Horner syndrome with transient visual impairment

Frank A. Orlando, Maria Elisa Lupi

Department of Community Health and Family Medicine, University of Florida, Gainesville, FL, United States

ABSTRACT

A 57-year-old female presented with headache, miosis, and ptosis diagnosed as Horner syndrome (HS). After delaying the recommended diagnostic imaging, she experienced transient, unilateral visual impairment in bright light. The patient was subsequently determined to have a spontaneous internal carotid artery dissection (ICAD) and secondary retinal ischemia with minimal cardiovascular risk factors and no history of preceding trauma. She wore dark glasses, received gabapentin for pain control, and was anticoagulated for a total of 4 months at which time the ICAD resolved despite a residual blepharoptosis and anisocoria.

Keywords: Amaurosis fugax, anisocoria, blepharoptosis, blindness, facial pain, headache, Horner's syndrome, Horner, Horners, internal carotid artery dissection, miosis, neck pain, ptosis, transient monocular vision loss

Introduction

HS is divided into pre-ganglionic (1\(^{st}\) and 2\(^{nd}\) order) and post-ganglionic (3\(^{rd}\) order) based on lesion location, and primary care physicians must understand the various presentations to narrow the differential diagnosis and guide study selection. 1\(^{st}\) and 2\(^{nd}\) order lesions have the classic triad: unilateral miosis, ptosis, and anhidrosis. 3\(^{rd}\) order lesions, presenting without anhidrosis, require expedited neuroimaging given the severe underlying etiologies: ICAD and cavernous sinus thrombosis. Other 3\(^{rd}\) order lesions include neck/nasopharyngeal masses and benign causes like otitis media and cluster headaches. We present a case of acute HS with neck pain and visual changes requiring ICAD workup.

Case Report

A 57-year-old, non-smoking female with hyperlipidemia presented to clinic with complaints of a gradually worsening, intense right-sided head pressure and eye swelling for 1 week associated with malaise, sore throat, nasal congestion, and nausea. On exam, right-sided findings included blepharoptosis [Figure 1], conjunctival injection, nasal congestion, neck tenderness without lymphadenopathy or bruit, and a miotic pupil that failed to dilate under dim lighting [Figure 2]. The neurologic and cardiopulmonary exams were otherwise normal.

She delayed STAT imaging insisting on a sinus infection and presented to the ER four days later dizzy, with tongue numbness, and seeing a “blue light” from her right eye. MRI/MRA and CTA showed right ICAD with high grade stenosis (at least 90%) entering the petrous bone [Figure 3]. She was admitted on enoxaparin and warfarin for anticoagulation, atorvastatin for stroke prevention, and gabapentin for pain. Exposure to bright light continued to cause a transient visual impairment, and she needed sunglasses indoors. Enoxaparin was stopped post-discharge after two therapeutic INRs, and warfarin was continued for 4 months when follow-up CTA showed ICAD resolution. She was then transitioned to ASA. Now 6 years later, the blepharoptosis and anisocoria have remained chronic.

Discussion

HS is a rare neurologic condition, and this case highlights the importance of primary care physicians making a timely clinical

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: WKHLRPMedknow_reprints@wolterskluwer.com

Received: 14-07-2020 Accepted: 02-10-2020
Revised: 17-09-2020 Published: 31-12-2020

How to cite this article: Orlando FA, Lupi ME. Horner syndrome with transient visual impairment. J Family Med Prim Care 2020;9:6273-5.
In addition to causing HS by injuring the adjacent sympathetic plexus, an ICAD can compress local cranial nerves via pseudo-aneurysm 8-16% of the time, particularly IX-XII and infrequently V and VII. Cavernous sinus thrombosis also causes a 3rd order HS but with ophthalmoplegia, particularly cranial nerve VI with no other brainstem signs. An MRI cavernous sinus should be performed in these patients.

Even though patients with carotid artery dissection may have a subtle history of antecedent neck trauma, it can also occur spontaneously, especially in those with hypertension, smoking, on oral contraception, or with a connective tissue disorder. Furthermore, the majority of carotid artery dissections are idiopathic. Therefore, when acute HS presents with symptoms of ICAD but no recent trauma and a lower than expected cardiovascular risk, it is still critical for primary care physicians to follow through with STAT imaging, even if the patient is hesitant, hoping for a benign cause. In this case, MRI/MRA with fat suppression is the best initial screening test and is non-invasive, although digital subtraction angiography is still considered the gold standard. Axial, T1-weighted, pre-contrast, fat-suppressed MRI is necessary to diagnose ICAD when the vessel lumen is not narrowed, making the dissection undetectable on CTA and MRA. Importantly, pharmacological testing by an ophthalmologist is generally only useful for isolated anisocoria, otherwise it could delay diagnosis and treatment. Stroke is a major risk in ICAD patients for two weeks following HS onset. Patients with high-grade ICA stenosis or ICA occlusion rarely present with an episodic vision impairment-related exclusively to light exposure, a symptom related to transient retinal ischemia (retinal claudication).

The differential diagnosis of headache and transient visual impairment also includes amaurosis fugax, a transient monocular vision loss (TMVL) or binocular blindness lasting seconds or minutes. TMVL has a diverse etiology, but some use amaurosis fugax to exclusively describe its most common vascular causes, retinal ischemia from ICA stenosis or embolism, which have a 2-3% per year stroke risk. In such cases, the TMVL is classically described as descending over the field of vision “like a curtain or shade,” or less commonly ascending from below, but can also be shaded, black, or blurred vision.

Retinal artery stroke is among a group of ocular arterial occlusive disorders that cause a permanent monocular blindness rather than HS. Ocular arterial occlusive disorders include central or branch retinal artery occlusion and ocular ischemic syndrome, each of which has a cerebral stroke risk that is significantly increased independent of internal carotid stenosis. Some feel the terms “ocular migraine” and “retinal migraine” are oxymorons that should not be used in the medical vernacular because migraines causing binocular vision loss are a cortical process while vasospasm causing TMVL is a retinal process. Moreover, migraines classically produce positive visual phenomena such as scintillations rather than visual loss without scintillations.
although TIA can also produce positive visual phenomena.[14]
While migraine alone will not produce HS, cluster headaches can cause HS and have a similar presentation to ICA.[15]

We were granted institutional permission to do clinical research and obtained informed consent to take and publish this patient's photographs. The patient's miosis was difficult to reproduce in a photograph until we reverse lit the eye in a totally dark room before an infrared light source; then the anisocoria showed up dramatically [Figure 2]. Because of this process, this photo must be black and white, and there is a reflection on her nose from a camera artifact. This artifact could not be eliminated without significant cropping, even when we consulted an expert professional photographer.

Early recognition of HS remains challenging for primary care physicians in the outpatient setting. A clinical diagnosis of HS obligates quick neuroimaging to confirm the etiology followed by prompt treatment to prevent chronic stroke sequelae and death.

Acknowledgement
We would like to thank photographer Harry Rosa, Ariel Pomputius, Emma Djabali, Marie Fucci, Sandy Campbell, and Drs. Sonal Tuli, Eric Grieser, and Carlos Diez-Freire for their support.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

References
1. CADISS trial investigators, Markus HS, Hayter E, Levi C, Feldman A, Venables G, Norris J. Antiplatelet treatment compared with anticoagulation treatment for cervical artery dissection (CADISS): A randomised trial. Lancet Neurol. 2015;14:361-7. doi: 10.1016/S1474-4422(15)70018-9.
2. Milhaud D, de Freitas GR, van Melle G, Bogousslavsky J. Occlusion due to carotid artery dissection: A more severe disease than previously suggested. Arch Neurol 2002;59:557-61.
3. Hakimi R, Sikavummar S. Imaging of carotid dissection. Curr Pain Headache Rep 2019;23:2.
4. Majeed A, Ribeiro NPL, Ali A, Hijazi M, Farook H. A rare presentation of spontaneous internal carotid artery dissection with Horner’s syndrome, VIIth, Xth and XIIth nerve palsies. Oxf Med Case Rep 2016;2016:255-8.
5. van der Zwet KVM, Brown AV, Bakker SLM. Internal carotid artery dissection presenting with dysgeusia, Horner syndrome, and hypesthesia of the fifth cranial nerve: A case report. J Emerg Med 2020;58:e27-9.
6. Micheli S, Paciaroni M, Corea F, Agnelli G, Zampolini M, Caso V. Cervical artery dissection: Emerging risk factors. Open Neuro J 2010;4:50-5.
7. Mansukhani SA, Eckel LJ, Wu KY, Hassan MB, Van Loon JA, Chen JJ, et al. Horner syndrome due to internal carotid artery dissection with normal vascular imaging: A radiological conundrum. J Neuro-Ophthalmol 2020:1-3. [in press]. doi: 10.1097/WNO.0000000000000981.
8. Morris NA, Merkler AE, Gialdini G, Kamel H. Timing of incident stroke risk following cervical artery dissection presenting without ischemia. Stroke 2017;48:551-5.
9. Furlan AJ, Whisnant JP, Kearns TP. Unilateral visual loss in bright light: An unusual symptom of carotid artery occlusive disease. Arch Neurol 1979;36:675-6.
10. Donders RC, Dutch TMB Study Group. Clinical features of transient monocular blindness and the likelihood of atherosclerotic lesions of the internal carotid artery. J Neurol Neurosurg Psychiatry 2001;71:247-9.
11. Tadi P, Najem K, Margolin E. Amaurosis Fugax. StatPearls [internet] Treasure Island (FL): StatPearls Publishing; 2019.
12. Aver MB, Magal I, Kherani A, Mitha AP. Risk of stroke in patients with ocular arterial occlusive disorders: A retrospective Canadian study. JAHA 2019;8:1-7.
13. Pula JH, Kwan K, Yuen CA, Kattah JC. Update on the evaluation of transient vision loss. Clin Ophthalmol 2016;10:297-303.
14. Winterkorn JM. Retinal migraine is an oxymoron. J Neuroophthalmol 2007;27:1-2.
15. Elhnawy AM, Solymosi L, Sommer C. Carotid dissection presenting as a prolonged cluster-like headache in a patient with episodic cluster headache. BMJ Case Rep 2017;2017:bcr2017220845. doi: 10.1136/bcr-2017-220845.