarity.*

Keywords

Bilateral Hypoplasia of Internal Carotid Arteries (HICA), Pregnancy, Patent Foramen Ovale, Transient Ischemic Attack (TIA).

Introduction

Congenital agenesis, aplasia, and hypoplasia of the internal carotid artery (ICA) are rare, with less than 0.01% of the population believed to be affected [1-3]. Fewer than 200 cases have been described in the literature, of which there were less than 30 demonstrated bilateral abnormalities [3-5]. Hypoplasia of the ICA (HICA) is often asymptomatic due to collateral circulation, making its true incidence difficult to ascertain. Nonetheless, it has been associated with malformation and aneurysm of the Circle of Willis, as well as transient ischemic attack (TIA) and stroke [6-8]. It is possible that factors inducing vasculopathy and coagulation, such as smoking or pregnancy, as well as PFO, can further heighten the risk of adverse events in patients with HICA [9].

Case Report

A 33-year-old Hispanic female presented to the emergency department with one hour of right upper extremity weakness and numbness. These symptoms were preceded by constant right-sided headache and intermittent blurring of vision throughout the day. The patient was 28 weeks pregnant, with four previous pregnancies. She had a past medical history of obesity, hypertension, and PFO. Notably, the patient suffered a stroke three years prior, which presented with headache and right upper extremity weakness. She was not pregnant at the time of her previous stroke. She had no prior surgeries, and denied ever smoking or consuming alcohol. Due to her history, she had been started on enoxaparin for her current pregnancy. All symptoms other than the headache resolved

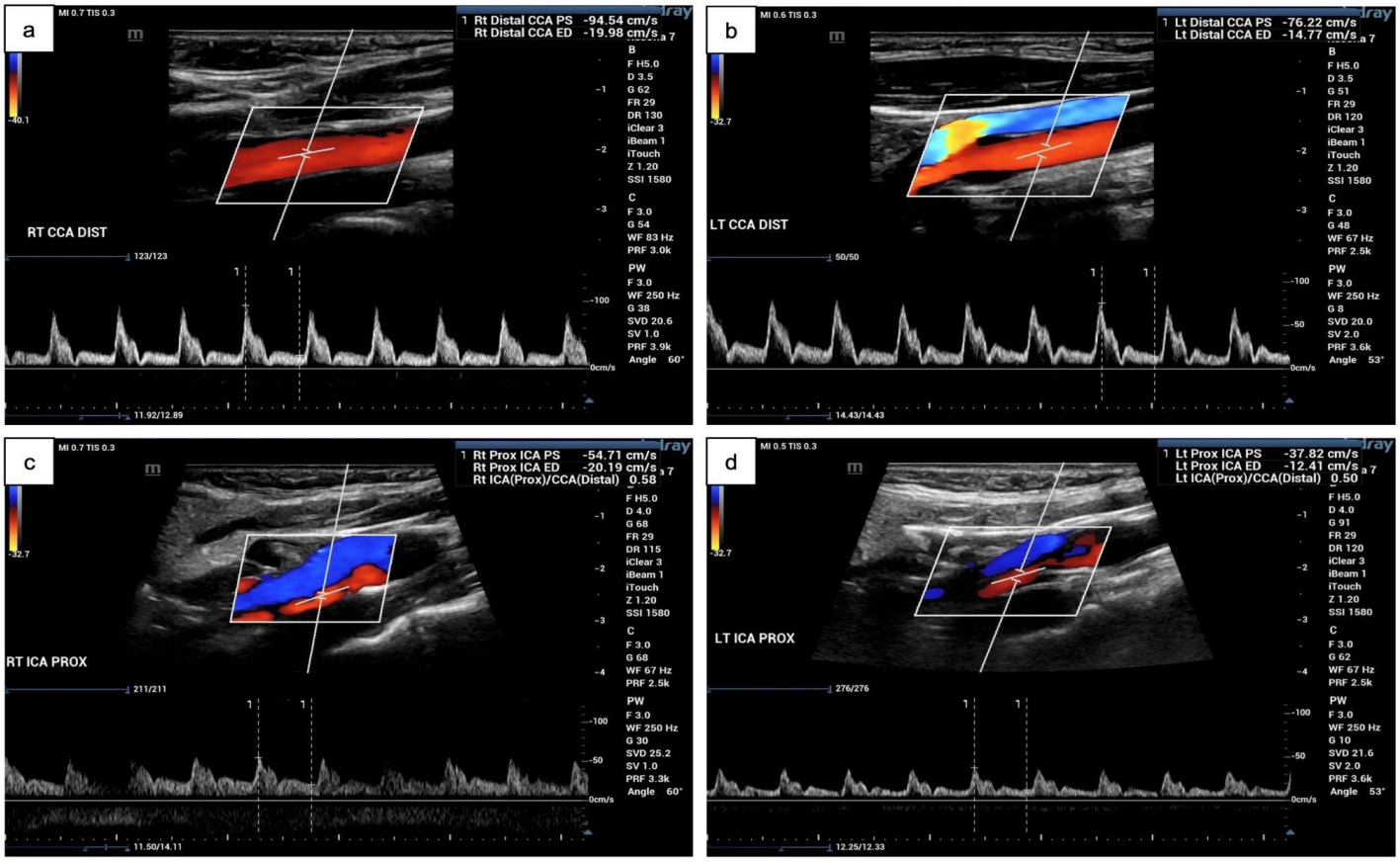

Figure 1: Extracranial duplex ultrasound, longitudinal field, demonstrating (a) normal right common carotid artery, (b) normal left common carotid artery, (c) attenuated flow with uniform small caliber in the right internal carotid artery, and (d) attenuated flow with uniform small calibern the left internal carotid artery.

by the time she was evaluated in the emergency department, but given her presentation and history, the patient was admitted for further workup.

Physical examination on admission revealed an initial blood pressure of 159/91 and an obese body habitus, with no other significant findings. There were no significant laboratory findings, including cardiac markers and coagulation studies. Electrocardiogram was unremarkable. Non-contrast head CT and MRI in the emergency department demonstrated no acute intracranial pathology while old infarcts were noted to the right basal ganglia and centrum semiovale. Doppler ultrasound of the extracranial carotids revealed diffuse smooth narrowing with low peak systolic velocities in both ICAs, with the right measuring 0.4 cm and the left 0.3 cm (Figure 1) in maximum diameter. No plaque formation was noted at the carotid bifurcations or ICAs.

Non-contrast CT showed narrowed bony carotid canals, with the external openings averaging 4 mm in diameter bilaterally and the petrous portions measuring 2.8 mm wide on the right and 3.1 mm wide on the left (Figure 2). CT and MR angiography of the neck confirmed smooth narrowing of the bilateral ICAs just distal to the carotid bifurcations, worse on the left, consistent with congenital hypoplasia (Figure 3). Similarly, CT and MR angiography of the head demonstrated reduced luminal size of the anterior and middle cerebral arteries (Figure 4). No aneurysms or other abnormalities were detected.

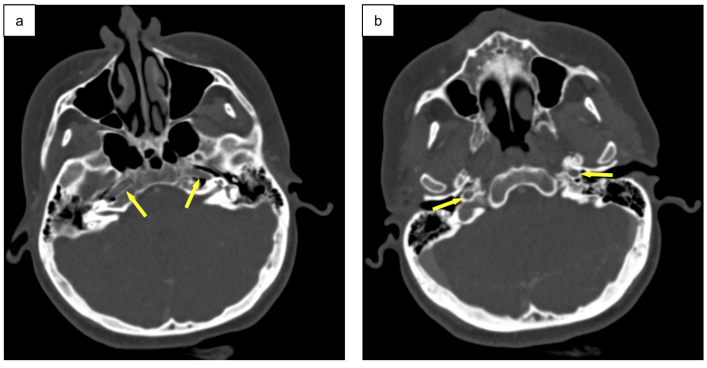

Figure 2: Axial non-contrast CT, using multiplanar reformation to correct for head tilt, demonstrating (a) bilateral narrowing of the petrous portions of the carotid canals (yellow arrows), with the right measuring 2.8 mm in width and the left measuring 3.1 mm; and (b) bilaterally reduced caliber of the external openings of the carotid canals (yellow arrows), both averaging 4.0 mm in diameter.

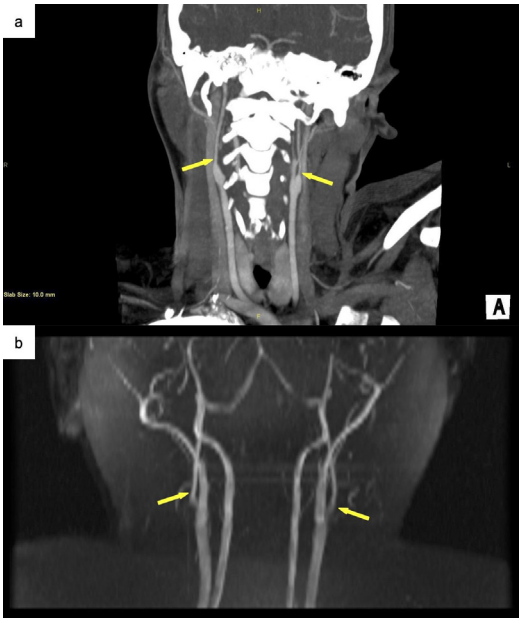

Figure 3: (a) CT angiography of the neck, 3D MIP coronal reconstruction, demonstrating markedly reduced caliber of the bilateral internal carotid arteries (yellow arrows) (b) MR angiography of the neck, 3D MIP coronal view, similarly showing reduced caliber of the internal carotid arteries (yellow arrows), with greater signal reduction noted to the left.

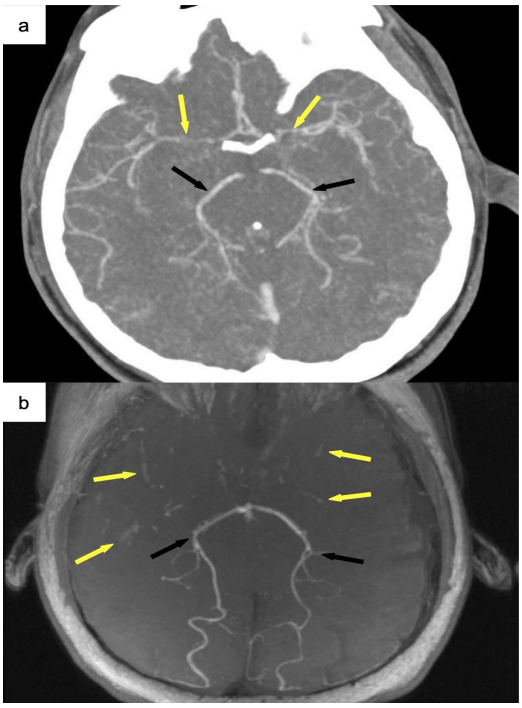

Figure 4: (a) CT angiography of the head, 3D MIP axial reconstruction, demonstrating attenuation of the bilateral middle cerebral arteries (yellow arrows) as compared to the posterior circulation (black arrows). (b) MR angiography of the head, 3D MIP axial reconstruction, demonstrating absence of flow-related signal intensities in the anterior circulation of the Circle of Willis. Nearly non-perceptible signal observed within the segments of the bilateral middle cerebral arteries (yellow arrows). Normal flow is present within the posterior circulation (black arrows).

Given the complete resolution of her symptoms and ongoing pregnancy, the patient was anticoagulated with enoxaparin and discharged home with a diagnosis of TIA with plans for close multidisciplinary follow-up.

Discussion

To the best of our knowledge, this is the second reported case of bilateral HICA associated with PFO, and the first in a currently pregnant patient [10]. While PFO by itself has not been shown to increase the risk of ischemic stroke, it is nonetheless found in up to 46% of cryptogenic stroke cases in patients younger than 55 years [11-13]. Miranda et al. describe three mechanisms by which PFO may cause brain ischemia: paradoxical embolism, in which emboli are shunted from the venous to arterial circulation via the PFO; thrombosis within the PFO leading to arterial embolism; and dysrhythmia of the left atrium due to a sufficiently large PFO, causing thrombus and embolus formation [13]. Although the relationship between PFO and stroke remains controversial, it is possible that HICA patients are more vulnerable to emboli secondary to PFO. Zhu et al. have reported a case of central retinal artery occlusion in a patient with bilateral HICA and PFO due to paradoxical embolism; narrowing of the ICA apparently led to dilation of the vertebrobasilar artery system, allowing the embolus to pass more easily to the affected artery [10]. Additionally, an embolus that might normally have traveled more distally and affected a smaller distribution in a patient with normal ICA patency may instead cause a devastating occlusion of the entire ICA distribution in patients with HICA. It is crucial to evaluate patients with HICA for potential sources of embolism, as such alterations to the regional vasculature may make them more susceptible to embolic insult.

Pregnancy and puerperium significantly increase the risk of TIA and stroke, both ischemic and hemorrhagic [14-16]. Pregnancy creates a state of relative venous stasis and hypercoagulability that persists months after delivery, making thrombosis and embolism more likely [15]. Over the past two decades, the incidence of stroke during pregnancy and puerperium has increased with the increasing prevalence of other established cardiovascular risk factors, including hypertension, diabetes, and obesity [14-16]. Given that so few cases have been reported during pregnancy or puerperium, precisely how these factors interact with HICA and affect the chance of brain ischemia has yet to be explored. Gupta et al. described a patient with bilateral HICA who developed frontal infarct and a small subarachnoid hemorrhage shortly after delivery [9]. These findings are consistent with compromised blood flow to the areas of the anterior and middle cerebral arteries. Similarly, our patient demonstrated reduced flow to the anterior circulation on CT and MR angiography. HICA patients experiencing pregnancy and puerperium may be particularly susceptible to strokes in these regions, necessitating careful evaluation of risk factors and treatment options. It is important to note that our patient first had a stroke at the age of 30 when not pregnant, suggesting that any propensity she may have for developing cerebral ischemia exists independent of gravidity. Given the aforementioned close

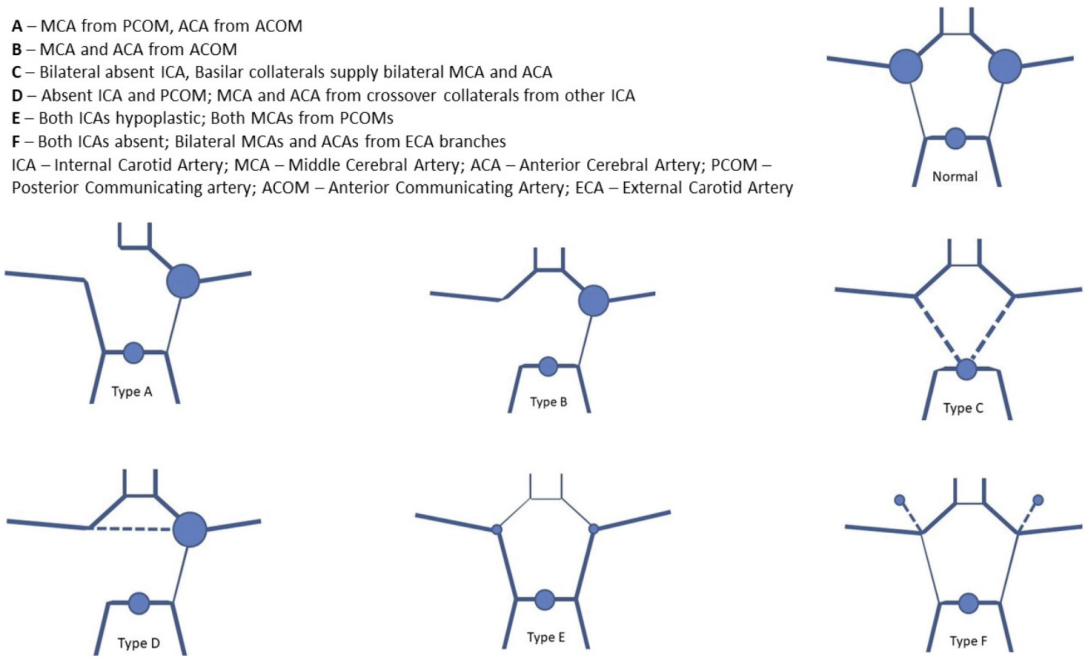

Figure 5: Schematic illustration of six patterns of collateral circulation in HICA proposed by Lie, (courtesy Dr. Steven Lev, MD).

relationship between PFO and ischemic stroke, the potential for bilateral HICA to increase her risk must be considered.

The embryologic origins of HICA remain controversial. Development of the aorta and its branches from mesenchyme begins in the 3rd week of development, with the proximal ICA being derived from the third aortic arch and the distal ICA from the dorsal aorta [17,18]. HICA may represent incomplete development of the dorsal aorta specifically, given how consistently the ICA maintains a normal width for up to 2 centimeters distal to the carotid bifurcation; this can be appreciated in our patient [7]. Multiple mechanisms have been proposed for unilateral HICA, most of which involve insult to the embryo between 4 - 8 weeks of development. These include exaggerated folding of the embryo to one side, constriction of the embryo by amniotic bands, and secondary regression of ICA precursors [3,6,17-18]. There has so far been no explanation for the development of bilateral HICA specifically [3,17].

A narrow or absent carotid canal on imaging indicates congenital ICA abnormality, as the development of the carotid canal depends on the proper formation of the ICA. It can therefore be used to distinguish between congenital HICA and acquired causes of ICA narrowing, such as atherosclerosis, chronic dissection, fibromuscular dysplasia, and vasculitides [3,17,19]. Few studies have examined the normal dimensions of the bony carotid canal. One study reported that the external openings of the carotid canal measured 5.41 mm anteroposteriorly and 7.52 mm mediolaterally on average [20]. Other studies reported mean diameters of the carotid canals ranging from 5.07 +/- 0.69 mm to 5.62 +/- 0.61 mm [21-23]. Our patient had external openings averaging 4.0 mm in diameter and petrous portions of the carotid canals averaging 3.0 mm in width without findings indicative of the other aforementioned conditions, increasing the strength of our diagnosis of bilateral HICA.

Another important consideration in the differential diagnosis of HICA would be moyamoya disease, which is associated with both ICA stenosis and narrowed carotid canals [22,23]. It typically presents with ischemic strokes in children and cerebral hemorrhage in adults [22,24]. Unlike HICA, moyamoya disease is understood to affect other vessels, including the anterior, middle, and posterior cerebral arteries. Additionally, ICA stenosis in moyamoya disease primarily occurs in the distal end of the vessel [23]. Moyamoya disease is best characterized by a small net of collateral vessels on direct cerebral angiography, said to resemble a puff of smoke in appearance [24]. The presence of diffuse stenosis restricted to the ICAs and the absence of "puff of smoke" appearance in CT and MR angiography, together with her presenting with cerebral ischemia as opposed to hemorrhage, makes moyamoya unlikely in our patient.

Lie classified six patterns of collateral circulation that arise with congenital ICA abnormalities, as illustrated in Figure 5 [10,25,26]. Our patient with bilateral HICA would most likely be classified as Type E; however, imaging did not demonstrate the prominent posterior communicating arteries (PCOM) that would be expected. There are likely patterns of collateral circulation beyond what Lie has described [4].

Conclusion

Our case demonstrates that HICA can be associated with stroke and TIA in young patients, especially when further complicated by vascular and hematologic factors. It is crucial that HICA be considered in the differential diagnosis in such cases despite its rarity, as variant anatomy has potential therapeutic implications.

Doppler ultrasound offers a simple and cost effective first step for evaluating a patient for HICA, while CT and MR angiography can confirm the diagnosis and characterize vascular abnormalities beyond the classically defined categories.

Consent Statement

Informed consent was obtained from the patient for the publication of this case report, including accompanying images.

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