

BACKGROUND

- Horner syndrome is characterized by the classic triad of ptosis, miosis, and anhidrosis (1)
- The “Harlequin sign” is a distinct hemifacial flushing that can occur with ocular findings (such as Horner syndrome) or without (referred to as “Harlequin syndrome”) (2)
- Most cases of the Harlequin sign also involve abnormalities of the pupil (3)

PURPOSE

- To report a case of Horner syndrome with a positive Harlequin sign in an adolescent male due to a presumed traumatic cause

SETTING

- Initial work-up occurred in the emergency department of a Saskatchewan tertiary-care center. Follow-up occurred at a community ophthalmology clinic.

METHODS

1. Case report including a review of the patient’s clinical record (history of presenting illness, clinical examinations, investigations, and imaging)
2. Review of PubMed articles describing Horner syndrome and the Harlequin sign

CASE REPORT

History of Presenting Illness: An 11-year-old boy presented to the emergency department with a history of uneven facial flushing. The boy’s parents described the left side of his face becoming flushed with exercise while the right side remained pale.

Clinical Exam: Right-sided ptosis & miosis, with pupils measuring 5.0 mm on the right and 8.0 mm on the left (Figure 1). Pupils otherwise round, reactive, and without afferent pupillary defects. Anhidrosis was suspected on the right side but was not formally assessed. The boy had 20/25 corrected vision in both eyes, complete extraocular movements, and full confrontational fields. Anterior and posterior segment examinations were normal. General examination revealed normal tone, reflexes, and cranial nerve function.

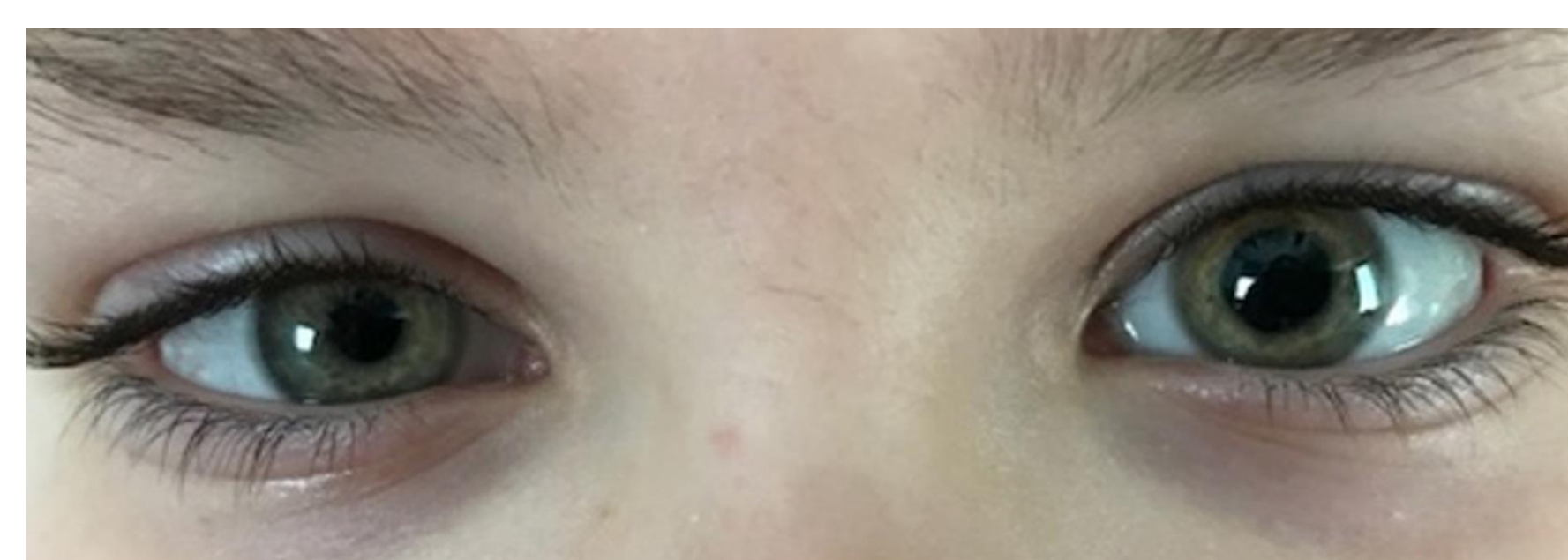


Figure 1: Right-sided ptosis and miosis of Horner syndrome.



Figure 2: Photograph of the patient after exercise with the hemifacial flushing of the Harlequin sign.

Follow-up: At four-years, the family reports that the ptosis and flushing has resolved, and only mild anisocoria remains.

Lab Investigations: Unremarkable

Neuroimaging: MRI and MRA, of the brain, neck, and upper chest, with and without contrast were unremarkable

Collateral: A photograph taken by the parents before presenting to the emergency department showed distinct hemifacial flushing (Figure 2).

Diagnosis: Horner syndrome with a positive Harlequin sign.

Etiology: Remains unclear. The parents recall a recent history of right shoulder injuries at a trampoline park and in a snowmobile accident, suggesting a possible traumatic cause.

DISCUSSION

- Horner syndrome arises due to disruption of the sympathetic chain
- The Harlequin sign arises due to sympathetic dysfunction that prevents thermoregulatory vasodilation on the denervated side
- The normal side exhibits a compensatory flushing reaction to the lack of sudomotor and vasomotor function on the denervated side
- In our case, we suspect a lesion on the right side, correlating to a right-sided ptosis and miosis, with a contralateral flushing reaction on the left side.

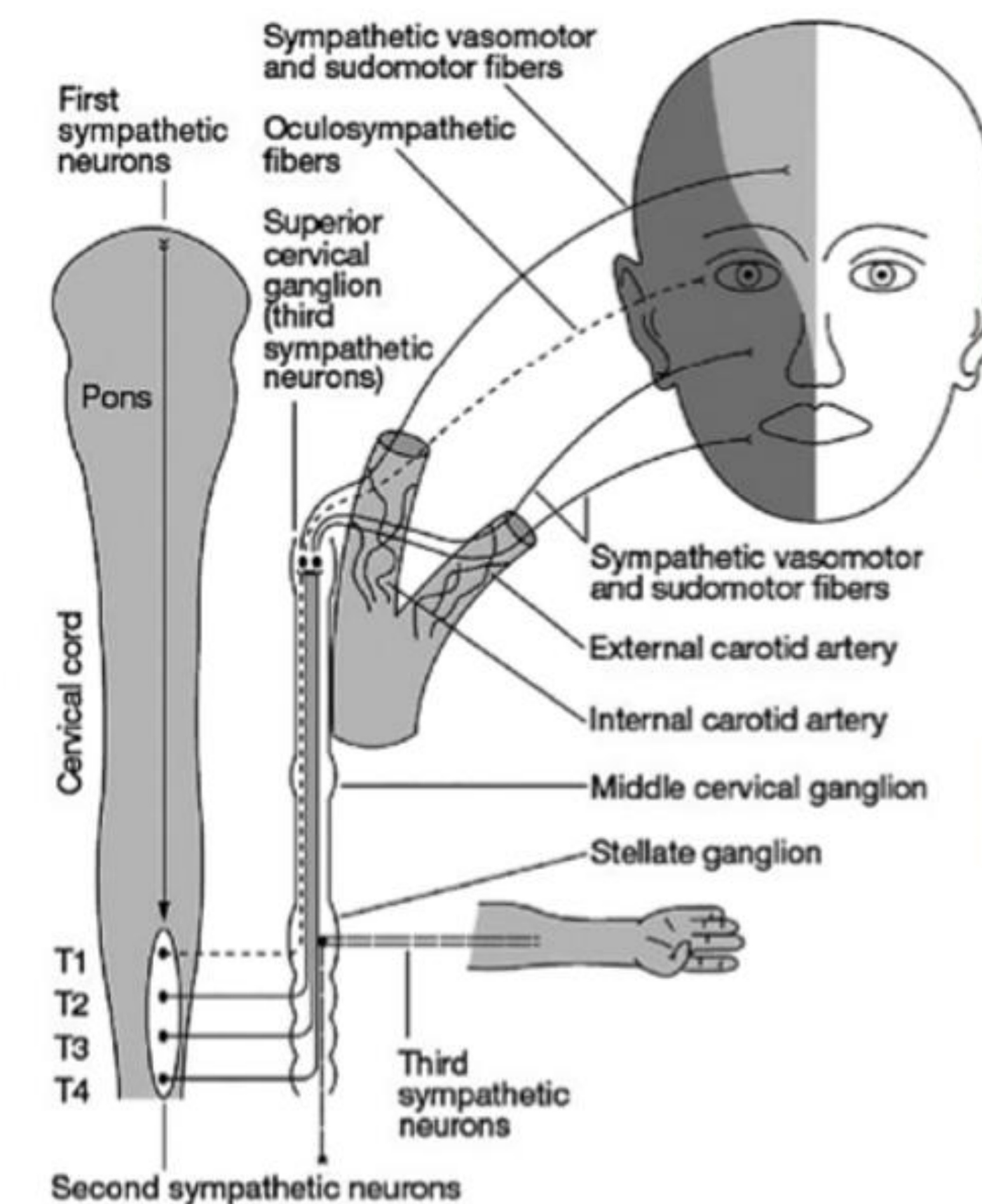


Figure 3: Oculo-sympathetic pathway and possible lesion locations

REFERENCES

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2. Miquel J, Piyaraly S, Dupuy A, Cochat P, Phan A. Congenital Cases of Concomitant Harlequin and Horner Syndromes. *J Pediatr.* 2017; 182:389–92.
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