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ISOLATED OCULOMOTOR NERVE PALSY IN SPONTANEOUS INTERNAL CAROTID ARTERY DISSECTION

Case report

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ABSTRACT - Partial oculosympathetic palsy followed by ischemic manifestations in brain or retina are the main symptoms of extracranial internal carotid artery (ICA) dissection. Unusually, cranial nerves may be affected. Isolated oculomotor nerve palsy is found only rarely. *Case:* We present a 50-year-old nondiabetic man who experienced acute onset of right occipital headache which spread to the right retro-orbital region. Five days later he noticed diplopia and right blurred vision sensation. Neurologic examination disclosed only impaired adduction and upward gaze of right eye, slight ipsilateral pupillary dilatation, without ptosis. Brain MRI was normal. Angiography showed right internal carotid artery dissection with forward occlusion to the base of the skull. Intravenous heparin followed by warfarin was prescribed. The headache and the oculomotor nerve deficit gradually resolved in the next three weeks. *Discussion*: Isolated oculomotor nerve palsy is underrecognized as a clinical presentation of extracranial ICA dissection. If the angiographic evaluation is incomplete without careful study of extracranial arteries, misdiagnosis may lead to failure to initiate early treatment to prevent thromboembolic complications. For this reason we draw attention to the need for careful evaluation of cervical arteries in patients with oculomotor nerve palsy. Mechanical compression or stretching of the third nerve are possible mechanisms, but the direct impairment of the blood supply to the third nerve seems to be the most plausible explanation.

KEY WORDS: third nerve; oculomotor palsy; carotid artery; dissection; angiography.

Paralisia isolada do nervo oculomotor na dissecção de artéria carótida interna: relato de caso

RESUMO - A paralisia oculosimpática parcial (síndrome de Horner) seguida por manifestações isquêmicas cerebrais ou retinianas são os principais sintomas da dissecção da artéria carótida interna (ACI) extracraniana. O acometimento de nervos cranianos é incomum. Apenas raramente a paralisia isolada do nervo oculomotor pode ser encontrada. *Caso*: homem de 50 anos, sem diabetes, apresentou cefaléia occipital de início súbito irradiada a seguir para a região retro-orbitária direita. Após 5 dias começou a notar diplopia e visão embassada no olho direito, quando procurou o hospital. O exame neurológico evidenciou apenas prejuízo da adução e do olhar vertical para cima no olho direito, com discreta dilatação pupilar ipsilateral, sem ptose. RNM foi normal. A angiografia digital revelou dissecção da ACI direita com oclusão a montante até a base do crânio. A anticoagulação foi iniciada. A cefaléia e a movimentação ocular melhoraram em 3 semanas. *Discussão*: A paralisia isolada do nervo oculomotor dificilmente é reconhecida como um possível sinal clínico de dissecção da ACI. Se o estudo angiográfico for incompleto, sem a avaliação cuidadosa das artérias extracranianas, esse diagnóstico deixa de ser feito, impedindo assim o início precoce da anticoagulação para a prevenção dos eventos tromboembólicos. Por isso, reforçamos a importância do estudo das artérias cervicais nos pacientes com paralisia do nervo oculomotor. A compressão mecânica ou o estiramento do nervo são mecanismos possíveis, mas o prejuízo direto do suprimento sanguíneo arterial para o nervo é a explicação mais plausível.

PALAVRAS-CHAVE: terceiro nervo; nervo oculomotor; artéria carótida; dissecção; angiografia.

Extracranial internal carotid artery (ICA) dissection is an important cause of ischemic stroke in young and middle-aged patients[1]. Although found rarely, the classical clinical picture is of a patient who presents unilateral pain in the head, face or neck and partial oculosympathetic palsy, which may be followed by ischemic manifestations in the brain or retina some hours or days later[1]. Cranial nerves may

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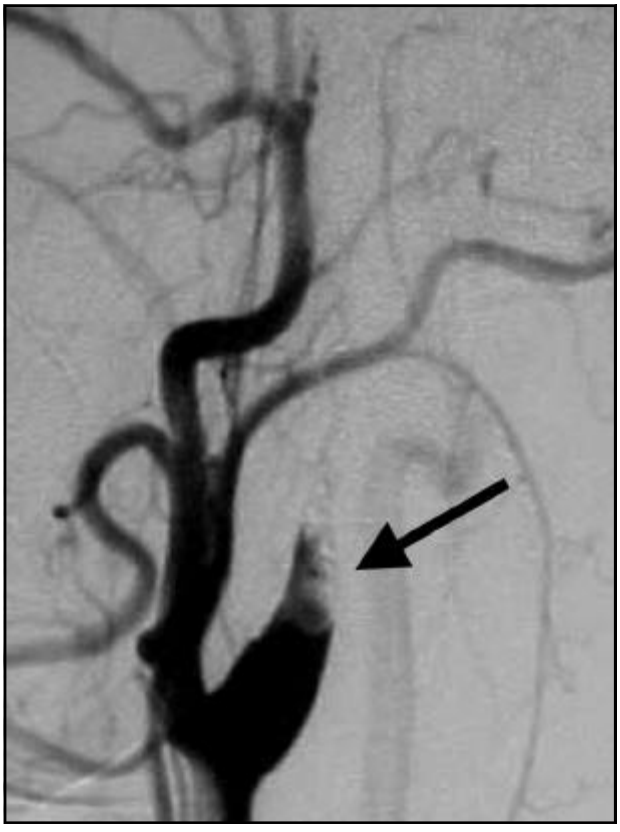

Fig 1. Lateral angiographic view of the right carotid artery showing a tapered, flame-like occlusion (black arrow) suggestive of artery dissection.

be affected in 6% to 12% of patients with carotid artery dissection[2]. The lower cranial nerves are the most commonly affected[3]. Ocular motor dysfunction due to involvement of the third, fourth or six cranial nerve is found only occasionally[4-6] and isolated oculomotor nerve palsy is rarely found.

We describe a patient with this atypical form of presentation of ICA dissection with no associated cerebral or retinal ischemic symptoms. We discuss possible mechanisms involved and draw attention to the appropriate angiographic investigation.

CASE

A 50-year-old nondiabetic man experienced acute onset of intense right occipital headache which spread in hours to the right retro-orbital region. Five days later he noticed acute onset of diplopia and right blurred vision sensation. His medical history was significant for cardiac Chagas' disease diagnosed 10 years before when he presented syncope and cardiac arrhythmia. He was being treated with amiodarone. There was no history of previous trauma and arterial hypertension. He denied transient monocular blindness or hemispheric ischemic attacks. His blood pressure was 120/80 mm Hg. There was no cervical or cranial bruit and cardiac auscultation was normal. Neurological examination revealed impaired adduction and upward gaze of right eye (paresis of the medial rectus and superior rectus muscles) without ptosis. There was a slight ipsilateral pupillary dilatation with poor reaction to bright light. Funduscopic examination, visual acuity and the remainder of the neurological examination were normal.

Findings on head CT were normal. Brain MRI demonstrated no focal lesion, the magnetic resonance angiography showed occlusion of right internal carotid artery. Digital subtraction angiography showed diminishing of the right internal carotid artery caliber suggestive of dissection (Fig 1) with forward occlusion to the base of the skull. There was hypoplasia of the right P1 indicating presence of fetal variant. As ICA blood flow was impaired, the right middle cerebral artery was being supplied by the right posterior communicating artery (Fig 2). Electrocardiogram revealed right branch block and antero-superior division block. Echocardiogram disclosed difuse involvement of heart with mild left ventricle enlargement, but no thrombus was detected.

Intravenous heparin followed by warfarin was prescribed. The headache and the oculomotor nerve deficit gradually resolved over the following three weeks.

DISCUSSION

The most common ophthalmologic manifestations of carotid dissection are oculosympathetic palsy (Horner's syndrome) and transient monocular blindness[7] due to the involvement of the internal part of the pericarotid sympathetic plexus[8] and thromboembolic event respectively. Cranial nerve palsies due to ocular motor dysfunction are uncommon manifes-

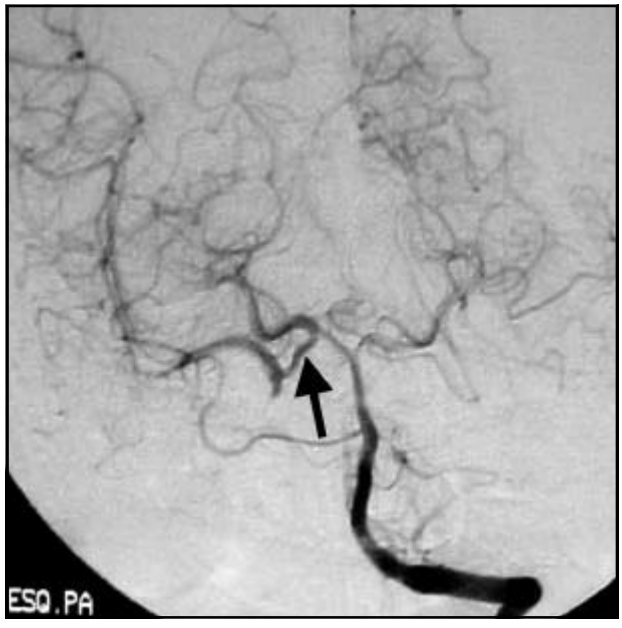

Fig 2. Postero-anterior view of angiography of left vertebral artery showing hypoplasic right P1 - fetal variant (black arrow).

tations of internal carotid artery (ICA) dissection, occuring in only 2.6% of patients[4]. Isolated oculomotor nerve deficit is rare[2,4]. Pupillary involvement is not a rule[4,9], but in some cases mydriasis may be the only manifestation for the few days preceding the complete oculomotor nerve palsy[10,11]. In other patients the pupil tends to be miotic, presumably because of the concurrent oculosympathetic palsy due to the pericarotid sympathetic plexus involvement[4,8]. This may explain the slight mydriasis observed in our patient.

The internal carotid artery is in proximity with oculomotor nerve only within the cavernous sinus. For this reason, mechanical compression or stretching of the third nerve would be a possible mechanism if the dissection of ICA extends into this site. However, according to some authors, another possible mechanism is the impairment of the blood supply to the third nerve[4,12]. An anatomical study of the extraneural blood supply to the intracranial oculomotor nerve showed that its proximal part receives nutrient arterioles mainly from thalamoperforating arteries arising from the posterior cerebral artery[13]. Small arteries arising from the basilar artery and posterior comunicating artery may also contribute[14]. The distal (intracavernous) portion of the oculomotor nerve is supplied mainly by arterioles originating from the inferior cavernous sinus artery and, to a lesser degree, by the meningohypophyseal trunk, both derived from the intracavernous internal carotid artery, which may be prone to embolic, hemodynamic or both mechanisms of ischemia[13].

Our patient had a fetal pattern of the right posterior cerebral artery (PCA). Like a previous patient described in the literature[10], this fact could explain the unusual susceptibility of the oculomotor nerve to ischemia due the compromised blood flow of small arteries arising from PCA and from the posterior communicating artery. We believe that in our patient the simultaneous onset of oculomotor and pupillary disfunction evidenced by acute diplopia and right blurred vision sensation must be probably related to a hemodynamic event that interrupted the blood flow to the arterioles supplying the proximal portion of the third nerve where oculomotor and pupillary fibers run together. Another possible mechanism to explain the third nerve palsy could be the known occurrence of microembolisms in these small arteries[15].

Our patient presented Chagas' disease. In theory, a thromboembolic event due to Chagas' cardiomyopathy could be the source of the oculomotor nerve ischemic lesion. Cardiac embolism as an etiologic factor in this patient has been ruled out by echocardiographical evaluation that disclosed only a mild contractile disfunction but no thrombus. Moreover, cervical angiography pointed to the diagnosis of carotid artery dissection.

In a patient with headache and complete oculomotor nerve palsy, the suspicion of a posterior communicating artery aneurysm is strongly suggested and angiographic evaluation is mandatory to confirm the diagnosis. In our patient, aneurysm was the first supposed diagnosis, but angiography did not show any intracranial aneurysm or other vascular lesion. Unexpectedly, a dissection of ICA was found.

Isolated oculomotor nerve palsy is underrecognized as a clinical presentation of extracranial ICA dissection. If the angiographic evaluation is incomplete and a careful study of extracranial arteries is not performed, the correct diagnosis may be missed or delayed. As early treatment with intravenous anticoagulation therapy may prevent thromboembolic complications related to cervical dissections[1], we draw attention to the importance of thorough angiographic study and careful evaluation of cervical arteries.

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