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Resident and Fellows

Harlequin Syndrome in Cluster Headache

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A 63-year-old male experienced 3 years of excruciating, right retro-orbital, 15–90 minute headaches occurring 1 to 3 times daily associated with ipsilateral lacrimation, ptosis, and facial congestion. The episodes began after a complicated right root canal procedure. He also reported separate episodes of distinct left sided facial flushing triggered by exertion as evidenced by a photo that he provided (see Fig. 1) that clearly demonstrates the distinct line of demarcation down the middle of his forehead. On exam, he had right sided miosis (only in darkness), ptosis, tongue deviation, and head tilt. No pharmacologic pupil function tests were performed. MRI brain with and without contrast and CTA head/neck were negative. Cluster headache was diagnosed and verapamil significantly improved headaches. The patient was also diagnosed with Harlequin syndrome, a rare and striking phenomenon of sudden-onset hemifacial or hemibody flushing in responses to heat, exertion, or emotion.

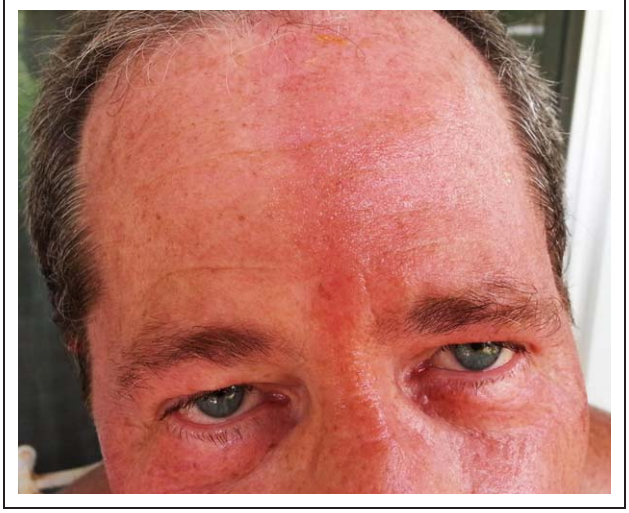

Fig. 1.—Unilateral facial flushing in Harlequin syndrome.

Typically, the unilateral increased discoloration focuses the attention on that side as abnormal; however, the underlying sympathetic neural injury occurs on the non-flushed side of the face.[1] Ipsilateral sympathetic fiber injury can be the underlying etiology due to brainstem infarction, thyroid disease, internal carotid artery dissection or aneurysm and perineural spread of a malignant skin tumor; appropriate testing is important to rule these conditions out.[2,3]

The strictly LEFT-sided facial flushing that occurred with exertion in this patient demonstrates

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the patient's loss of sweating on the RIGHT, non-flushed side of the face, suggestive of sympathetic dysfunction on the RIGHT and consistent with Harlequin syndrome.

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