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Case Report

Internal Carotid Artery Dissection Presenting with Transient or Subclinical Horner Syndrome

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Keywords
Horner syndrome · Internal carotid artery dissection · Transient anisocoria · Transient eyelid ptosis

Abstract
Introduction: The most frequently encountered symptoms in internal carotid artery dissection (ICAD) are head or neck pain and cerebral ischemia. Ocular symptoms or signs have been reported as the presenting feature in up to 50% of patients, with (painful) Horner syndrome being the most frequently associated. Horner syndrome is part of the classic triad that depicts the characteristic presentation of ICAD and that consists of pain in the ipsilateral neck, head and orbital regions, (partial) Horner syndrome, and cerebral or retinal ischemia. All patients presenting with painful Horner syndrome should therefore require prompt investigations to rule out carotid artery dissection. In patients with confirmed diagnosis, treatment should be started early to prevent permanent ocular or cerebral complications. ***Case Presentation:*** Case 1: A 61-year-old woman presented with right temporal headache, an episode of transient visual loss and drooping of the right upper eyelid. Examination revealed anisocoria, which was more important in darkness. Reversal of anisocoria was observed after instilling drops of apraclonidine 0.5%. Neuroimaging demonstrated intrapetrous ICAD. Headaches, eyelid ptosis, and anisocoria all had resolved the next day. Apraclonidine pharmacologic testing a few weeks later was no longer dilating the previously smaller pupil. Case 2: A 48-year-old man presented with drooping of the right upper eyelid and right occipital headache and facial pain that all started one day after an intense yoga workout. Anisocoria was noticed upon examination, with topical cocaine 10% pharmacologic testing confirming a right Horner syndrome. Neuroimaging revealed ICAD. The patient reported resolution of his eyelid ptosis a few days later. Eyelid ptosis and anisocoria had indeed resolved at a follow-up examination a few weeks later. However, cocaine drop testing still produced anisocoria, compatible with subclinical Horner syndrome. ***Conclusion:*** Transient or

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subclinical Horner syndrome can be the presenting feature in ICAD; in such cases, the characteristic eyelid ptosis and anisocoria may be short-lived and resolve in only a few days. If suspected by clinical history, pharmacologic testing may be helpful in identifying subclinical cases.

Introduction

Horner syndrome is caused by an interruption of the sympathetic innervation to the eyes and is typically described as a clinical triad of unilateral ptosis, miosis, and anhidrosis. While painful Horner syndrome is the most common ocular sign of internal carotid artery dissection (ICAD) (reported in up to 58% of patients in large series) [1], only a handful of cases of transient or subclinical (positive pharmacologic test without clinical evidence) Horner syndrome have been reported in association with ICAD [2–4]. We hereby present two cases. The CARE Checklist has been completed by the authors for this case report, attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000535475).

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Case 1

A 61-year-old woman presented to the ophthalmology department with a 10-day history of right temporal headache. She had also experienced an episode of seeing positive visual phenomena consisting of zigzag lines starting in the left side of her visual field but then gradually expanding across the entire visual field, and this was accompanied by the appearance of a mosaic of colored spots. This episode lasted for about 15 min and occurred 2 days before presentation. In addition, she had noticed drooping of the right upper eyelid, starting a few hours afterward. Past medical history was positive for hemolysis, elevated liver enzymes, and low platelets (HELPP) syndrome, poorly controlled arterial hypertension, miscarriage, pyelonephritis, and hypokalemia. She was also known for migraine headaches but had not had a single episode for the last 15 years or any episode accompanied by visual aura. Furthermore, the characteristics of her migraine headaches in the past were different from the current headache, and contrary to her migraine episodes, she had no photophobia.

At presentation, best corrected visual acuity was 20/20 in both eyes. Slit lamp and eye fundus examination were unremarkable. However, a mild ptosis of the right upper eyelid was noticed, as well as a difference in pupillary size with a smaller pupil in the right eye (Fig. 1a). Anisocoria was more important in darkness. Eye movements were full and painless. Reversal of anisocoria and resolution of upper eyelid ptosis of the right eye were observed after instilling three drops of apraclonidine 0.5% in both eyes, suggestive of a right Horner syndrome (Fig. 1b). Same-day computed tomography angiography with specific attention to the supra-aortic and cerebral vessels revealed no apparent narrowing of the carotid artery lumen and was initially considered normal. Headaches, eyelid ptosis, and anisocoria were all found to be resolved during examination at the neurology department the next day, and a provisional diagnosis of migraine complicated with Horner syndrome was proposed by the neurologist. A repeat exam at the ophthalmology department the next week confirmed

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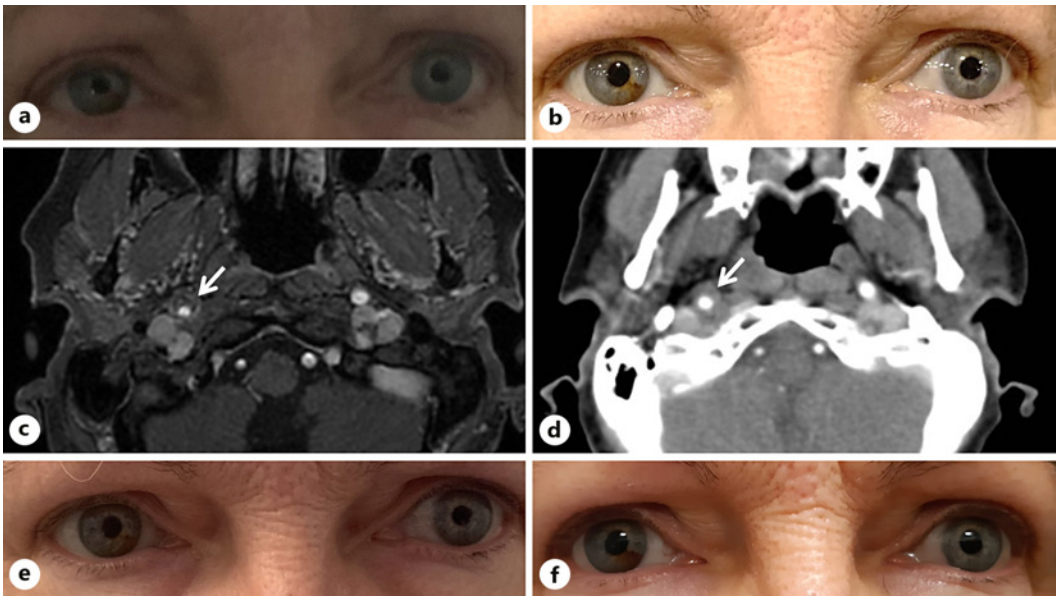

Fig. 1. Photographs of patient 1 at presentation demonstrating Horner syndrome of the right eye with miosis and ptosis of the upper eyelid (**a**), followed by reversal of anisocoria and resolution of upper eyelid ptosis after instillation of topical apraclonidine 0.5% (**b**). Cerebral magnetic resonance angiography (**c**) and computed tomography angiography (**d**) demonstrating dissection of the right intrapetrous internal carotid artery (white arrows). Photographs at follow-up showing no more evidence of clinical (**e**) or subclinical (**f**) (using apraclonidine 0.5% pharmacologic testing) Horner syndrome.

complete resolution of the right eye eyelid ptosis and miosis (Fig. 1e). However, magnetic resonance angiography was obtained the next day and showed a narrowed eccentric lumen surrounded by a crescent-shaped mural hematoma at the intrapetrous segment of the right internal carotid artery, compatible with dissection (Fig. 1c, arrow). Antiplatelet treatment with acetylsalicylic acid (80 mg daily) was started, and her antihypertensive treatment was intensified. Apraclonidine 0.5% pharmacologic testing was repeated 6 weeks later and did no longer unilaterally dilate the right pupil (Fig. 1f). Review of the initial CT scan did reveal mild narrowing of the lumen of the intrapetrous part of the right carotid artery, surrounded by a non-enhancing crescent-shaped thickening of the vessel wall (Fig. 1d, arrow). A repeat magnetic resonance angiography after 6 months of follow-up showed no signs of cerebral ischemic disease and demonstrated complete recanalization of the right internal carotid artery, after which antiplatelet treatment was discontinued.

Case 2

A 48-year-old man presented to the emergency department with a 2-day history of drooping of the right upper eyelid, right occipital headache, and right facial pain; all starting one day after a 3-hour-long intense yoga workout. Past medical history was positive for syphilis (treated 2 years before).

At presentation, drooping of the right upper eyelid was confirmed, as well as an inverse ptosis of the right lower eyelid and a difference in pupillary size (with a smaller pupil in the right eye) (Fig. 2a). As was the case in our other patient, anisocoria was more important in darkness. Best corrected visual acuity was 20/20 in both eyes. Slit lamp and eye fundus examination were unremarkable. Ocular motility was full and painless. Cocaine 10% pharmacologic testing showed asymmetric dilation of the pupils, suggestive of a right Horner syndrome (Fig. 2b). Urgent CT angiography was performed and was initially considered

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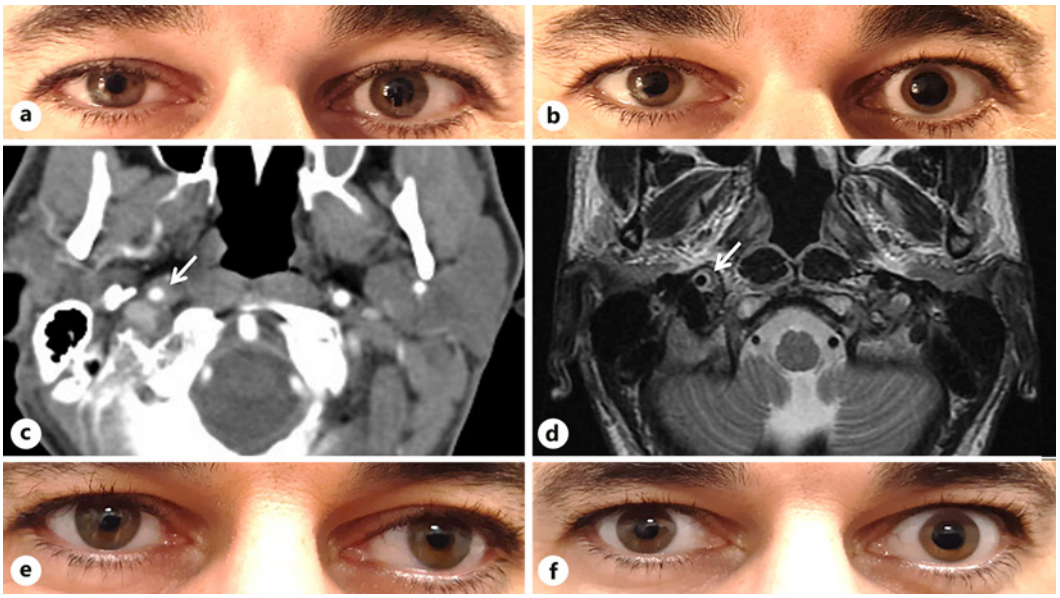

Fig. 2. Photographs of patient 2 at presentation demonstrating Horner syndrome of the right eye with miosis, ptosis of the upper eyelid, and inverse ptosis of the lower eyelid (**a**), followed by an asymmetric dilation of the pupils after instillation of topical cocaine 10% (**b**). Head computed tomography angiography (**c**) and MRI (**d**) demonstrating dissection of the right internal carotid artery at the cervical segment (white arrows). Photographs at follow-up showing no clinical signs of Horner syndrome (**e**), whereas cocaine 10% pharmacologic testing still produced anisocoria by dilating the uninvolved left pupil, compatible with a right subclinical Horner syndrome (**f**).

normal. A closer look, however, revealed mild narrowing of the lumen at the cervical segment of the right internal carotid artery, surrounded by a crescent-shaped thickening of the vessel wall (Fig. 2c, arrow), suggesting dissection. These findings were confirmed the next day by cerebral MRI (Fig. 2d, arrow). Head CT scan as well as MRI also demonstrated a small lacunar ischemic lesion located at the right thalamus. Antiplatelet treatment with acetylsalicylic acid (80 mg daily) was subsequently initiated. The patient reported resolution of his eyelid ptosis a few days after initial presentation. At a follow-up examination 3 weeks later, eyelid ptosis and inverse ptosis had indeed resolved (subtle preexisting dermatochalasis of the right upper eyelid was still present), as well as the anisocoria (Fig. 2e). However, cocaine 10% pharmacologic testing still produced anisocoria (only dilating the uninvolved left pupil) and was therefore compatible with a right subclinical Horner syndrome (Fig. 2f). Doppler ultrasound 2 months and 6 months after initial presentation showed no more signs of carotid dissection, and antiplatelet drugs were discontinued after a total of 6 months of treatment (during which the patient experienced no new symptoms).

Discussion

Ocular symptoms or signs are frequently associated with and are often the presenting features in ICAD. The classic triad consists of pain in the ipsilateral neck, head and orbital regions, (partial) Horner syndrome, and cerebral or retinal ischemia. Patients presenting with painful Horner syndrome or transient monocular visual loss should therefore require prompt investigations to rule out carotid artery dissection. In patients with confirmed diagnosis, treatment should be started early to prevent permanent ocular or cerebral complications [1].

Transient or subclinical (positive pharmacologic test without clinical evidence) Horner syndrome has been reported in association with ICAD [2–4]. None of the published cases both had clinical and pharmacologic confirmation of Horner syndrome at presentation and early (weeks rather than months) repeat pharmacologic tests (in search of a subclinical syndrome) after complete resolution of the clinically visible Horner syndrome [2–4]. Similar to our two cases, clinical presence of anisocoria and eyelid ptosis can be as short-lived as a few days [3, 4]. Horner syndrome in ICAD is presumed to be the result of ischemia (through the vasa nervorum) or compression or stretching (by the enlarged occluded carotid) of the pericarotid sympathetic fibers [1]. Its transient nature in our patients might therefore potentially be explained by relief of the compression/stretching effect on the arterial wall after the formation of a more solid intramural hematoma.

A case of transient Horner syndrome associated with intrapetrous carotid dissection developing during a migraine attack has already been reported in a patient with a known migraine history [4]. Contrary to our first case, the migraine attack associated with the Horner syndrome in this patient did not differ from the usual ones. The authors hypothesized that in migrainous patients some predisposing facilitating factors may exist, causing spontaneous dissections to occur even in relationship to trivial stimulation (such as the coughing during a pharyngitis in their patient) [4]. In our first patient, the dissection itself could have been precipitated by important fluctuations in blood pressure (with values as high as 200/130 mm Hg around the time of presentation) [5]. Repeated hyperextension, hyperflexion, and/or forceful rotation of the neck during an intense yoga workout could have been the causative mechanism in our second patient [6, 7].

Similar to our first patient, transient visual loss in ICAD can indeed mimic migraine visual aura with positive visual phenomena such as "scintillations," "flashing lights," "bright spots," or "scintillating scotoma." These symptoms are typically limited to the side of the dissection [1], although binocular cases have been reported [8]. They can often be evoked by postural changes (for example, trying to sit up) or by exposure to bright lights [1].

Initial radiologic interpretation of the CT angiography in both patients was mainly focused on volume-rendered 3D reconstruction images and revealed no apparent narrowing of the carotid arteries. However, the association of Horner syndrome and orbital and/or ipsilateral head or neck pain of acute onset is so characteristic that ICAD should be considered until proven otherwise [1]. Indeed, depending on the tissue space into which the blood dissects, there can be no narrowing of the lumen so that Doppler and angiographic studies may be interpreted as normal, causing diagnosis to be missed [1, 9]. Hemorrhage into the subintimal space of the vessel often does cause narrowing of the vessel lumen, whereas hemorrhage into the subadventitial layer may produce a pseudoaneurysm that does not narrow the lumen and does not produce stenosis. A crescent-shaped mass in the vessel wall may be identified with MRI in both subintimal and subadventitial hemorrhage [1, 9].

Patients usually have resolution and healing of the blood vessel on follow-up imaging 6 months after diagnosis, and radiological changes are seldom observed beyond 1 year [10, 11]. Although the dissected blood vessel can recanalize completely, residual stenosis, occlusions, or aneurysms may also persist [10, 11]. Currently, there is no consensus on optimal treatment. As outlined before, the main purpose of treatment is to prevent stroke. The most likely mechanism for stroke in patients is thought to be artery-to-artery embolism from an intraluminal thrombus or from fragments of a thrombus that forms in the false lumen [11, 12]. Therefore, in patients with ischemic stroke or transient ischemic attack after an extracranial carotid or vertebral arterial dissection, guidelines from the American Heart Association (AHA)/American Stroke Association (ASA) recommend treatment with antithrombotic therapy for at least 3 months [12]. These guidelines also

state that the relative efficacy of anticoagulation versus antiplatelet therapy is unknown, but current data seem to suggest no significant differences in outcome events or the rate of recanalization when comparing antiplatelet therapy with anticoagulation [11–13]. Endovascular procedures may be considered in patients refractory to medical treatment (patients who have recurrent ischemic events despite antithrombotic therapy) and to treat complications such as pseudoaneurysms [12].

Conclusion

Transient or subclinical Horner syndrome can be the presenting feature of ICAD. While straightforward when clinically evident, the characteristic eyelid ptosis and anisocoria may be short-lived and resolve in only a few days. If suspected by clinical history, pharmacologic testing may still be helpful to identify subclinical cases. Even then, identifying the underlying lesion may sometimes be a radiological challenge. Regardless, all patients should be recognized early to prevent permanent ocular or cerebral complications.

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Statement of Ethics

Written informed consent was obtained from the patients for publication of this case report and any accompanying images. Approval by the Ethics Committee of the CHU Saint-Pierre hospital in Brussels was acquired on December 7, 2021 (reference number CE/21-12-05).

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

T.B. and L.S. wrote the manuscript, conducted the relevant literature search associated with it and were involved in the diagnostic process and clinical follow-up of patient 1, while T.B., J.S., and M.O.H. were involved in the diagnostic process and clinical follow-up of patient 2. S.K. offered expert opinion. F.W. is the senior author who advised and oversaw this case and manuscript. All authors read and approved the final manuscript.

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

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