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Case Report

Persistent isolated mydriasis as an early sign of internal carotid artery dissection: Pourfour du petit syndrome

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ABSTRACT

Background: The dissection of the internal carotid artery (ICA) is commonly associated with miosis in Bernard-Horner syndrome (BHS). The presence of mydriasis is exceptional but can occur in the context of Pourfour du Petit syndrome (PDPS), a rare entity opposite of BHS accompanied by eyelid retraction and hyperhidrosis and caused by hyperactivity of the sympathetic cervical chain.
Aim: To report on a case of PDPS as the first manifestation of an ICA dissection.
Method: A 54-year-old man presented with isolated left mydriasis with no other abnormalities in the examination. Six months later, he suffered an ischemic stroke in the left middle cerebral artery territory secondary to a left ICA dissection.
Results: The initial study with Intracranial computed tomographic angiography and brain magnetic resonance imaging ruled out compressive cause of the third cranial nerve or structural lesion in the midbrain. The absence of hypersensitivity to Pilocarpine discarded postganglionic parasympathetic involvement.
Conclusions: In the presence of unilateral mydriasis and once common causes are ruled out an imaging examination of the supra-aortic trunks should be completed, since it could represent the first sign of carotid pathology in the context of PDPS.

1. Introduction

The dissection of the internal carotid artery (ICA) accounts for up to 20% of strokes in young patients and is frequently associated to head or neck pain, cerebral ischemia or Bernard-Horner syndrome (BHS) presenting with miosis, ptosis and enophthalmos. Pourfour du Petit syndrome (PDPS) is a rare entity that presents as a reverse BHS, with mydriasis, eyelid retraction and hyperhidrosis. Like BHS, it is caused by injuries in the sympathetic cervical chain [1]. We report on the case of a patient with PDPS that presented with unilateral persistent isolated mydriasis six months prior to an ischemic stroke secondary to a spontaneous ICA dissection. Previous literature on the topic is reviewed.

2. Case report

A 54-year-old man had sudden painless blurred vision. He reported no current or previous history of headache, neck pain or trauma. On the examination (Fig. 1), the only sign was anisocoria, with left pupil larger than right and unresponsive to light (Fig. 1A and B). The visual acuity was 20/20 and 20/25 on the right and the left eye respectively. Pupil diameters were 2 and 4 mm with photopic conditions in the right and left eye respectively; and 4 mm in both eyes with scotopic conditions. Eye movements were normal, and there was no evidence of ptosis, eyelid retraction or facial hyperhidrosis. Pharmacological ocular tests showed a negative response to Pilocarpine 0.125% but a significant contraction of the mydriatic pupil to the administration of Pilocarpine at 1% (Fig. 1C), indicating either sympathetic overactivity or preganglionar parasympathetic underactivity. Due to the main suspicion of non-reactive mydriasis as an initial symptom of a compressive III cranial nerve palsy, an intracranial computed tomographic angiography (CTA) at the level of the Circle of Willis was performed in order to rule out a posterior communicating artery aneurysm. No imaging of the supra-aortic trunks, including cervical carotid artery, was obtained at that moment. Besides, a brain magnetic resonance imaging (MRI) was

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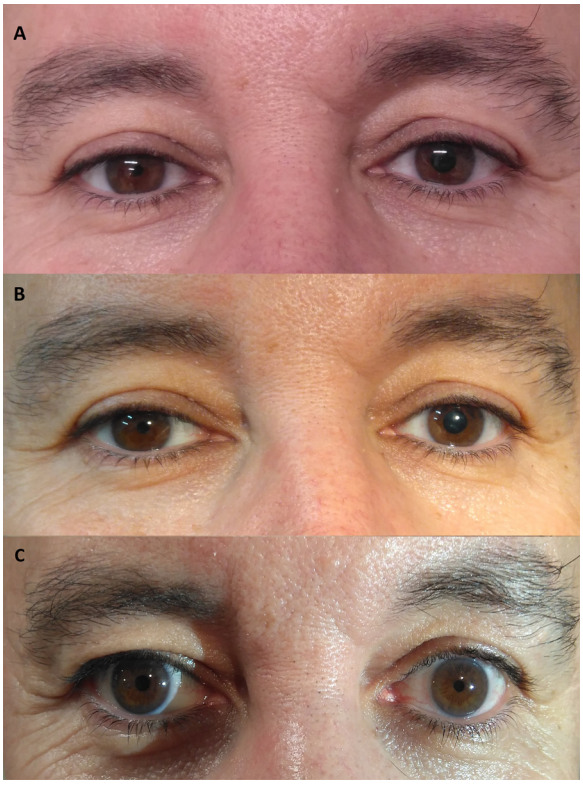

Fig. 1. Pupillary findings: Anisocoria due to left mydriasis in ambient light (1A). The dilated pupil is not reactive to light (1B). Post 1% pilocarpine shows a left pupil contraction (1C).

done in order to assess the upper brainstem and basal cisterns. Both examinations showed no significant findings. Antiplatelet therapy was added. On a follow-up visit four months later, the left mydriasis persisted without any change.

After 6 months, the patient suffered a sudden onset right hemiplegia, aphasia and right homonymous hemianopsia. Cranial computed tomography showed a hyperacute ischemic stroke involving the left middle cerebral artery (MCA) territory. The CTA showed a dissection of the left internal carotid artery (ICA) on its extracranial portion (Fig. 2) and a distal thrombus placed at the proximal segment of the ipsilateral MCA. Treatment consisted on intravenous fibrinolysis, mechanical thrombectomy and carotid stenting. The clinical evolution was good. Left eye mydriasis persisted on the last examination 9 months from onset.

3. Discussion

PDPS is a somewhat unknown entity also known as reverse Horner syndrome. This term was coined for the first time by Biffi in 1846 in honor of Pourfour du Petit, a French military surgeon [1]. The syndrome is characterized by unilateral mydriasis, eyelid retraction, and abnormal sweating response with hyperhidrosis. However, the clinical manifestations vary from cases with complete PDPS to others with isolated mydriasis or hyperhidrosis. The clinical course is variable, from transitory self-limited cases to permanent ones. The exact mechanism is not clear, but it is believed to be due to a sympathetic hyperstimulation caused by an irritation of the cervical sympathetic chain [1].

Various pathologies affecting the cervical sympathetic pathway have been associated with this syndrome: Nonpenetrating injuries of the cervical spine, traumatic pseudo-aneurysm of the internal carotid artery, cervical and thoracic tumors, and iatrogenic causes, such as complications after central venous catheterization or positional post-surgical side-effect.

There is a close relationship between PDPS and BHS, indeed the same etiologies can produce any of both conditions. It is believed that PDPS would be the clinical pattern whenever the insult irritates the sympathetic fibers, while BHS would be associated to deficitary states. It is however not possible to predict which phenomenon will occur after a certain damage.

Although the causes of a unilateral mydriasis are multiple, including compressive or ischemic neuropathy of the III cranial nerve and migraine, only few cases of isolated mydriasis as the first symptom of a carotid dissection have been reported [2]. The dissection of ICA is often associated to ocular pathology like amaurosis fugax and BHS. Mydriasis

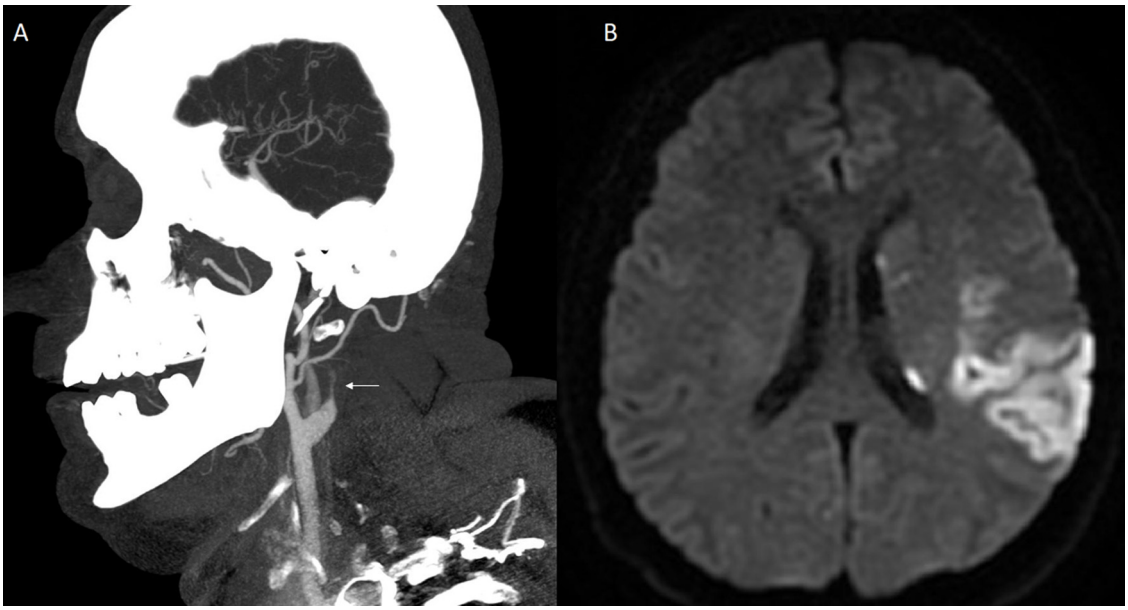

Fig. 2. Computed tomographic angiography performed in the Emergency Department shows a progressive narrowing of the eccentric lumen of the left internal carotid artery, surrounded by a crescent-shaped mural thrombus, with complete distal obstruction. This is known as the “string sign” and suggests a dissection (arrow in 2A). Diffusion weighted imaging of the magnetic resonance imaging performed in the acute phase shows areas of restricted diffusion in the posterior territory of the left middle cerebral artery representing an acute stroke (2B).

is exceptional and attributed to either a sympathetic overactivation or a parsympathetic underactivation. Rarely, a carotid pathology can produce a parasympathetic underactivation triggering a unilateral mydriasis. In some of these cases the mydriasis is usually followed by ophthalmoparesis, due to the damage of the third cranial nerve by hemodynamic mechanisms [3]. Also, a compressive mechanism due to compensatory hypertrophy of the posterior ipsilateral communicating artery in an ICA agenesia case has been reported [4]. Both causes were ruled out in our case given the normality of CTA and the absence of concomitant ophthalmoparesis. Other times, transient mydriasis is associated with a parasympathetic dysfunction secondary to ischemia in the ciliary ganglion, a structure irrigated by the ophthalmic artery through the ciliary arteries. ICA dissection is reported cause of ischemia of this structure [5]. This etiology was also discarded in our patient given the absence of hypersensitivity to low doses of pilocarpine (0.125%) on eyedrops testing which is characteristically observed in cases of a tonic pupil.

In our case, we propose that the spontaneous dissection of the internal carotid artery affected in the first term the cervical sympathetic pathway, causing the irritation of the oculo-sympathetic fibers. This was confirmed by the response to Pilocarpine at 1% but not at 0,125%. The long interval between the onset of the first symptoms of ICA dissection and the secondary stroke in this case is not such unusual since widely variable intervals ranging from hours to months have been reported in the literature. In our case, the stable degree of mydriasis, even after correct treatment with carotid stenting, could be explained by an aberrant regeneration of the oculosympathetic pathway, as suggested in previous literature.

4. Conclusions

Internal carotid artery dissection is a serious condition and it is therefore crucial to recognize its atypical neuro-ophthalmological manifestations such as the Pourfour du Petit Syndrome. Although the symptoms are opposed to those of Horner's syndrome, it has the same diagnostic value. Importantly, as shown here, it might precede in months the appearance of other symptoms due to ICA dissection. Then, in the presence of unilateral mydriasis, once ruled out commonest causes, CTA of the supra-aortic trunks should be performed.

Conflicts of interest

The authors declare that they have no conflict of interest.

Ethical standard

This study was performed in accordance with ethical standards, and informed consent was waived.

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