Teaching NeuroImages:

Multiple clinical manifestations of a ganglionic sympathetic defect

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Figure 1 Episodic deformation of left pupil

(A) Left pupillary deformation known as tadpole pupil. This is presumably sympathetic overactivity causing segmental spasm of the iris dilator. Some tadpole pupils evolve to permanent Horner syndrome (sympathetic deficit). (B) Absence of facial flushing and sweating on the left side during sport activities indicating sudomotor fiber dysfunction.

A 39-year-old woman with longstanding asymmetric facial flushing noticed episodic deformation of her left pupil (figure 1), which 2 months later became miotic. Despite no ptosis, a left Horner syndrome was pharmacologically confirmed (figure 2). She also had left cheek pain, present only at mealtime. Radiologic investigations were normal.

This case illustrates many manifestations of a unilateral sympathetic deficit to the head and eye including harlequin sign,¹ tadpole pupil, Horner syndrome, and masticatory pain from parasympathetic overactivation of secretory myoepithelial cells, typically upon the first bite.² The site of dysfunction localizes to the superior cervical ganglion; the cause remains unknown.

AUTHOR CONTRIBUTIONS

Myriam Ladaïque: study concept and design, acquisition of data, analysis and interpretation of data. Aki Kawasaki: study concept

and design, acquisition of data, analysis and interpretation of data, study supervision, critical revision of manuscript for intellectual content.

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Figure 2 Left Horner syndrome pharmacologically confirmed

(A) Baseline anisocoria (pre-apraclonidine) in room light. (B) After instillation of 1 drop of topical apraclonidine in each eye, the miotic left pupil became mydriatic; the right pupil remained unchanged. This reversal of anisocoria is due to adrenergic denervation hypersensitivity of the left iris dilator and confirms a Horner syndrome.

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