Bilateral internal carotid artery agenesis: a case report

Agenesia bilateral de artéria carótida interna: relato de caso

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Abstract

Agenesis of the internal carotid artery (ICA) is defined as a congenital absence of the carotid canal and occurs in less than 0.01% of the population. This anomaly is usually diagnosed as an incidental finding or after a cerebrovascular event. We present the case of a 36-year-old woman, with bilateral agenesis of the ICA who had suffered a subarachnoid hemorrhage due to ruptured aneurysm of the basilar artery. Therefore, agenesis of the ICA is a condition that, although rare, should be considered since there is a risk of coexistence with other life-threatening conditions such as aneurysms.

Keywords: internal carotid artery; carotid artery diseases; intracranial aneurysm.

Resumo

Agenesia da artéria carótida interna (ACI) é definida como uma ausência congênita do canal carotídeo e ocorre em menos de 0,01% da população. O diagnóstico dessa anomalia ocorre geralmente como um achado incidental ou depois de um evento cerebrovascular. Apresentamos o caso de uma mulher de 36 anos, hipertensa e tabagista, com agenesia bilateral de ACI, que abriu quadro com hemorragia subaracnoide decorrente de rotura de aneurisma de artéria de basilar. A agenesia da ACI é, portanto, uma condição que, embora rara, deve ser lembrada por estar associada a outras más formações potencialmente fatais, como os aneurismas cerebrais.

Palavras-chave: artéria carótida interna; doenças das artérias carótidas; aneurisma intracraniano.

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INTRODUCTION

Agenesis of the internal carotid artery (ICA) is defined as a congenital absence of the carotid canal and is very often found in conjunction with other arterial abnormalities in the Circle of Willis.[1]

Agenesis, aplasia and hypoplasia of the ICA are rare congenital anomalies, occurring in less than 0.01% of the population.[2] In the majority of cases, absence of one or both of the internal carotid arteries does not manifest in any symptomology. This is because of collateral vessels, such as persisting embryonic vessels, the normal anastomosis routes via the Circle of Willis or anastomoses with the external carotid arteries. As a result, the anomaly is generally diagnosed as an incidental finding or after a cerebrovascular event, such as a stroke or a subarachnoid hemorrhage after rupture of an aneurysm.[3]

Other patients, in contrast, may exhibit signs related to cerebrovascular insufficiency or vascular complications caused by a concomitant arterial malformation. In these cases, patients can display a variety of symptoms ranging from recurrent headaches and blurred vision to hemiparesis and lowered states of consciousness.[4]

Although it can be detected using computed tomography angiography (angio-CT) or magnetic resonance angiography (angio-MRI), the gold standard examination for diagnosis of ICA agenesis remains conventional angiography.[5]

It is important to point out that ICA agenesis, when present, is generally unilateral and more commonly on the left.[5] The ratios between prevalence rates of ICA agenesis on the right-hand side, on the left-hand side, and bilaterally are 1:3:1, respectively.[3]

CASE DESCRIPTION

We describe the case of a 36-year-old female smoker, already known to have hypertension, for which she was not receiving regular treatment, who presented with a sudden case of strong intensity holocranial headaches combined with projectile vomiting and lowered state of consciousness.

An initial examination conducted on admission to a walk-in center at a tertiary hospital found that she was drowsy, arousing when called, exhibited confused speech, withdrew all four limbs in response to painful stimulus and exhibited discrete brachio-crural hemiparesis on the left and rigidity at the base of the skull. In view of the above, the patient was scored 11 on the Glasgow coma scale and classified as grade III on the Hunt-Hess scale.

A cranial computed tomography scan was conducted, demonstrating meningeal hemorrhage on the right and bilateral agenesis of the carotid canal. In view of this, the patient underwent angiography, which showed an absence of opacification of the internal carotid arteries, bilaterally from the origins, in addition to an irregular saccular dilation at the top of the basilar artery (Figures 1, 2 and 3). Endovascular treatment was conducted using platinum coils and hydrocoil, achieving total occlusion of the aneurysm (Figure 4).

The patient was admitted to the intensive care unit for postoperative recovery, where she remained for four days with no intercurrent conditions. She was discharged from hospital seven days after admission. Currently, two years after the procedure, she is in outpatients follow-up and exhibits discrete

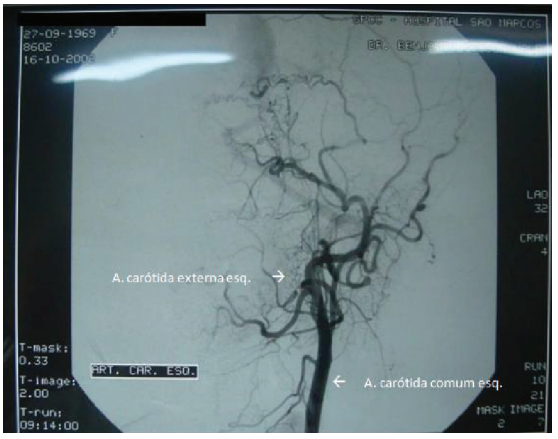

Figure 1. Selective arteriography of the left carotid system, cervical portion, lateral view; the left internal carotid artery is not seen in contrast.

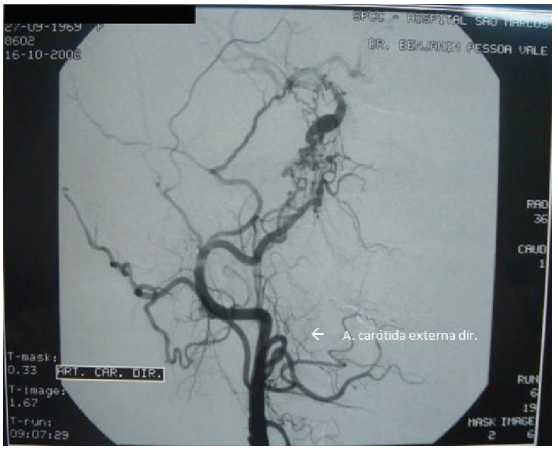

Figure 2. Selective arteriography of the right carotid system, cervical portion, lateral view; the right internal carotid artery is not seen in contrast.

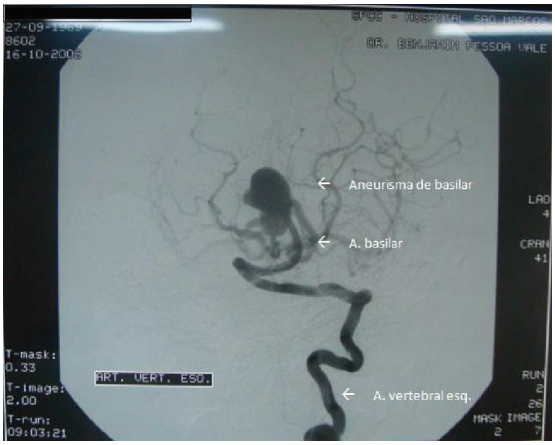

Figure 3. Selective arteriography of the left vertebrobasilar system, intracranial portion, anteroposterior view: aneurysm at top of basilar artery (preoperative image).

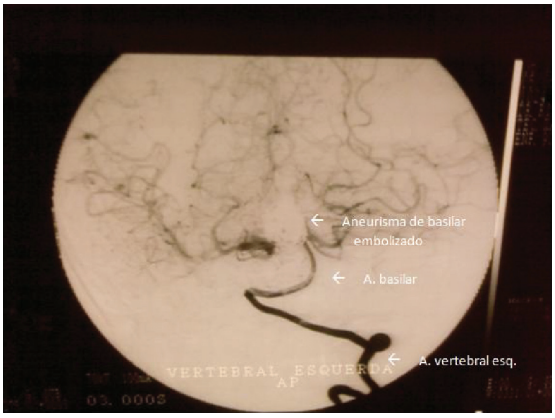

Figure 4. Selective arteriography of the left vertebral-basilar system, intracranial portion, anteroposterior view: aneurysm at top of basilar artery after endovascular treatment with platinum coils and hydrocoil.

hemiparesis on the left which does not limit her daily activities.

DISCUSSION

Agenesis of the ICA is a rare congenital anomaly, with a little over 100 cases reported since the first was described by Tode, in 1787, after dissection of a cadaver.[3,6] We were able to locate just 26 cases of bilateral internal carotid artery agenesis in the literature.[7]

The ICA originates from the cranial portion of the dorsal aorta and the terminal segments of the primitive third aortic arch and is fully developed by around the sixth week.[8] Agenesis or aplasia of the internal carotid artery is therefore related to atresia or involution of the third aortic arch and the distal portion of the dorsal aorta, while still in the initial embryonic phase. In turn, the base of the skull begins to form after the fifth or sixth week of fetal life. Therefore, if the embryonic precursor of the ICA has not developed by the third to fifth week, the ICA and the carotid canal will never develop.[3]

No precise explanation has been found for development of internal carotid artery anomalies, but it is believed that all variants are the result of insults during embryo development. Keen e col. have suggested that mechanical injuries to developing embryos, such as excessive bending of the embryo to one side and the effects of pressure or restriction by amniotic bands, may be conducive to absence of the internal carotid artery.[4]

The natural history of patients with absent internal carotid artery is not entirely clear.[9] It is known that the majority of patients who have this anomaly remain asymptomatic because arterial supply is compensated by collateral circulation.[10] It is also known that there is a greater risk of cerebral aneurysm in the Circle of Willis among patients with internal carotid artery agenesis.[9] As a result, initial clinical presentation tends to be a cerebrovascular event such as a stroke or a subarachnoid hemorrhage after rupture of an aneurysm, as occurred in the case described here.[10]

It is estimated that the prevalence of intracranial aneurysms in the general population ranges from 2 to 4%. In contrast, among patients with congenital absence of the ICA, this range increases to 24 to 34%.[4] Patients with bilateral internal carotid artery agenesis are at high risk of development of aneurysms, particularly in the posterior communicating and basilar arteries, as was seen in the case described here.[9]

The greater likelihood of intracranial aneurysm formation may be the result of abnormal hemodynamic forces, related to collateral compensation in arterial vessels or to congenital defects of the integrity of the internal carotid artery or other arteries.[4]

Conditions such as ICA agenesis which involve a certain degree of hemodynamic impairment may comprise an additional trigger factor for development of significant comorbidities.[10] Many different authors stress the importance of conducting periodic imaging studies of the cerebral vessels of patients with ICA agenesis, with the objective of screening for development of intracranial aneurysms.[9] Studies have shown that early recognition of anomalies involving the carotid system can prevent potentially fatal complications.[10]

Conventional angiography is more sensitive than angio-CT or angio-MRI for detection of cerebral aneurysms. Notwithstanding, angio-MRI is recommended for screening and monitoring of aneurysms in children because it is a noninvasive technique.[9]

Congenital ICA absence can be seen in association with other syndromes, particularly in the pediatric population. Associations that have been described previously include mastoid hypoplasia, Horner syndrome and congenital hypopituitarism.[9]

CONCLUSIONS

Internal carotid artery agenesis is a condition that, although silent, can be associated with other potentially serious malformations and disorders. In general, patients with bilateral ICA agenesis are at high risk of development of intracranial aneurysms. Diagnosis of anomalies involving the carotid system should be the signal for rigorous follow-up focused on early identification of potentially fatal changes, such as cerebral aneurysms.

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