

CASE REPORT

Left internal carotid artery agenesis associated with communicating arteries anomalies. A case report

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Abstract

Agenesis, aplasia and hypoplasia of the internal carotid artery are rare congenital malformations. They are usually asymptomatic and incidentally discovered through ultrasound or imagistic tests. The aim of this study is to improve their management in our Departments. We report here the case of a 39-year-old woman addressed to our ambulatory in 2013 for benign symptoms like dizziness and headache. Imagistic findings (magnetic resonance imaging of the brain, and cervical spine, and magnetic resonance angiography of the head and neck) indicated a very rare condition: left internal carotid artery agenesis accompanied by the absence of the pre-communicant part of the left anterior cerebral artery and of the right posterior communicating artery. Internal carotid artery agenesis is an uncommon congenital anomaly and it could be misdiagnosed as stenosis/occlusion of this artery. This condition is important to be recognized due to the associated hemodynamic changes and in order to discover and evaluate other accompanying vascular malformations (aneurysms, collateral channels) and their life threatening potential risks (subarachnoid hemorrhage or ischemia). Also, it has a special importance in case of planning carotid or trans-sphenoidal hypophyseal surgery.

Keywords: agenesis, carotid artery, communicating arteries, magnetic resonance angiography.

Introduction

Agenesis, aplasia and hypoplasia of the internal carotid artery (ICA) are very rare congenital malformations occurring in about 0.01% of the general population [1–3]. These anomalies are often asymptomatic and can be occasionally identified through duplex ultrasonography or imagistic tests [4, 5].

These conditions may be confused with other carotid arteries pathologies as dissection, atherosclerosis or fibromuscular dysplasia [6].

The subjects diagnosed with these types of malformations can lead a normal life due to the development of an extensive collateral vascularization, but this adaptation does not come without risks in the form of potential pathological modification of the collateral vessels (e.g., aneurysms) which can cause subarachnoid hemorrhage [2].

The aim of this study is to improve the management of this rare malformation, emphasizing the importance of understanding their anatomical and imaging features.

Case presentation

We report the case of a 39-year-old woman presented in our Ambulatory Care Unit in October 2013 for recurrent uncharacteristic dizziness and headaches with a long, undetermined onset, but an increasing frequency in the last few months. Our patient had no relevant medical or surgical history, no allergies and a non-contributory

family history. The general physical examination was within normal limits. The neurological examination was also nonspecific. The patient was awake, alert and cooperative; there was no evidence of language disturbance or aphasia; the cranial nerves (I–XII) were intact; the patients had normal bulk and tone and the muscle strength was 5/5 throughout; the reflexes were 2+ in the upper and lower extremities, plantars downgoing bilaterally; the sensation was intact including a negative Romberg; there were no evidence of dysmetria or dysdiadochokinesia; the gait was also normal – the patient being able to toe, heel and tandem walk without difficulty.

In order to exclude brain and cervical spine conditions which could cause these nonspecific symptoms in a young woman, the medical staff performed magnetic resonance imaging (MRI) of the brain and of the cervical spine, and magnetic resonance angiography (MR-A) of the head and neck.

Brain MRI revealed no pathological modifications.

MR-A of the head confirmed the absence of several arterial vessels: left ICA, pre-communicant (A1) part of the left anterior cerebral artery (ACA) and right posterior communicating artery (PCOA); it revealed compensatory increased size of bilateral vertebral artery (VA), basilar artery (BA), and left PCOA; it showed the compensatory collateral circulation to the left ACA through a patent anterior communicating artery (ACOA) and to the left middle cerebral artery (MCA) from the posterior circu-

lation through the enlarged left PCOA, and did not find any aneurysms or any other malformations (Figures 1–3).

MR-A of the neck revealed a filiform trajectory of the left common carotid artery (CCA) and no flow in the left ICA starting with its emergence from the left CCA (Figure 4).

On cervical spine MRI were found a mild C5–C6 posterolisthesis and a C7 radiculopathy (consecutive to a degenerative C6–C7 spinal stenosis) (Figure 5).

The symptoms accused by our patient resolved under medical symptomatic treatment and, until now, no complications of the vascular abnormalities described have been registered.

The discovery of these vascular malformations being incidental, no surgical or interventional corrections were considered necessary. However, we recommended annual clinical and imaging evaluation.

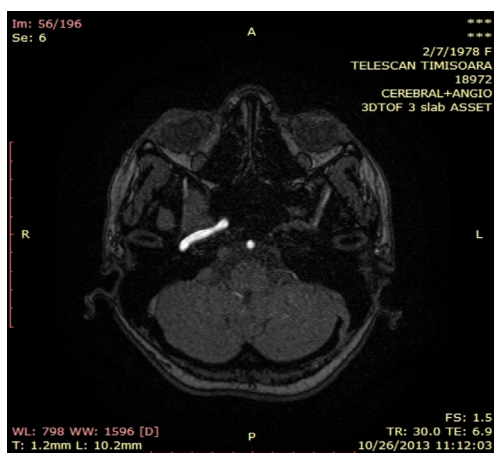


Figure 1 – 3D-TOF MR-A of the head (circle of Willis) (fast) shows normal flow in the right intracranial ICA and no visibility of the left intracranial ICA and carotid canal. 3D-TOF: Three-dimensional time-of-flight; MR-A: Magnetic resonance angiography; ICA: Internal carotid artery.

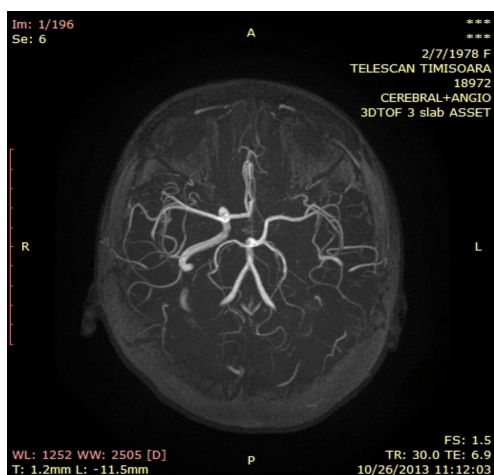


Figure 2 – 3D-TOF MR-A of the head (circle of Willis) (reconstruction) shows the default of left ICA, A1 part of the left ACA and right PCOA; in addition, it demonstrates a compensatory increased size of both VAs, BA, ACOA and left PCOA. 3D-TOF: Three-dimensional time-of-flight; MR-A: Magnetic resonance angiography; ICA: Internal carotid artery; ACA: Anterior cerebral artery; PCOA: Posterior communicating artery; VAs: Vertebral arteries; BA: Basilar artery; ACOA: Anterior communicating artery.

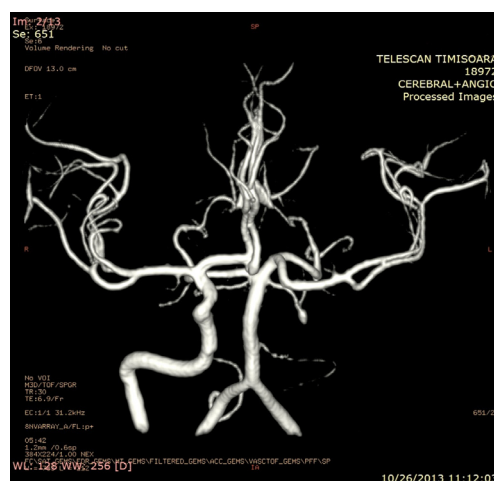


Figure 3 – MIP MR-A of the head (circle of Willis) shows collateral circulation to the left ACA (only A2 post-communicant ACA) through a patent ACOA and to the left MCA from the posterior circulation through an enlarged PCOA. MIP: Maximum-intensity projection; MR-A: Magnetic resonance angiography; ACA: Anterior cerebral artery; ACOA: Anterior communicating artery; MCA: Middle cerebral artery; PCOA: Posterior communicating artery.



Figure 4 – MR-A of the neck (coronal section) shows a reduced caliber of the left CCA and absence of flow in the left ICA from its emergence. MR-A: Magnetic resonance angiography; CCA: Common carotid artery; ICA: Internal carotid artery.

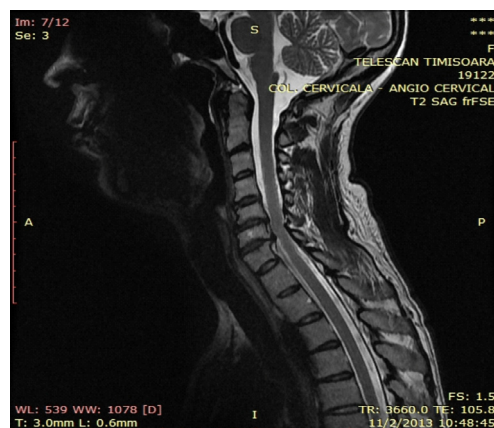


Figure 5 – Cervical spine MRI. T2 sagittal images reveal a mild C5–C6 posterolisthesis and a C7 radiculopathy consecutive to a degenerative C6–C7 spinal stenosis. MRI: Magnetic resonance imaging.

Discussions

The ICA forms from the dorsal aorta and the third aortic arch (4–5 mm embryonic stage) in the 24th day of embryogenesis and it will be completed by the 6th week of embryonic life [3, 7–9]. The definitive CCA, external carotid artery (ECA) and its branches are formed by the 40th day (16–18 mm stage). The carotid canal formation occurs at the same time and it is dependent on the development of the ICA, therefore, the congenital absence of the artery is accompanied by the absence of the carotid canal [6, 8, 10].

The development anomalies of the ICA can be classified as follows: (a) agenesis (complete failure of arterial development – absence of both ICA and carotid canal); (b) aplasia (lack of development – presence of vestiges of non-patent vessels and also of the carotid canal), or (c) hypoplasia (incomplete development of the artery – one of the ICA with a reduced but patent vascular lumen accompanied by a smaller but normally structured carotid canal) [2, 3, 9, 11]. The absence/presence of the bony carotid canal can be observed on the computed tomography (CT) scan of the skull base. This information has a particular importance since it will differentiate aplasia from agenesis and also hypoplasia (associated with a diminutive carotid canal) from acquired conditions of the ICA where the carotid canal has a normal size and structure [3, 6, 12, 13].

The causes of agenesis/aplasia of the ICA (secondary to a possible atresia/involution of the third aortic arches and the distal portion of the dorsal aortas) could be mechanical (pressure effects), excessive bending of the cephalic end of the embryo to one side or the other, or constrictions by amniotic bands [1, 8, 10, 14].

The congenital developmental malformations of the ICA are very rare. Moreover, it is reported that the aplasia/agenesis of the left ICA is three times more frequent than the right or the bilateral ICA's anomalies [1, 5].

The circle of Willis forms during the 7 to 24 mm stage of embryonic development. If the ICA is absent, the collateral system pattern necessary for a proper cerebral vascularization depends on the stage in which the disruption occurs. Thus, according to Cali *et al.*, the collateral will originate from primitive pathways (e.g., inter-cavernous anastomoses) if the disruption occurs before the 24 mm stage, and, respectively, from circle of Willis after its complete development (the most frequent) [3, 15].

According to Lie, there are six possible pathways of collateral circulation associated with the absence of the ICA:

(a) Type 1 – collateral circulation to the ipsilateral ACA through a patent ACOM and to the ipsilateral MCA from the posterior circulation through a hypertrophied PCOA (also our case);

(b) Type 2 – the ipsilateral ACA and MCA are supplied across a patent enlarged ACOA;

(c) Type 3 – bilateral agenesis of the ICA with supply to the anterior circulation *via* carotid-vertebro-basilar anastomoses through hypertrophied PCOAs;

(d) Type 4 – unilateral agenesis of the cervical portions of the ICA with an inter-cavernous communication to the ipsilateral carotid siphon from the contralateral ICA;

(e) Type 5 – the ACAs are supplied by bilateral hypoplastic ICAs and the MCAs are supplied by enlarged PCOAs;

(f) Type 6 – collateral flow provided *via* transcranial anastomoses from the homolateral internal maxillary branches of the ECA system (*rete mirabilis*) [3, 5, 11].

Others simplified the six collateral pathways mentioned above into three main types as follows:

(1) The fetal type – the ACOA supplies the ACA, and the PCOA supplies the MCA (the most common type and also the one present in the case we described);

(2) The adult form, where the ACOA supplies both ACA and MCA;

(3) Posterior collateral pathways through anastomosis from the ECA, contralateral ICA or from some primitive vessels (the rarest) [1, 3, 6, 9, 16, 17].

Duplex ultrasonography (US) is usually the first test performed on a patient suspected of having congenital or acquired carotid condition. However, for a correct diagnosis and assessment of the possible brain lesion or cerebral perfusion defects (very rare despite the altered vascular anatomy), there are needed more imaging tests including: MR-A/CT angiography (CT-A)/catheter angiography, a skull base CT scan and, if available, a single-photon emission computed tomography (SPECT) [1, 2, 5].

We chose to use brain MRI and brain and neck MR-A in order to investigate the possible cerebral and vascular abnormalities involved in our case.

MR-A as an assessment tool of neck and cerebral vessels proved to be able to identify and differentiate between various lesions and abnormalities in the carotid arteries and the arteries around the circle of Willis. Also, due to its high diagnostic yield and noninvasive nature, MR-A is frequently preferred over conventional angiography [18].

The congenital anomalies of ICA must be differentiated from acquired carotid pathologies, such as craniocervical artery dissection, fibromuscular dysplasia, severe atherosclerosis, moyamoya disease and arteritis [3, 5, 10].

Craniocervical artery dissection

Craniocervical artery dissection is a major cause of ischemic symptoms in young adults, but it frequently remains undiagnosed due to its mild and nonspecific symptoms (headache, dizziness, etc.). In these cases, the MR-A may reveal: smooth or irregular vessel wall, string and pearl sign (focal narrowing with a distal site of dilatation), flame-like occlusion, pseudoaneurysm, intimal flap, double lumen (visible in <10%) or distal branch occlusion. Also, the presence of the periarterial rim of intramural hematoma is demonstrated on the MRI as a hyperintense signal on T1- and T2-weighted images [19, 20].

Fibromuscular dysplasia

Fibromuscular dysplasia is an autosomal dominant disorder, affecting up to 5% of the population. In ~65% of the cases it was reported a bilateral involvement of the ICAs (usually the C2 segment). The angiographic characteristic finding is multifocal alternating stenosis and adjacent dilatations causing a *string of beads* appearance. Less frequently, there is focal concentric, long-segment tubular stenosis or diverticular outpouching present [20, 21].

Carotid atherosclerotic disease

The carotid atherosclerotic disease was easily excluded in our case considering the age of the patient and the absence of cardiovascular risk factors. Also, the MR-A informed the involvement of the carotid bulb and the presence of multifocal atherosclerotic plaques [20].

Moyamoya disease

Moyamoya disease is a hereditary genetic abnormality causing intimal thickening in the walls of bilateral distal ICAs and circle of Willis. The MR-A criteria for the diagnosis of moyamoya disease are (a) bilateral stenooclusive change of the carotid fork and (b) development of moyamoya vessels (dilated perforating arteries in the basal region). It is important to mention that in the very early and advanced stages these moyamoya vessels are difficult to detect. In patients with moyamoya fluid-attenuated inversion recovery (FLAIR) images and post-contrast T1 images may show a linear pattern of increased signal in the leptomeninges and perivascular spaces ("ivy sign"). MRI can also reveal ischemia, infarction, generalized cerebral atrophy, and ventriculomegaly [18, 19, 22, 23].

Arteritis

The arteritis is commonly characterized by narrowing or occlusion of multiple arteries, frequently involving the aorta, its branches, and/or various arteries in the proximal upper or lower extremities (*e.g.*, Takayasu disease). These changes are usually focal or segmental and include stenosis, occlusion, or aneurysms [19]. In addition, it is important to mention that the pathological findings in arteritis like Takayasu disease or large-vessel giant cell arteritis may disappear with corticosteroid therapy [5, 24].

Due to the collateral pathways, the congenital anomalies of ICA are frequently asymptomatic and incidentally found on imaging tests, but some patients present nonspecific symptoms like recurrent headache, blurred vision, hearing impairment, hemiparesis, cranial nerve palsy and convulsions due to modifications in collateral flow with consecutive vascular insufficiency and/or cerebral ischemia [1, 9, 12, 13].

Our patient mentioned only mild symptoms and the diagnosis was incidental, but there were reported cases of cerebrovascular events, such as subarachnoid hemorrhage after rupture of an aneurysm or cerebral infarct. There is an increased risk of aneurysm formation associated with ICAs congenital development anomalies (23–45% compared to 2–4% in general population). The aneurysmal dilatations are more common along ACOA, PCOA, posterior cerebral arteries (PCAs) and the BA. In these cases, the aneurysms formation may be due to increased flow through collateral vessels associated with an increased hemodynamic load on the normal side in the presence of additional risk factors, such as systemic hypertension and vessel wall weakness. The high rate of aneurysms formation is the main reason for a close clinical and imagistic surveillance in these patients [1–3, 10].

Several conditions associated with congenital ICA development malformations were reported. These include congenital hypopituitarism, Horner's syndrome, ischemic

optic neuropathy, spasmodic torticollis (due to an enlarged VA), agenesis of the *corpus callosum*, arachnoid cyst, meningocele, neurofibromatosis, Klippel–Feil syndrome, coarctation of the aorta and anomalies of the BA [2, 6, 9].

Recognition of the congenital anomalies of the ICAs are important in case of thromboembolic disease (emboli in one cerebral hemisphere may come from the contralateral CCA or from the vertebrobasilar system), planning carotid endarterectomy (both cerebral hemispheres may be dependent on the single, atheromatous ICA) or trans-sphenoidal hypophyseal surgery (the absent ICA can be associated with intracavernous collateral) [3, 5, 25].

Conclusions

We reported a case of agenesis of the left ICA associated with communicating arteries anomalies, but without aneurysmal modifications or brain lesions. Agenesis of the ICA is a rare but important condition that is usually found incidentally. Duplex US is generally the first test used in the assessment of ICA pathology, but for an accurate diagnosis of congenital development malformations of the ICAs imaging tests such as MR-A, CT-A, and CT of the skull base are needed. In our opinion, the ICA development anomalies have to be promptly recognized and periodically evaluated both clinically and imagistically, due to the possible associated hemodynamic changes and the possible implications during carotid endarterectomy or trans-sphenoidal hypophyseal surgery.

Conflict of interests

The authors state that there is no conflict of interests.

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