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Review Article

Congenital Hypoplasia of the Internal Carotid Artery (HICA): A Short Review

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ABSTRACT

Introduction: Hypoplasia of the Internal Carotid Artery is a rare developmental abnormality. Because of its rarity, the association, diagnosis and treatment are not uniformly described.

Discussion: We discuss the rarity, etiology, clinical presentation, associated conditions, diagnosis and treatment. We review the literature to provide a guide on the approach and management of this silent yet possibly ominous disease.

Conclusion: Physicians need to be aware of this rare anomaly and all the various associated conditions, and should use a multi-disciplinary approach with careful long-term follow-up.

Keywords: Congenital hypoplasia of the internal carotid artery, Collateral circulation, Diagnosis & management

INTRODUCTION

Developmental abnormalities of the Internal Carotid Artery (ICA) are uncommon. The incidence is less than 0.01% of the population [1–3]. However, because of its silent nature, the true incidence remains unknown.

Congenital anomaly of the ICA can fall under the category of agenesis, aplasia, or hypoplasia [4,5]. Agenesis is the complete absence of the ICA, whereas aplasia is the lack of development of the ICA despite its precursor primordial structure existing at one point during development, and finally hypoplasia is the incomplete development of the ICA [1,6–8]. Since 1836, there have only been little more than 100 described cases in the literature [5,7–10]. Unilateral abnormalities are much more common than their bilateral counterpart [1,7]. An understanding of the pathogenesis associated conditions and potential complication is necessary for vascular surgeons and neuro radiologists so that this rare condition could be recognized and managed appropriately when encountered.

LITERATURE REVIEW

Pathogenesis

There have been competing views as to how such an anomaly can occur embryologically. Aortic arches form from mesenchyme between days 26 and 30. The third aortic arches become the common carotid arteries and proximal segments of the internal carotid arteries. The distal segments of the internal carotid arteries are derived from the dorsal aorta between the first and third arches [11].

It has been postulated that unilateral HICA is a result of some mechanical or hemodynamic stresses on the developing embryo [8]. In addition, Bhat et al hypothesized it could be due to an exaggerated folding of the embryo at 4-8 weeks towards one side or due to constriction from amniotic band(s) [1]. Nardone proposed that the abnormality is secondary to spontaneous regression of the ICA precursors [5]. Although there are possible explanations for unilateral HICA, the origin of the bilateral version remains unknown [8]. Some authors suggest that the form of collateral circulation depends on whether the disruption occurs prior to the development of the Circle of Willis (CoW), or after. If the disruption is prior to the development of CoW, Intercavernous collaterals develop [12].

Skull base development occurs at 5-6 weeks of fetal life [12]. If the ICA has not developed by 5-6 weeks, carotid canal atresia, absence of the sympathetic plexus, and other abnormalities will occur. The ophthalmic artery will have an aberrant origin [13]. Carotid canal under development may not always be present in unilateral hypoplasia (Figure 1).

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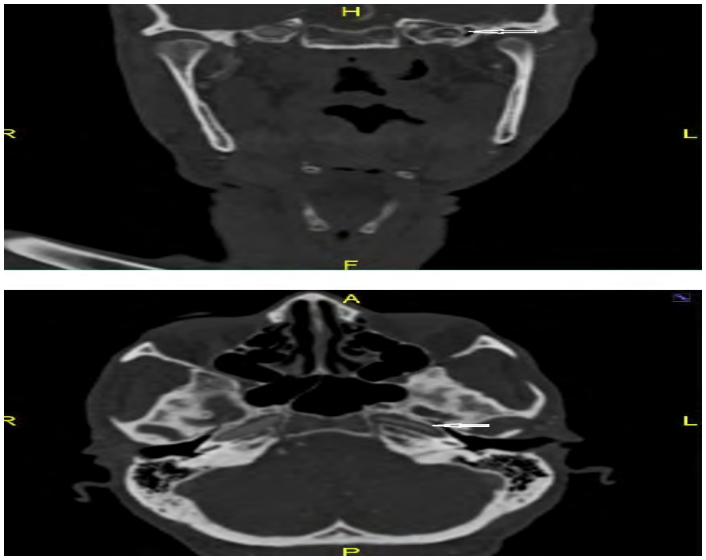

Figure 1: Coronal and axial CT images of the skull base demonstrate that the carotid canals have the same width bilaterally (arrows). The petrous portion of the left ICA, however, has diminished calibre relative to the right.

Clinical presentation

Due to the narrowing of the ICA from the incomplete development, collateral circulation will develop to compensate for the decreased perfusion to the brain [14]; as a result, HICA is most commonly detected incidentally [5,10,15]. However, HICA can have an eclectic range of presentation and symptomatology in a minority of cases. The presentation is largely dependent on the associated conditions that occur with HICA. The presentation may depend on the age group, with younger patients presenting with developmental delays (due to pituitary gland hypoplasia secondary to hypoperfusion) and symptoms of compression or bleeding from dilated or aneurysmal collateral arteries, and the older patients presenting with atherosclerotic cerebrovascular disease [13]. The patients can present with headaches, transient ischemic attacks (TIA)/strokes, seizures, intracranial aneurysms, subarachnoid hemorrhage, or parenchymal hemorrhage [1,7]. HICA can present with Horner's syndrome [9] or even with hearing or visual loss [3].

Associated disease

Intracranial anomalies are commonly associated with HICA. The two most common associations are intracranial aneurysms and aberrations of the CoW [4,13]. The general population has a 2-4% risk of developing intracranial aneurysms, while those with HICA have a 25-34% risk. Kaya et al have estimated that the risk can be as high as 67%. The aneurysm commonly develops on the basilar arteries and/or the posterior cerebral arteries (PCA) due to the increased hemodynamic stress from the increased flow through the collateral circulation [1,3,7,14]. The aneurysm formation is not limited to those two arteries and can occur in other intracranial vasculature as described in a report of an aneurysm developing in the anterior cerebral artery (ACA) in a 4-month-old infant [16]. Due to the altered flow dynamics the aneurysms in these patients have an increased tendency to bleed [17].

In addition to aneurysm formation, variations in the CoW are also commonly seen. More often than not, the aberrations in the CoW are the main sources of the collateral circulations in the brain [3,8].

For example, the hypo perfused areas of the brain secondary to unilateral HICA can be supplied by the communicating arteries as described by Akfirat et al in which the middle cerebral artery (MCA) ipsilateral to the HICA is supplied by the basilar artery and the posterior communicating artery [6]. In addition, the anterior communicating artery can supply the ACA [9] or the ACA can have collaterals from the external carotid artery (ECA) branches via the rete mirabiles [17]. Figure 2 shows a case with cross filling from the anterior communicating artery.

Furthermore, Tanaka et al also described a case of anomalous arteries from the posterior communicating artery and ICA supplying the ipsilateral MCA [18]. In the setting of bilateral HICA, the main vascular supply to the brain and CoW is via the vertebro basilar system [15].

Uncommon forms of collaterals that are unrelated to the CoW can also be present, such as transcranial anastomosis from the contralateral ECA or ICA [7,9], persistent embryological vessels [8] and even inter cavernous anastomosis [15]. Furthermore, the vascular anomalies can even extend extracranially; for example, Kaya et al report a case in which the right vertebral artery was the first branch to arise the aortic arch in the setting of HICA [3]. Lie had classified the collateral pathways into six categories [19]. We present here our schematic representation of Lie's classification (Figure 3).

Although rare, other miscellaneous anomalies have been described. For example, Horner's Syndrome has been documented to be associated with HICA in various age groups – Momtchilova et al describe a 5-month-old male infant presenting with Horner's syndrome with enophthalmos and iris hypopigmentation on the ipsilateral side as the HICA with no history of any birth trauma [20]. Another published case is that of a 15 year-old female who presented with Horner's syndrome with intermittent mydriasis since birth with an incidental finding of HICA ipsilaterally [9]; there is a reported case of a 67 year-old male with HICA that was associated with Horner's syndrome with heterochromia iridis [21]. Hasegawa et al described a 22 year-old female with HICA had multiple associated anomalies such as intraventricular hemorrhage from a ruptured collateral vessel and an ipsilateral encephalocele [22]. Afifi's infant was found to have cerebral hemihypoplasia in the setting of HICA [16].

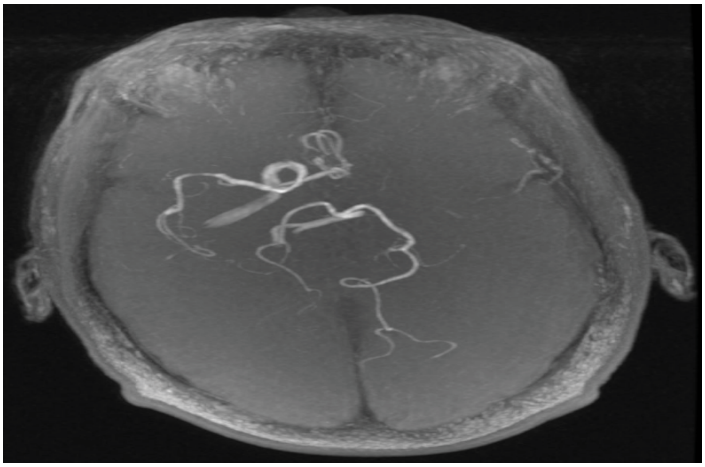

Figure 2: CTA of the Circle of Willis (CoW) shows lack of visualization of the left ICA and left MCA. There is cross-filling of the left ACA via the anterior communicating artery.

A – MCA from PCOM, ACA from ACOM
B – MCA and ACA from ACOM
C – Bilateral absent ICA, Basilar collaterals supply bilateral MCA and ACA
D – Absent ICA and PCOM; MCA and ACA from crossover collaterals from other ICA
E – Both ICAs hypoplastic; Both MCAs from PCOMs
F – Both ICAs absent; Bilateral MCAs and ACAs from ECA branches
ICA – Internal Carotid Artery; MCA – Middle Cerebral Artery; ACA – Anterior Cerebral Artery; PCOM – Posterior Communicating artery; ACOM – Anterior Communicating Artery; ECA – External Carotid Artery

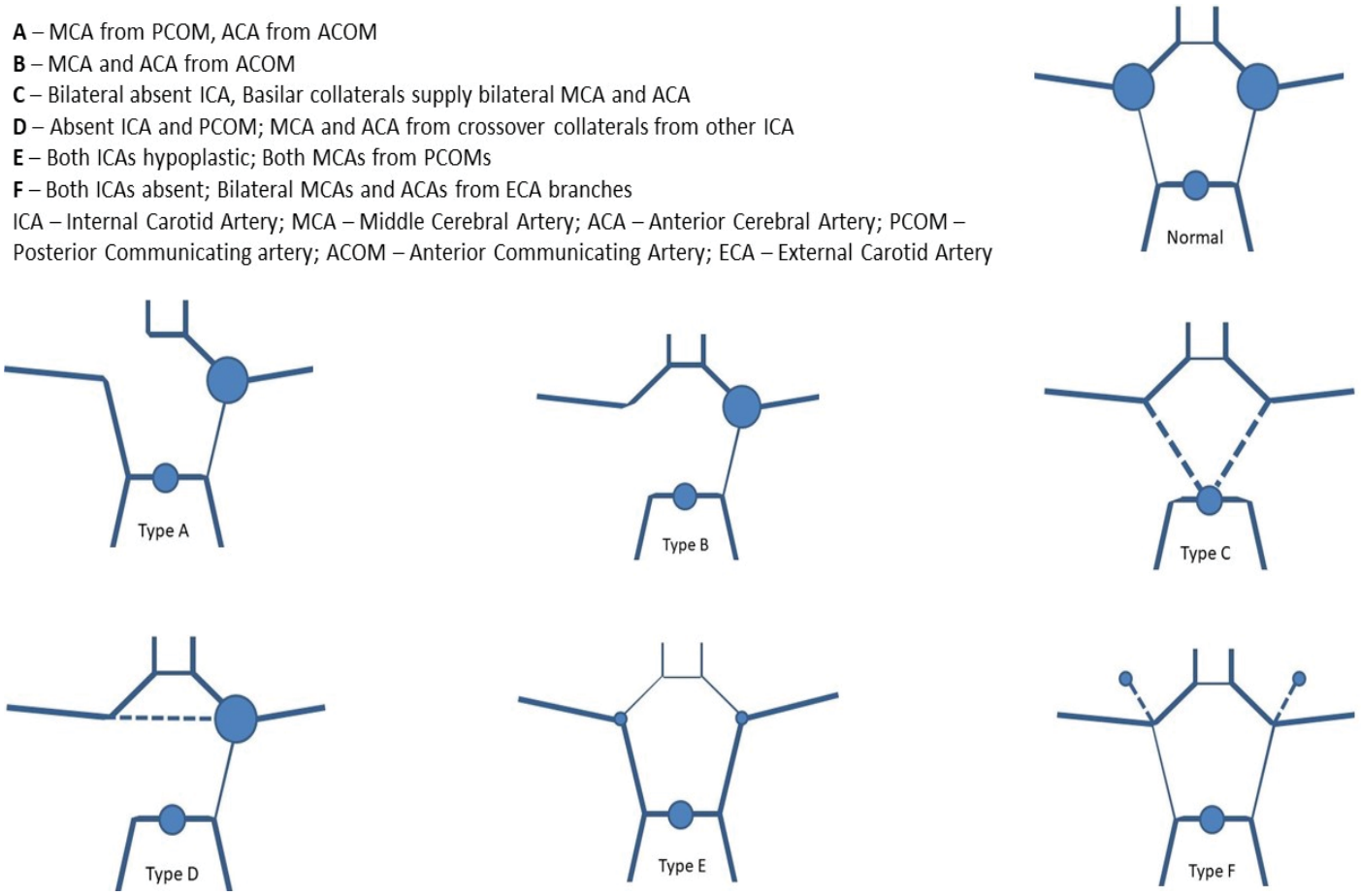

Figure 3: Schematic representation of Lie's classification scheme.

Diagnosis

Diagnosis is largely based on various imaging modalities; however, the most difficult challenge is differentiating HICA from acquired disease of the ICA. The differential diagnosis should include severe atherosclerosis, carotid dissection, fibromuscular dysplasia, arteritis, radiation-induced angiopathy, etc. [1]. Ultrasound can show diffuse luminal narrowing of the ICA, however that finding is non-specific [10]. The use of ultrasound Color Doppler (Figure 4) can suggest HICA if there is an absence of wall thickening (which is associated with acquired disease) and an absence of flow disturbances [23]. CTA and/or MRA will show diffuse narrowing of the ICA (Figures 5 and 6).

Depending on the age of presentation, there may be atherosclerotic disease at the carotid bifurcation. Embryologically, the ICA is an absolute necessity to the proper formation of the carotid canal, thus an underdeveloped ICA in the setting of HICA can present with a smaller-than-normal carotid canal [1] which can be detected by MRA or CTA [10].

Digital Subtraction Angiography (DSA) is not only invasive, but also vulnerable to confusing HICA with acquired disease causing diffuse luminal narrowing [23]; In addition, since bilateral HICA causes the CoW to receive its blood supply only from the vertebrobasilar system [15], DSA is inadequate due to the superimpositions secondary to that aberrant blood supply [7]. Unlike the other imaging modalities, CTA is advantageous in that it is less invasive, less expensive, and can better define the associated anomalies such as aneurysms [7]. Therefore, it is important that 3-Dimensional CTA be used to confirm the diagnosis.

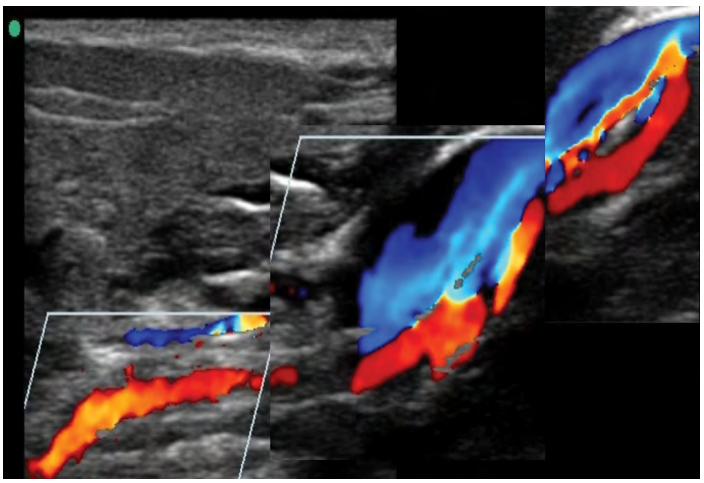

Figure 4: Color Duplex Scan demonstrates plaque formation at the left carotid bifurcation (arrow) and a smoothly narrowed ICA (arrowhead) throughout its cervical course (collage of images). The diameter of the ICA was 2 mm.

Treatment

Treatment approach has not been standardized because of the rarity of the condition. However, in a few reported cases, treatment has been focused on medical prevention, i.e., control of TIA and stroke risk factors, with promising results. A patient who presented with TIA symptoms in the setting of HICA, and was medically treated with antiplatelet therapy, statins, and blood pressure lowering medications had been free of symptoms for 7 months [4]. In addition, two patients who were treated with antiplatelet therapy - Aspirin 325 mg daily – for symptomatic HICA did not report

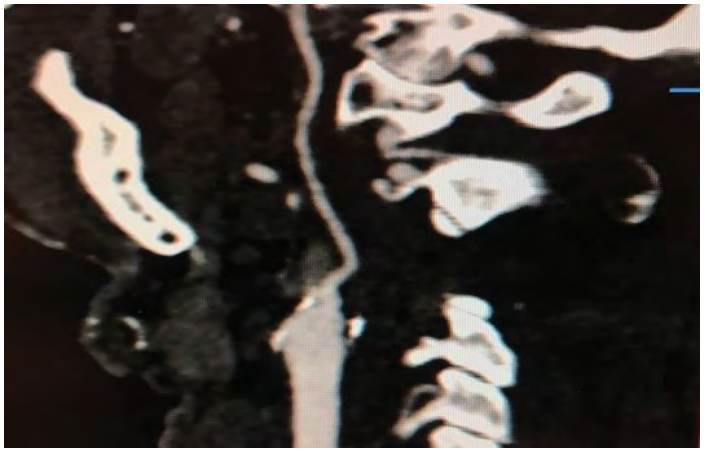

Figure 5: Maximum intensity projection (MIP) image from neck CTA demonstrates smooth tapering of the left internal carotid artery (ICA) from its origin (arrow). The left ICA has a small uniform calibre throughout its entire length.

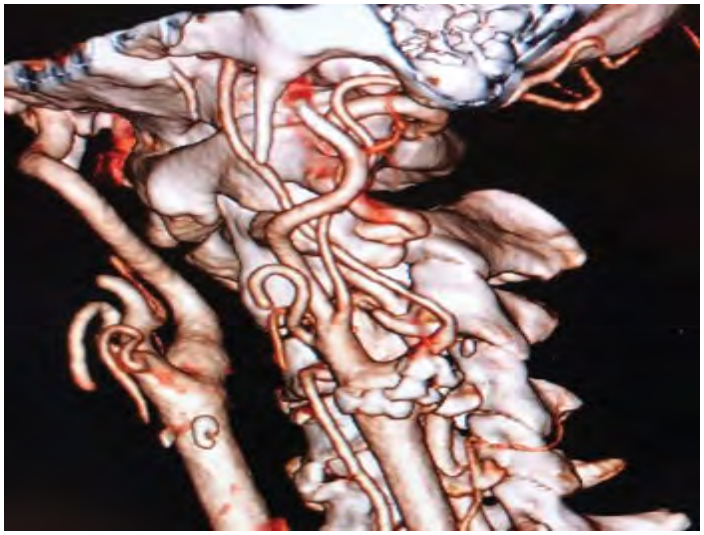

Figure 6: 3D reconstruction from neck CTA shows a constant diminished caliber of the entire cervical carotid artery (arrow). Calcific plaque is present at the carotid bifurcation (arrowhead).

symptoms at 13 months and 28 months respectively, along with no change in both their neuro radiological re-evaluations [5].

Furthermore, because of the intimate relationship between intracranial aneurysms and HICA, it has been suggested that treatment should also focus on evaluation and intervention of those aneurysms [7]. The use of endovascular stents and surgical clipping remain to be valid options even in the setting of HICA with its associated vascular variations [17] and also in the patients with bilateral agenesis of the ICA [24]. Finally, because of the associated aberrant intracranial vasculature, it is important to recognize each patient's unique anatomy since it will have great implications in the setting of surgical planning such as transsphenoidal hypophyseal surgery and carotid endarterectomy [10]. The hypoplastic ICA could be mistaken for a string sign and may be diagnosed only on direct surgical exploration, if not recognized preoperatively [25]. Recognition of HICA is important when surgical clamping of the contralateral ICA is planned. Awareness of this condition and the collateral pathways will also help in understanding that atheromatous emboli from the ICA can affect contralateral hemisphere. It should also be recognized that the collateral circulation that may be adequate under normal conditions could become inadequate at times of increased metabolic need such as development of cerebral metastases.

CONCLUSION

HICA, although rare and generally asymptomatic, can be troublesome to detect, manage, and monitor. Long term consequences of this congenital disease remain unknown especially where medical intervention is involved. Therefore, physicians need to be vigilant for all the various associations, and must utilize a multi-disciplinary approach with careful follow-up.

DISCLOSURE

There is no conflict of interests.

CONSENT

Images used are that of a deceased patient.

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