CASE REPORT

Left internal carotid artery agenesis in a patient with headache

Agenesia da artéria carótida interna em um paciente com cefaleia

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ABSTRACT

Sometimes in clinical neurology, we diagnose a very rare case. We report on a patient who presented with crisis of headache and vomiting (clinically diagnose as migraine). Computed tomography (CT) scan of the head did not reveal any structural lesion. Magnetic resonance angiography showed absence of left internal carotid artery associated with absence of the left middle cerebral artery (MCA).

Keywords: Carotid artery; Agenesis; Middle cerebral artery; Magnetic resonance angiography; Headache

RESUMO

Às vezes em neurologia clínica, nós diagnosticamos um caso muito raro. Nós relatamos o caso de um paciente que apresentava crises de dor de cabeça e vômitos (diagnóstico clínico de enxaqueca). Tomografia computadorizada da cabeça não revelou qualquer alteração estrutural. Angiografia por ressonância magnética mostrou ausência de artéria carótida interna esquerda associada à ausência de artéria cerebral média esquerda.

Palavras-chaves: Artéria carótida; agenesia; artéria cerebral média, Angiografia por ressonância magnética; cefaleia

INTRODUCTION

Agenesis of internal carotid artery (ICA) is a rare congenital anomalywith an incidence of 0.01%.[1,2] Most of the patients are asymptomatic and is usually discovered incidentally by computed tomography (CT) or magnetic resonance imaging (MRI).

In 1954, Tode et al.[3]described the first case of this condition and, in 1954, Verbiest et al.[4]demonstrated the absence of the ICA by angiography for the first time.

The most common source of collateral circulation in the case of the ICA agenesis is via the circle of Willis.[5] In these cases, the basilar artery or the contralateral ICA supplies the middle cerebral artery (MCA) and anterior cerebral artery (ACA) on the side of the absent ICA.[5]

The cause of the carotid agenesis is not known, but may be secondary to an insult to the developing embryo.[5]

The left internal carotid artery is reported to be affected by dysgenesis three times more often than the right one.[6]

We herein report a case of left ICA agenesis presenting with a history of chronic headaches for the preceding 14 years.

CASE REPORT

A 33-year-old woman, presented to our Headache Ambulatory with a history of chronic headache since 1999. She described pulsatile, right headache,of moderate to strong intensity, worsened by physical activities, associated to nausea, vomiting, photophobia, phonophobia and osmophobia, 1 to 2 crises (episodes) per month. She had family history of migraine and took oral contraceptives.

She came to the emergency room many times because of crisis of acute headache, refractory (resistant) to usual pain killers. In the late months the pain became daily,

soften, bilateral and pressing. She related abuse of pain killers.

A psychiatric treatment was indicated because she had anxiety and insomnia. Also, she was going through dental control due to myofascial pain. She is using stabilizing splint and does physiotherapy for temporo-mandibular joint dysfunction.

Her pain was refractory to various pain-killers and non-steroidal anti-inflammatory drugs, such as naproxen; cefalium®; naratriptan; sumatriptan; ibuprofen and ergotamine.

She used many preventive medicines against migraine, with partial results, e.g. fluoxetina; bupropion; venlafaxine; alprazolam; clonazepan; atenolol and cyclobenzaprine.

In the beginning, she made CT scan (computerized tomography), which was normal.

Angio RM (magnetic resonance angiograph) of the brain and of the carotid and vertebral arteries in 3.September.2013 showed left ICA agenesis (Figures 1 and 2).

Nowadays she is taking mirtazapine (30mg/day), sumatriptain and ketorolac with good response.

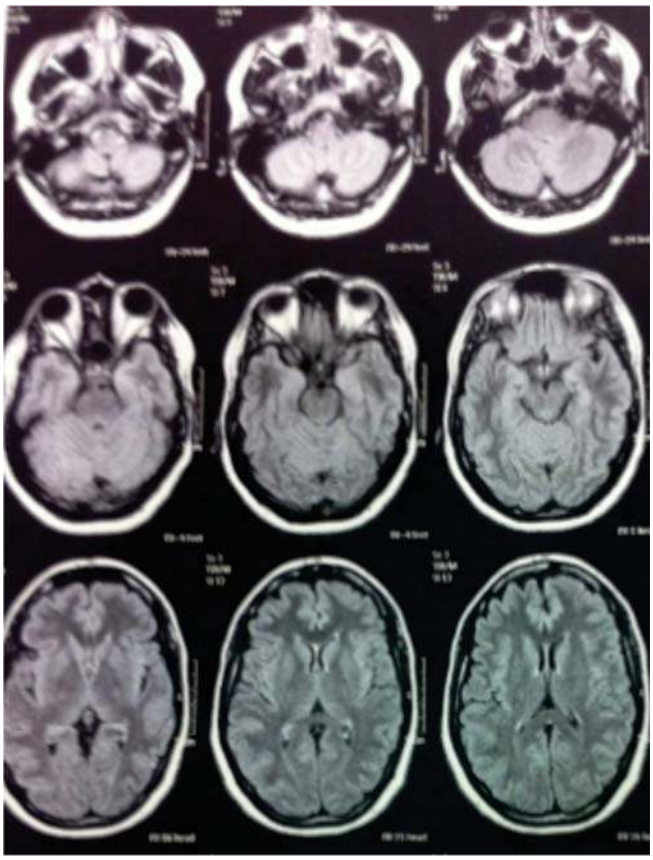

Figure 1. Magnetic resonance imaging.

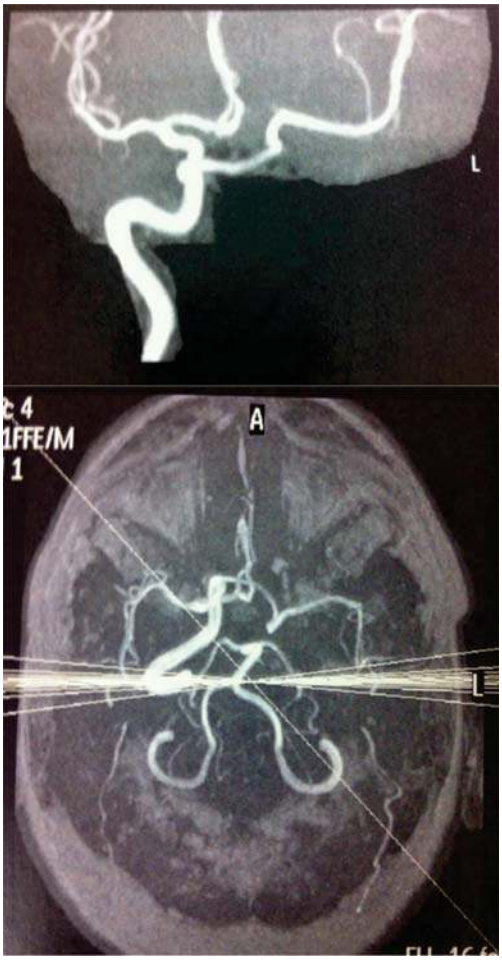

Figure 2. Angio-MR.

DISCUSSION

Embryologic considerations

Alterations of the embryologic development of the cerebral arterial circulation may lead to agenesis or hypoplasia of the carotid arteries.

According to Streeter,[7]] Padget[8]and McLone and Naidich,[9] two branches of the primitive ICA develop early in the embryogenesis, originating from the third aortic arch. In its primitive form, the internal carotid reaches the cephalic region up to the level of the Rathke`s pouch where two primary divisions occur. One cranial branch extends anteriorly to supply the developing forebrain via the anterior choroidal, middle cerebral, anterior cerebral and primitive olfactory arteries.

A second posterior branch gives rise to the posterior choroidal, diencephalic and mesencephalic arteries. As these branches advance caudally, anastomoses are made

with the developing longitudinal neural arteries supplied by the trigeminal artery connections to the primitive ICA. Agenesis or aplasia of ICA results from abnormal regression of the first and third aortic arch[10,11].

Lie[5] referred to agenesis as a complete absence of the entire ICA and carotid canal.

Clinical and radiological consideration

ICA agenesis is a rare congenital anomaly, with an estimated incidence of 0.01%, with more frequent symptom encountered in adults rather than in children or adolescent patients.[1,2] Relatively a small number of symptomatic cases of absence of the ICA have been reported in children, suggesting that initially the collateral pathways are sufficient to support cerebral perfusion[5]. This condition is known to occur more frequently in left sided ICA`s. The ratio between right and left ICA is 1:3[12].

Patients with agenesis of the ICA are often asymptomatic because of the collateral pathways. However, patients may present with headaches, seizures or neurological deficits secondary to cerebral ischemia, or intracranial hemorrhage[13]. Our patient had recurrent headache increases clinically diagnosed as migraine.

Diagnosis of ICA agenesis entails documenting the absence of the ICA by conventional angiography, MRA or MSCT angiography, and the absence of the carotid canal by CT. Collateral flow is most commonly supplied through the circle of Willis, but may be also provided by embryonic vessels or the external carotid artery. MRA appears to be an effective and sensitive method for detecting absence of the ICA. ICA agenesis is usually encountered during the diagnosis process.

Our patient has no complaints at present and is being followed up with periodic physical and neurological examination. We believe that these findings in our patient are instructive and will help to further our understanding of the embryologic development of the carotid arteries.

CONCLUSIONS

ICA agenesis is a rare vascular anomaly and is a typically discovered incidentally as patient are often asymptomatic. In our case, this patient has presented in our ambulatory with complaint of crisis of refractory headache, diagnosed as migraine.

This rare anomaly must be distinguished from ICA stenosis or occlusion.The collateral circulation should also be documented by conventional angiography, particularly prior to carotid endarterectomy, transphenoidal hypophyseal surgery or inducing hypotension. Collateral flow is most commonly supplied through the circle of Willis, but may be also provided by embryonic vessels or the external carotid artery.

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