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Case Report

Internal left cartoid agenesis: case report and literature review

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ABSTRACT

Internal carotid agenesis (ICA) is an exceptionally rare condition. It is typically discovered incidentally, often during evaluations for other conditions, and generally asymptomatic due to compensatory collateral circulation. In this case A 57-year-old female with chronic background of anxiety disorder and auricular tachycardia; congenital malformations: imperforate anus with multiple surgical interventions until the age of 15 years old; right ovary agenesis and right kidney hypoplasia.

Keywords: Internal carotid agenesis, Neurosurgery

INTRODUCTION

Intern carotid agenesis (ICA) is extremely rare; might occur in 0.01% of the population, being more frequently appear in the left side in (3:1) relation.[1] It's usually found as an incidental diagnosis, such as in a fetal life form anastomosis and collateral flood it's usually made, which create enough irrigation to the affected area.[2] In most cases it doesn't show any symptoms but might be associated to further complications. In this article, we will review the existing literature and report of a clinical case of a 57-year-old female patient with agenesis of the left internal carotid artery.

CASE REPORT

A 57-year-old female with chronic background of anxiety disorder and auricular tachycardia; congenital malformations: imperforate anus with multiple surgical interventions until the age of 15 years old; right ovary agenesis and right kidney hypoplasia. A year before the current evaluation, she experienced two episodes of syncope with loss of urinary sphincter control, no specific aphasia, and each lasting no longer than five minutes. Additionally, she had tachycardia and vertiginous syndrome with dyspnea. The thorax shows normal dynamics with no signs of respiratory distress. Lung fields exhibit good breath sounds with no additional noises. Heart sounds are rhythmic, of good intensity and tone, with no murmurs or abnormal sounds. Limb strength is decreased in the thoracic limbs, rated ++++/+++++.

As part of the study protocol, a cardiological evaluation was performed using a Holter monitor, which showed monofocal atrial tachycardia. A carotid Doppler examination conducted on 08/30/2023 reported a left common carotid artery caliber of 4.4 mm, compared to 6.7 mm on the contralateral side. This also indicated a slight decrease in flow rate relative to the contralateral artery. An accurate bifurcation was not observed, and there was inappropriate flow in the proximal segment of the internal aorta, with a velocity of 37 cm/s and reduced diastolic flow."

Angio tomography of the supra-aortic branches was requested for anatomical delineation and to rule out any underlying causes. On 12/22/2023, the report identified an anatomical variant of the supra-aortic branches, classified as type 2 according to Natsis. Additionally, it observed hypoplasia of the left carotid artery at the base of the cranium. The posterior right communicating artery, the internal left carotid artery, and the posterior cerebral artery were not visible (Figures 1A and B). This concludes that the origin of the syncope and collapses is associated with low cardiac output related to the previously described anatomical variants of the cerebral circulation, despite the patient continuing with her normal daily activities.

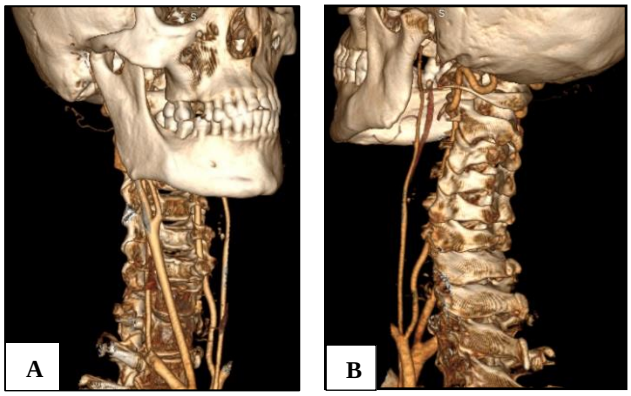

Figure 1: Three-dimensional reconstruction of computed tomography in the arterial phase at the cervical level. (A) Right oblique lateral view comparing the bifurcation of the common carotid artery. (B) Left oblique posterior view comparing the external carotid artery from its origin.

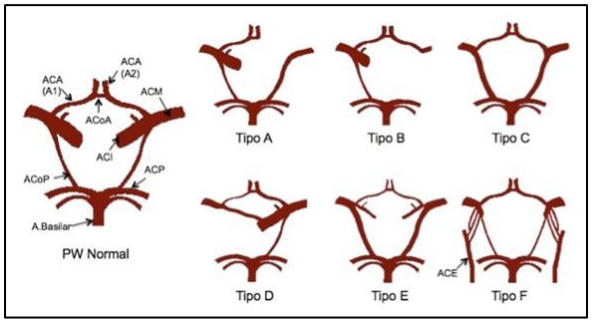

Figure 2: Classification of carotid dysgenesis patterns (adapted from lie).

DISCUSSION

Cerebral artery variants are relatively common and can include a range of anomalies. Among the most frequently described variants are anomalous origins of arteries, agenesis, sectoral hypoplasia, duplications, segmental fenestrations, and the absence of vessels, as well as variations in irrigation territories. One of the rarest variants is agenesis of the internal carotid artery.[3]

Agenesis of the internal carotid artery is usually unilateral, with the contralateral internal carotid artery compensating for the absence of the main blood supply.[4] There are 6 original collateral pathways: Type A-There is a unilaterally absent internal carotid artery with collateral circulation through the anterior cerebral artery (Acom A) via the anterior communicating artery (ACom) and the ipsilateral middle cerebral artery (MCA) via the posterior communicating artery (PCOM), which is generally hypertrophic. Type B-There is a unilaterally absent internal carotid artery combined with collateral circulation through the ACA and MCA via the ACom. Type C-The agenesis is bilateral, and the anterior circulation is supplied through the vertebrobasilar system via a hypertrophic PCOM. Type D-Characterized by unilateral agenesis of the cervical portion of the internal carotid artery and of the intercavernous collaterals of the intercavenous segment of the contralateral internal carotid artery. Type E-There is bilateral hypoplasia of the internal carotid arteries, and their ACAs and MCAs are supplied by hypertrophic PCOMs. Type F-There is bilateral absence, compensated by transcranial anastomoses and branches of the carotid artery.[5] (Figure 2). Clinical characteristics can be categorized according to age groups (0-20 years, 20-40 years, over 40 years). In the 0-20 years group, developmental delay (54%) and symptoms resembling subarachnoid hemorrhage (25%), such as headache, nausea, and vomiting, were the most common.

Developmental delay is often due to abnormal development of the pituitary gland or problems with its blood supply. Subarachnoid hemorrhage symptoms are attributed to the formation and rupture of aneurysms caused by altered intracranial hemodynamics. In the over-40 age group, nearly half of the patients experienced transient ischemic attacks and ischemic strokes, possibly due to decompensation of collateral circulation caused by atherosclerosis. In the 20-40 years group, although any of the mentioned symptoms may occur, most asymptomatic cases (12.5%) were observed in this group, possibly because the brain adapts to intracranial collateral pathways.[6]

The anomaly can be detected in a doppler ultrasound, where the carotid bifurcation is not visible, or in which hypoplasia of the ipsilateral common carotid artery is observed, as in the case described here. It is possible to differentiate between agenesis and carotid occlusion. In agenesis, the carotid canal will be absent, and when it has a malformation or absent, it prevents the development of the internal carotid artery (ICA).

The presence or absence of a carotid canal on CT imaging is key to distinguish between aplasia and agenesis. The absence of a carotid canal in the head on CT, along with the absence of the ICA on angiography, will confirm the diagnosis of ICA agenesis.[6] Conventional MRI may show that the flow void phenomenon is absent. Under normal conditions, blood

with high flow through the internal carotid artery cannot return the signal in time, and the flow does not appear in the scan. Digital subtraction angiography can show that the internal carotid artery disappears immediately after its origin, while the diameter of the external carotid artery may be larger than normal.[7]

Treatment

Currently, there is no effective treatment for congenital absence of the internal carotid artery (ICA). The main therapeutic approaches focus on maintaining compensatory blood flow and reducing blood flow resistance, which is achieved through symptomatic treatment and reconstruction of collateral circulation pathways. This may include carotid stenting or carotid endarterectomy in cases of extracranial ICA stenosis. Dolichoectasia of the collateral pathways and large aneurysms can cause neurological symptoms due to compression of normal brain structures. Treating aneurysms may relieve pressure on these normal brain structures.[8]

CONCLUSION

Congenital absence of the internal carotid artery (ICA) is an extremely rare condition. It is essential to consider vascular malformations when evaluating neurological symptoms. Although this condition generally presents with few symptoms, it is strongly associated with cerebral aneurysms. Early clinical signs of this malformation may include nonspecific symptoms such as headaches, syncope, transient ischemic attacks, or cerebral hemorrhage. Therefore, the possibility of carotid agenesis should be considered when more common causes have been ruled out. Early diagnosis is crucial for optimizing clinical management and planning potential future surgical interventions.

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