



Unilateral Hypoplasia of the Internal Carotid Artery

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Authors' contributions

This work was carried out in collaboration between all authors. Author LP wrote the draft of the manuscript. Author UT designed the figures, managed literature searches and contributed to the correction of the draft. Author BA managed the literature searches. Authors GY, DA and FG provided the cases. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Hypoplasia of the internal carotid artery (ICA) is a rare congenital anomaly. Most of the patients are asymptomatic. When clinically symptomatic it presents as cerebral ischemia or hemorrhage. Carotid artery hypoplasia should not be confused with other conditions which may have a similar appearance such as arterial dissection, atherosclerosis, vasculitis and tubular fibromuscular hyperplasia. Hypoplasia of the ICA is usually discovered incidentally by computed tomography or magnetic resonance imaging. We report three cases of ICA hypoplasia which were discovered incidentally on computed tomography angiography (CTA). One of the three patients described an ipsilateral amaurosis fugax associated with the hypoplasia of the right internal carotid artery.

Keywords: Aneurysm; hypoplasia; internal carotid artery; multidetector computed tomography.

1. INTRODUCTION

Agenesis and hypoplasia of the internal carotid artery are very rare congenital anomalies,

occurring in less than 0.01% of the population [1,2]. They are more often unilateral. The exact cause of the agenesis and hypoplasia has not

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been published but a sequel from an insult to the developing embryo is accepted as a theory [2]. Hypoplasia of the ICA is usually asymptomatic, because of the collateral circulation from the contralateral ICA or vertebrobasilar system via the circle of Willis. Less commonly, collateral flow is provided via embryonic vessels, intercavernous anastomotic vessels or communicating arteries from external carotid artery [3,4]. When symptomatic, subarachnoid hemorrhage, transient ischemic attack or cerebrovascular insufficiency may be seen. Hypoplasia of the ICA has to be recognised because of its frequent association with aneurysms and not to be confused with more serious carotid diseases [4,5]. We present three cases of unilateral hypoplasia of the ICA assessed with CT angiography.

2. PRESENTATION OF CASES

2.1 Case 1

A 56-year old female patient presented with headache. Her past medical history was

unremarkable with no comorbidities and no medical treatment taken. It was the first time she admitted to hospital and she wasn't checked with CT or MRI before. She had no vascular risk factors and no family history of atherosclerotic or cerebrovascular diseases. On admission, general physical and neurological examinations were normal. Blood tests, including items relevant to diabetes, dyslipidemia and vasculitis, were unremarkable. She was referred to the radiology department. Cranial magnetic resonance imaging showed ischemic and gliotic lesions in the periventricular white matters and semioval center. Computed tomography angiography revealed hypoplasia of the right CCA and ICA with a transvers diameter of 3.6 mm and 0.7 mm respectively (Figs. 1a and b). The A1 segment of the right anterior cerebral artery was hypoplastic. The caliber of the right posterior communicating artery was larger than the left posterior communicating artery (Fig. 1c). Left ICA, middle and anterior cerebral arteries, anterior communicating artery was normal. On bone window settings the right carotid canal was also hypoplastic (Fig. 1d).

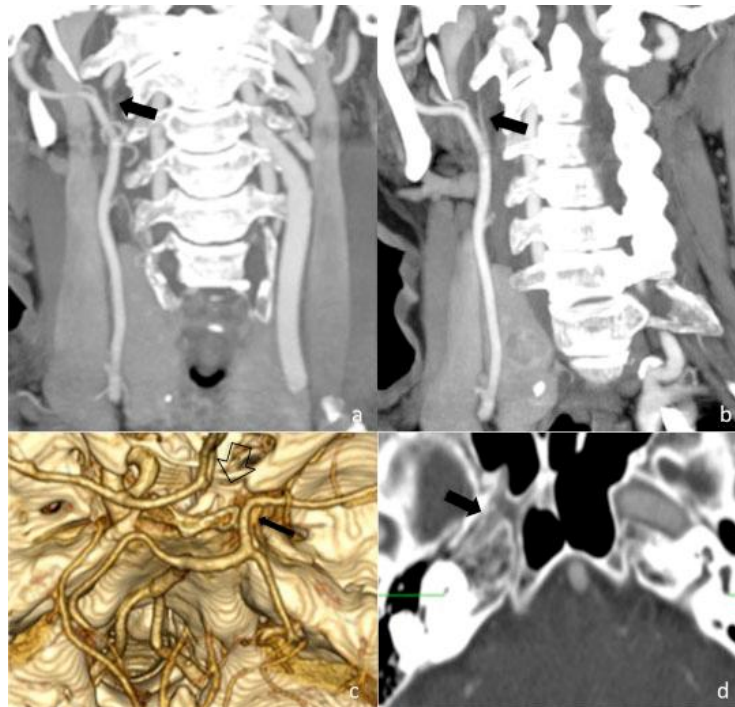


Fig. 1. (a) Coronal MIP CTA image shows the hypoplastic right ICA (arrow) and CCA. (b) sagittal oblique MIP CTA image demonstrates the hypoplastic right ICA (arrow). (c) three dimensional VR image demonstrates the large right posterior communicating artery (arrow) and hypoplastic ACA A1 segment (open arrow). (d) Axial CT scan demonstrates the hypoplastic right carotid canal

2.2 CASE 2

A 45- year old female patient admitted to the hospital with headache and hypoesthesia in the right mandibular and maxillary nerve innervation area. Physical and neurological examinations were normal. She had two births with no problems. She had medical treatment for hypertension and her tension was normal when she admitted to hospital. The laboratory tests were normal. Magnetic resonance imaging showed absence of the normal flow-voids in the cavernous and petrous portion of the right ICA.

On MR angiography all segments of the the right ICA were hypoplastic (Fig. 2a). CT angiography of the head and neck showed right aberrant subclavian artery (Fig. 2b). The right vertebral artery and right ICA were hypoplastic. The caliber of the right CCA was smaller than the left, measured 4.2 mm and 8.2 mm respectively. The right ICA was 1.2 mm in diameter and the right carotid canal was hypoplastic (Figs. 2c and d). CT angiography revealed a bilobulated 8x4 mm fusiform aneurysm of the P1 segment of the right posterior cerebral artery (Fig. 2e).

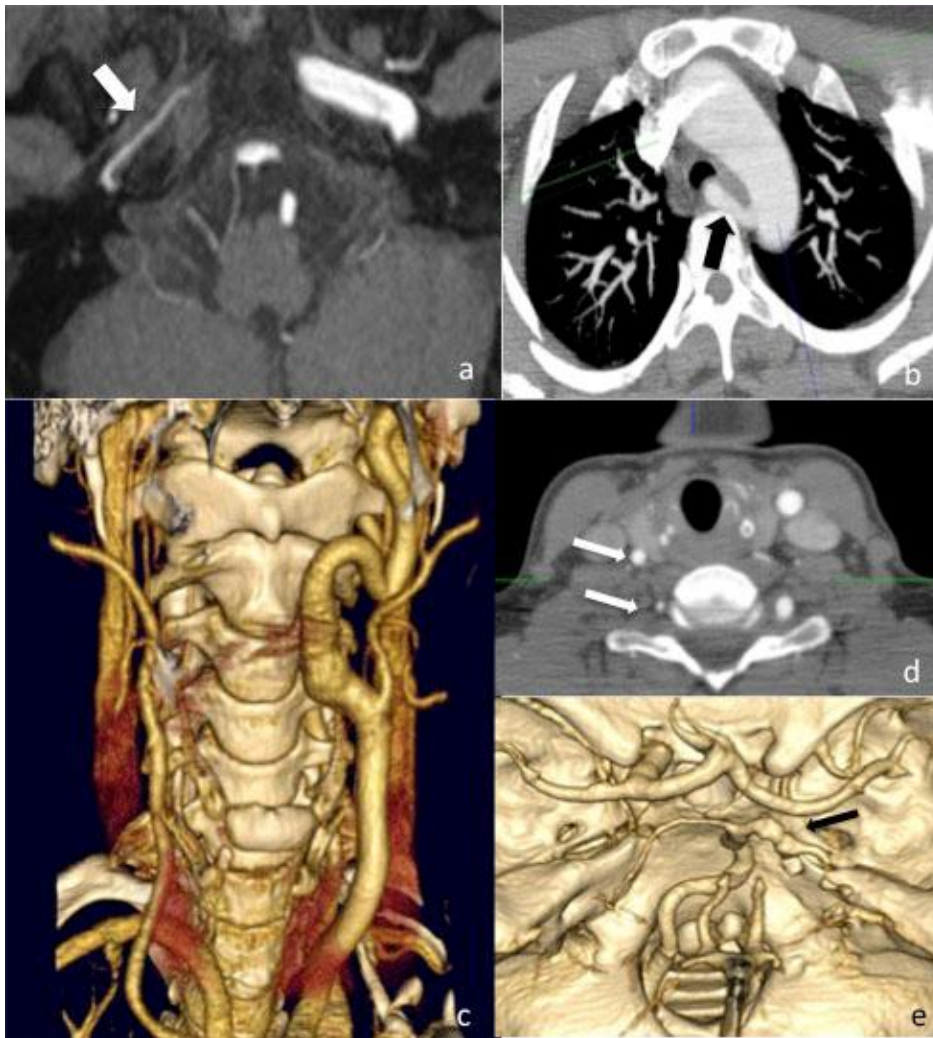


Fig. 2. (a) 3D TOF MR angiography demonstrates the petrous segment of the right hypoplastic ICA (arrow). (b) axial MIP CT image demonstrates the aberrant right subclavian artery. (c) 3D volume rendered CT angiographic image demonstrates the hypoplastic right ICA. (d) Axial CT image shows the hypoplastic right vertebral artery and right CCA (arrows). (e) 3D volume rendered CT image shows the bilobulated fusiform aneurysms of the right PCA P1 segment (arrow)

2.3 Case 3

A 49-year-old male patient was admitted to the hospital with complaint of transient right side visual loss lasting 5-10 seconds that had occurred two times in a week. Amaurosis fugax diagnosis was made and the patient was referred to the radiology department for doppler ultrasonography examination of the carotid arteries. On color Doppler ultrasonographic examination the right ICA caliber was 1.7 mm. Low amplitude and high resistant waveforms were determined in the right ICA so the patient referred to CT angiographic examination for seeking distal occlusion or serious stenosis. CT angiography revealed hypoplasia of the right ICA and the right carotid canal. The left CCA, middle and anterior cerebral arteries, anterior and posterior communicating arteries were normal (Figs. 3a and b).

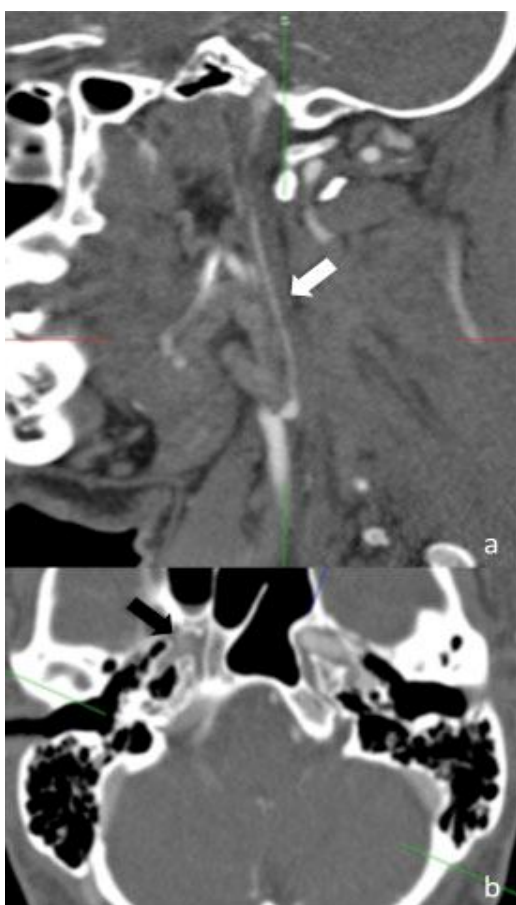


Fig. 3. (a) Sagittal oblique MIP image demonstrates the hypoplastic ICA. (b) axial CT image demonstrates the hypoplastic right carotid canal

3. DISCUSSION

ICA hypoplasia is extremely rare congenital anomaly and the incidence of ICA dysgenesis is estimated as % 0.01 (1,2). They have been classified by Lie into three groups: Agenesis, aplasia and hypoplasia (2). Hypoplasia of the ICA is distinguished from aplasia by the presence of patent but reduced caliber ICA. Aplasia is referred when the ICA and the carotid channel are completely absent (2). The mechanism of development of hypoplasia of the ICA is unclear. Some authors have suggested secondary regression of ICA following a phase of normal development, whereas others consider it to represent arrest of the development of ICA [6,7].

Hypoplasia of the ICA is usually asymptomatic due to the development of collateral vessels [2]. The collateral circulation may be in the form of a transcranial anastomosis with external carotid artery with persistent embryonic arteries or with anastomotic channels in the circle of Willis [8]. Hypoplasia of the ICA is usually discovered incidentally as in the presented cases. However, various clinical presentations are reported, such as, headache, epilepsy, cerebral ischemia, hemiplegia or intracranial haemorrhage [9,10].

Uniform narrowing of the luminal diameter of the ICA may result from developmental hypoplasia but also from a variety of acquired conditions such as atherosclerosis, tubular fibromuscular dysplasia, moyamoya disease, dissection and arteritis. Hypoplasia of the ICA must be differentiated from these acquired ICA stenoses because acquired stenosis could require proper treatment such as endarterectomy or anticoagulation therapy. The differentiation can be made by color doppler ultrasonography (CDS), CTA, magnetic resonance angiography or digital subtraction angiography (DSA). Owing to its cost, availability, noninvasiveness and accuracy in analysing vessel wall and flow, CDS is the first imaging method to rule out carotid disease. DSA is an invasive imaging method. Unlike DSA, both CDS and CTA can also detect the abnormalities of the vessel wall. CTA with its better spatial resolution and lack of flow related artefacts comparing with MRA is the appropriate imaging method for assessing ICA hypoplasia. In congenital hypoplasia, the entire ICA caliber is too small and there is no thickening of the vessel wall. On the contrary, in most acquired diseases, the external caliber of the ICA is normal and

intenal luminal narrowing is due to wall thickening [11,12]. Other important radiologic finding is carotid channel hypoplasia. The carotid canal closely linked to the development of the ICA during embryonic life. The skull base develops between the 5th and 6th weeks of embryonic life and is associated with ICA development. The presence of a developing ICA is essential for the formation of the carotid canal [3,13]. Thus the carotid canal is absent in ICA agenesis, small and not well developed in cases of hypoplasia, as in our cases. In acquired stenosis of the ICA, the carotid canal is normal in size.

The ICAs are derived from portions of the first and third aortic arches and paired dorsal aorta, when the embryo has attained the 3-mm stage [14]. The roots of the ICAs are formed from the third aortic arches. The dorsal aorta form the intermediate portions of the ICAs and the distal part of the ICAs are formed from the first aortic arches. Hypoplasia of the ICA depends on the abnormal regression or involution of the first and third aortic arches and dorsal aorta in the 20-24-mm embryonic stage (2). Collateral circulation accompanying ICA hypoplasia can be classified into three forms; 1) through the circle of Willis, 2) persistent embryonic vessels, 3) transcranial anastomosis from the ECA (4).

In cases of unilateral or bilateral ICA agenesis or hypoplasia, the posterior communicating artery most commonly enlarges and supplies the MCA and the ACA areas [15]. In the presented cases, ICA hypoplasias were seen on the right side. In one patient the right posterior communicating artery was larger than the left and the right MCA and ACA were supplied by the right posterior communicating artery. The posterior communicating arteries were normal in caliber in the other two patients with no persistent embryonic vessels and we thought the contralateral ICA supplied the right MCA and ACAs via the anterior communicating artery.

There is an increased risk of aneurysm formation in patients with hypoplastic or aplastic ICAs. The incidence of intracranial aneurysm in association with ICA agenesis and hypoplasia has been reported as % 25-43 (5). This could be due to increased hemodynamic stress as a result of increased flow through the collaterals or may be due to genetic disorder that has led to both hypoplasia of the ICA and vessel wall weakness, predisposing to aneurysm formation (4). We determined aneurysm of the P1 segment of the right posterior cerebral artery in one patient.

Amaurosis fugax is monocular visual loss ultimately occurs due to a temporary reduction in retinal artery, ophthalmic artery or ciliary artery according to the embolic and hemodynamic causes. Unilateral hypoplasia of the ICA or other vascular congenital anomalies (mainly cerebral aneurysms) is an infrequent cause of amaurosis fugax or TIA, with a clear remark that ischemic stroke or TIA secondary to embolization from vascular congenital anomalies was observed only in 2 of 70 cases in a clinical study of ischemic stroke of unusual cause [16].

4. CONCLUSION

ICA hypoplasia may be discovered incidentally. However, associated life-threatening conditions such as cerebral ischemia or subarachnoid hemorrhage can be seen. This rare anomaly must be distinguished from ICA stenosis or occlusion. The hypoplasia of ICA and carotid canal should prompt further evaluation to rule out the presence of intracranial aneurysms even in an asymptomatic patient. In addition, the collateral circulation and associated aneurysms hold a critical importance whilst performing manipulation around the sellar portion, carotid endarterectomy, transsphenoidal hypophyseal surgery or inducing hypotension, because such an iatrogenic manipulation may disturb compensatory collateral circulation.

CONSENT

All authors declare that 'written informed consent was obtained from the patients for publication of these case reports and accompanying images.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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