

## Bilateral Internal Carotid Artery Hypoplasia in Asymptomatic Patient. Case Report

*Hipoplasia Bilateral de Carótida Interna em Paciente Assintomática. Relato de Caso*

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### Introduction

Hypoplasia of the internal carotid artery is a congenital anomaly that involves narrowing of the arterial caliber 1 to 2 cm above the carotid bifurcation.<sup>1</sup> Internal carotid agenesis and hypoplasia are rare, occurring in less than 0.01% of the population.<sup>2</sup> Sixty cases of hypoplasia of the internal carotid have been reported, of which only 24 were bilateral hypoplasia.<sup>3</sup> We report the case of bilateral hypoplasia of the internal carotid artery in an asymptomatic patient.

### Clinical case

A 54-year-old woman sought medical emergency with occipital headache, right hemiface paresthesia, dysarthria and hypertensive peak of 200x120 mmHg. Captopril 50 mg and anxiolytic were administered and the patient progressed with BP improvement to 140x80 mmHg and total resolution of the clinical condition, being discharged 12 hours later. At admission, except for dysarthria, her neurological condition was normal. History of hypertension and bipolar disorder under irregular use of losartan 100 mg, quetiapine 50 mg, topiramate 50 mg, clonazepam 2 mg/day.

Family history of SAH (father, mother and two sisters); coronary artery disease (father with sudden death at 59); diabetes mellitus (mother, 1 sister, maternal grandmother, 2 maternal aunts).

She was referred for outpatient investigation, when laboratory tests revealed hypercholesterolemia. Electrocardiogram, chest X-ray, echocardiogram, cranial tomography and fundus with no abnormal findings. Carotid artery Doppler showed 1.2 mm thickening of the right bulb and possibly hypoplasia of the right and left internal carotid arteries. Cervical vessel computed tomography angiogram confirmed suspicion of bilateral hypoplasia of the internal carotid artery, revealing: type I aortic arch with minimal

calcifications; 8.6mm diameter brachiocephalic trunk, right common carotid artery 4.8 mm, right external carotid artery 2.9 mm, left common carotid artery 5.4 mm, left external carotid artery 3.5mm, all pervious, with regular walls, no lesions; absence of intrapetrous carotid path, reduced right and left carotid canal caliber and right and left internal carotid artery with hypoplasia throughout its path (Figure 1). Enlarged right vertebral artery caliber (V1 segment of 7.4 mm), left vertebral artery 6.5 mm, both pervious and with no wall lesion. Right-sided basilar artery formation crossing the midline and originating the posterior cerebral arteries to the left of the midline, complete Willis polygon. Pervious right and left anterior and middle cerebral arteries, with no lesions, filling completely through the posterior communicating system. Pervious right and left posterior cerebral arteries, with no lesions, filling directly through the vertebrobasilar system (Figure 2).

### Discussion

First, it is important to know the difference between agenesis, hypoplasia and aplasia of the carotid artery, since they represent different clinical conditions. Agenesis is when the development of the internal carotid artery (ICA) does not occur; hypoplasia when there is formation but development is incomplete obtaining a smaller diameter caliber and aplasia is used when there are traces of ICA. These abnormalities are congenital and extremely rare. In the literature so far, some 60 cases of ICA hypoplasia have been reported, of which 24 cases were bilateral,<sup>3</sup> such as the present case.

The Willis Polygon consists of two vascular systems — the vertebra-basilar and carotid system — is responsible for brain irrigation and is located at the base of the skull. This system is usually formed from anastomosed vertebral arteries originating the basilar artery, housed in the basilar gutter. It divides into two posterior cerebral arteries that irrigate the posterior inferior surface of each of the cerebral hemispheres. On each side, the internal carotid arteries originate a middle cerebral artery and an anterior cerebral artery. The anterior cerebral arteries communicate through a branch between them which is the anterior communicating artery. The posterior cerebral arteries communicate with the internal carotid arteries through the posterior communicating arteries.

Although the absence of one or both internal carotid arteries is rare, it is not usually symptomatic due to the formation of collateral circulation, such as persistent embryonic vessels, normal anastomotic pathways through the Willis Polygons

### Keywords

Carotid Artery, Internal; Hypoplasia; Congenital Abnormality; Computed Tomography Angiography.

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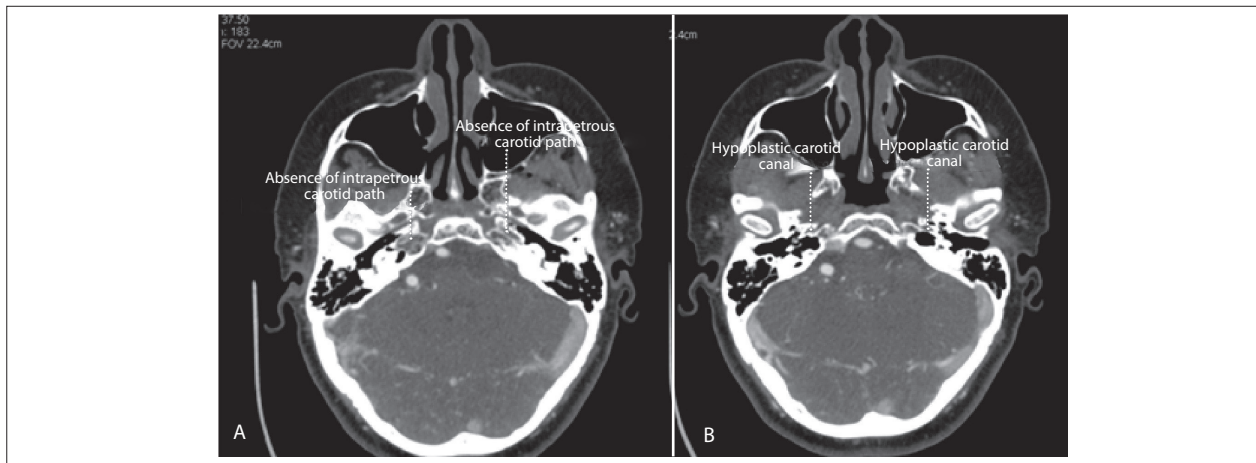
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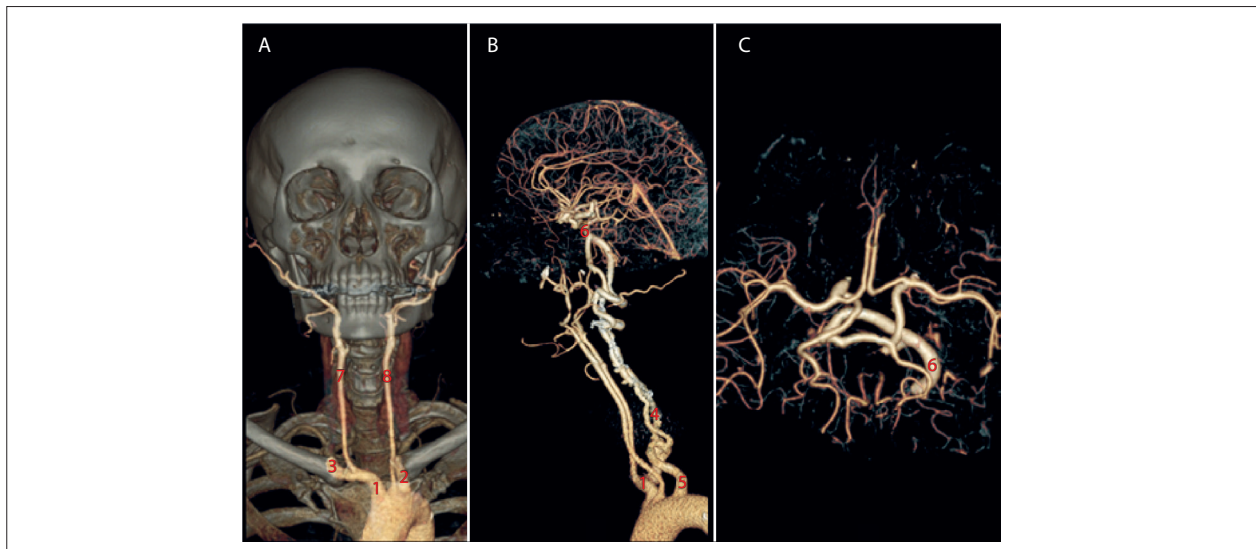
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## Case Report



**Figure 1** – Absence of intrapetrous carotid path (A) and hypoplastic right and left carotid canal (B).



**Figure 2** – Three-dimensional image of cerebral arterial circulation through the vertebral arteries (A and B) and complete Willis polygon (C) on computed tomography angiography. 1 = brachiocephalic artery; 2 = left subclavian artery; 3 = right subclavian artery; 4 = right vertebral artery; 5 = left vertebral artery; 6 = basilar artery; 7 = right carotid artery; 8 = left carotid artery.

or anastomosis with the external carotid arteries. Hypoplasia occurs about 1–2 centimeters above the bifurcation and may continue throughout its intracranial course, ending in the ophthalmic artery in some cases. It is assumed that the cause of ICA hypoplasia is incomplete development of the fetal dorsal aorta, which usually originates the distal cervical segment of the ICA up to the clinoid segment. A frequent abnormality is the widening of the posterior communicating basilar artery, in an attempt to create collateral circulation as a means of compensation. In cases with bilateral abnormalities, brain supply usually occurs from the vertebrobasilar system, while in unilateral ones, it usually occurs from the dominant contralateral carotid artery.<sup>1–6</sup>

In this case, our patient had right and left carotid canal with reduced caliber and hypoplasia of the right and left

internal carotid arteries throughout its course. Right and left vertebral arteries of increased caliber with basilar artery formation. Complete Willis polygon with right and left anterior, middle and posterior cerebral arteries originating from the vertebrobasilar system with no wall lesions.

The prevalence of this condition is not yet known. Tasar et al., reviewing 5,100 magnetic resonance angiography scans and/or brain angiography scans found 7 patients with congenital absence or hypoplasia of the ICA (0.13%), most of which with incidental diagnosis. Of those, only 1 case was bilateral ICA hypoplasia (0.01%).<sup>5</sup>

These anomalies are usually asymptomatic due to the presence of collateral circulation. However, some symptoms or abnormalities may appear in adulthood, such as subarachnoid hemorrhage (because of its association with

saccular aneurysm), transient ischemia and cerebral infarction. The main radiological findings are: smaller caliber of the internal carotid artery, forming the chordal sign seen by the hyperdensity of the artery, hypoplasia of the carotid canal in the temporal bone and vertebral-basilar system-dependent collateral circulation. These findings may also appear in other diseases, such as in carotid stenosis.

Our patient, in turn, was incidentally diagnosed at the age of 54.

## Conclusion

This case discusses this rare and usually asymptomatic congenital disorder, whose diagnosis is incidental. The suspicion evidenced in the carotid Doppler scan requires

computed tomography angiography or magnetic resonance angiography of cervical vessels and brain, which clearly enable the non-invasive identification of bilateral internal carotid hypoplasia and the whole intracranial circulation.

## Authors' contributions

Research creation and design: Caldas MA, Grilo LB, Soares LP, Custódio PS, Duarte LMV, Soares CEC. Data acquisition: Caldas MA, Grilo LB, Soares LP, Custódio PS, Duarte LMV, Soares CEC. Data analysis and interpretation: Caldas MA, Grilo LB, Soares LP, Custódio PS, Duarte LMV, Soares CEC. Manuscript writing: Caldas MA, Grilo LB, Soares LP, Custódio PS, Duarte LMV. Critical revision of the manuscript for important intellectual content: Caldas MA, Soares CEC.

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