Case Report

Painful Horner Syndrome as a Harbinger of Silent Carotid Dissection

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PRESENTATION of CASE

A 43-y-old white female presented to the hospital in July 2004 with pain in the left eye and left upper lid ptosis. She did not perceive any difference in perspiration between the two halves of her face. She was a nonsmoker and denied any history of head or neck trauma, or ocular, cardiac, vascular, or neurologic disease. Neuro-ophthalmological examination was normal except for 1 mm of left upper eyelid ptosis (drooping of the eyelid), miosis (constriction of the pupil), and mild enophthalmos (recession of the eyeball into the orbit) consistent with classic left-sided Horner syndrome (Figure 1). There was no carotidynia (a neck pain syndrome associated with tenderness to palpation over the carotid bifurcation) or carotid bruit. A chest radiograph obtained to rule out an underlying left apical superior sulcus tumor was normal. Magnetic resonance imaging/magnetic resonance angiography of the brain with cross-sectional imaging of the neck was obtained, which revealed extracranial left internal carotid artery dissection (Figures 2 and 3). The patient was treated with unfractionated heparin and coumadin and made an uneventful recovery. The patient was seen in the clinic a few months later and did not have any complications at follow-up.

DISCUSSION

Horner syndrome—characterized by the constellation of miosis, ptosis, anhidrosis (lack of sweating), enophthalmos, and anisocoria (unequal pupil size)—is present in up to 58% of internal carotid artery dissections [1]. Most patients experience neck, facial, and head pain ipsilateral to the lesion because of ischemia or stretching of the trigeminal pain fibers surrounding the carotid arteries [2]. Ophthalmic manifestations have been reported to occur in up to 62% of patients with internal carotid artery dissection [2]. Common findings in descending order of frequency are painful partial Horner syndrome (due to disruption of the third-order neuron oculosympathetic fibers) as seen in our patient, transient monocular vision loss, and permanent visual loss [2].

De Bray et al. studied the prognosis of 90 cases of isolated Horner syndrome due to internal carotid artery dissection [3]. They found that 91% of cases of Horner syndrome due to internal carotid artery dissection were painful. The risk of an early ischemic stroke within the first 2 wk was high (around 17%) without initial antithrombotic treatment [3].

Internal carotid artery dissection is a potentially life-threatening condition and carries a substantial risk of disabling stroke [4]. Carotid dissection is under-recognized as a cause of Horner syndrome and can be missed [5]. It is important to diagnose dissection because anticoagulation can prevent carotid thrombosis and embolism [5]. The investigation of choice is magnetic resonance imaging and angiography scan of the head and neck [5]. The treatment advocated for dissection is anticoagulation for 3–6 mo [5].
References

1. Bougousslavsky J, Despland PA, Regli F (1987) Spontaneous carotid dissection with acute stroke. Arch Neurol 44: 479–482.
2. Biousse V, Touboul PJ, D’Anglejan-Chatillon J, Levy C, Schaison M, et al. (1998) Ophthalmologic manifestations of internal carotid artery dissection. Am J Ophthalmol 126: 565–577.
3. de Brav J, Baumgartner RB, Guillon B, Dziewas R, Ringelstein E, et al. (2004) Prognosis of dissections with an initially isolated Horner’s syndrome [abstract]. 13th European Stroke Conference; Mannheim-Heidelberg, Germany; 2004 May 14. Available: http://www.esc-mannheim-2004.org/ Mannheim/m__84_oral.asp. Accessed 6 December 2004.
4. Dziewas R, Konrad C, Drager B, Evers S, Besselmann M, et al. (2003) Cervical artery dissection—Clinical features, risk factors, therapy and outcome in 126 patients. J Neurol 250: 1179–1184.
5. Chan CC, Paine M, O’Day J (2001) Carotid dissection: A common cause of Horner’s syndrome. Postgrad Med J 80: 104.

Learning Points

- Painful Horner syndrome should alert clinicians to the possibility of a silent carotid dissection until proven otherwise [6].
- Magnetic resonance imaging and angiography scan of the head and neck is the imaging modality of choice to look for dissection [5].
- For patients with carotid dissection, anticoagulation with warfarin and coumadin is recommended for 3–6 mo to prevent carotid thrombosis and embolism [5].