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Teaching case: Headache stigmata

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TEACHING CASE: HEADACHE STIGMATA

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Case Report

A 38-year-old man with past medical history of hypertension and obesity presented to our clinic for evaluation of severe headaches and multiple episodes of discoloration of the forehead on the same side as the headache.

The patient developed headaches starting in 2003. An inciting event might have been a traumatic eye injury. He was drinking a beer with a partially broken bottle and glass shards went into his left eye. On multiple occasions he had to have his eye washed out and suffered significant eye discharge and irritation at the time, but without significant headache. Two months after the accident he started to develop severe pain located at the left eye, forehead, and temple. The headaches resolved in about 1 month and recurred again a few months later. In May 2006, the pain recurred and has not resolved since that time. He experiences severe headaches lasting 30– 90 minutes about 3–4 times per week with nausea, photophobia, left eye tearing, redness, and ptosis. At times with the severe headaches, the patient experiences blurry vision of the left eye, anxiety, confusion, and concentration difficulties. The pain is described as throbbing, stabbing, and shooting. Many of the attacks wake him up at night and few attacks occur in the afternoon. During these severe attacks, the left forehead hurts to touch. He usually prefers to sit still with severe attacks, but the pain is not exacerbated with movement. Oral medications, such as hydrocodone, provided little relief, and he has been treated with amitriptyline, duloxetine, valproic acid, topiramate, riboflavin, and pregabalin for headache prevention with no clinical improvement. He also takes metoprolol for hypertension. In between attacks, there is a constant level of pain around the left eye that never resolves with more of a dull quality. This baseline pain is typically mild or moderate. Alcohol and stress are headache triggers.

Past medical history is notable for hypertension, obesity, and the eye injury as above. He also was seen by an ophthalmologist and found to have decreased visual acuity (20/70) of the left eye. No adequate explanation was given for the visual loss. He smokes about 1 pack/day. He works as a heavy equipment operator, but is unable to work due to headache. Family history is notable for hypertension and diabetes. Review of systems was significant for insomnia, fatigue, and weight gain.

Evaluation before initial visit including MRI brain with thin pituitary cuts, MR angiography of the head and neck, and MR venography of the head. All were unremarkable. Head CT showed only a deviated nasal septum. He was found to have a low testosterone level for his age and sex (50 ng/dL) and is treated with androgel with improvement in libido, but no significant headache improvement. A routine CBC and basic metabolic panel were normal and ESR was within normal limits.

The patient's initial physical exam was notable for obesity, elevated blood pressure of 150/100, left-sided eye ptosis and decreased visual acuity, and decreased temperature sensation of the left face and arm to temperature and pinprick. Corneal reflex was present

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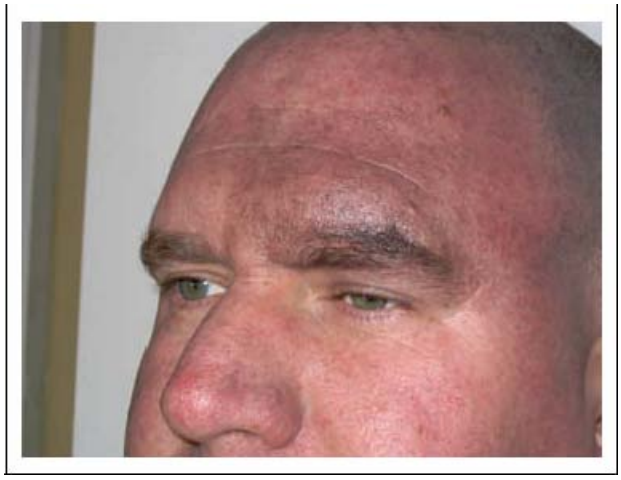
bilaterally. The remainder of the exam was normal.

The patient was placed on a brief course of prednisone and given subcutaneous sumatriptan and oxygen for acute attacks. For prevention, we elected to replace the patient's metoprolol with verapamil and discontinue riboflavin and amitriptyline.

On follow-up he reported oxygen was moderately helpful for severe attacks, but not the sumitriptan injection. He stopped the verapamil due to feeling off-balance and light-headed. He did experience dramatic short-term improvement while on prednisone. However, his headaches returned shortly after stopping the medication.

At the next office visit, he returned for evaluation of a new bruise on the left forehead (Fig. 1). The lesion developed after a sneeze and the patient did not notice it until he looked in the mirror. The color was initially bright red and the skin at that site was extremely painful to touch, darkened over the

Fig. 1



next few days and then gradually resolved (Fig. 2). Although the patient did have a moderate headache at the time, and he had a very severe headache the evening before, the lesion did not develop at a time of severe pain. This is at least the third time the patient has had this exact presentation of a bruise associated with sneezing and each time they have been on the left side.

EXPERT COMMENTARY

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The primary diagnosis in this case is cluster headache, initially episodic, with at least one remission. The headaches have probably become chronic since there has been no recorded remission after May of 2006. (See the criteria of the International Classification of Headache

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Disorders [ICHD]-2—Table 1).[1] In support of the diagnosis are the site, severity, and duration of attacks. The continuous low-grade pain between attacks of severe pain has been noted in the trigeminal autonomic cephalgias.[2] The frequency of 3–4 attacks per week is unusually low for cluster headache. The autonomic symptoms of left eye redness, tearing, and ptosis are typical of cluster headache, and the frequent occurrence of wakening the patient from sleep is similarly characteristic. The patient preferred to sit still during an attack; presumably he did not like to lie down. Often, patients during cluster headache will sit and rock. The patient's gender, alcohol as a trigger, and smoking history are all recognized features of cluster headache.

But, this is not a "pure" case. There are associated migrainous features, such as nausea, photophobia, and allodynia. The co-mingling of features of migraine and cluster headache is not unusual.[3] The dramatic response to prednisone is commonly noted in patients with cluster headache, but this patient's lack of response to subcutaneous sumatriptan is un-

Table 1.—(ICHD-2) Cluster Headache

Diagnostic Criteria:

A. At least five attacks fulfilling criteria B–D.
B. Severe or very severe unilateral orbital, supraorbital, and/or temporal pain lasting 15–180 minutes if untreated.
C. Headache is accompanied by at least one of the following:
   1. Ipsilateral conjunctival injection and/or lacrimation.
   2. Ipsilateral nasal congestion and/or rhinorrhea.
   3. Ipsilateral eyelid edema.
   4. Ipsilateral forehead and facial sweating.
   5. Ipsilateral miosis and/or ptosis.
   6. A sense of restlessness or agitation.
D. Attacks have a frequency from 1 every other day to 8 per day.
E. Not attributed to another disorder.

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usual. Under these circumstances, an opioid may be necessary to abort the acute attack until prophylactic measures become effective. Verapamil is the drug of choice for prophylaxis of cluster headache. I suggest that it be tried again at a low dose and the dose slowly built-up. Verapamil typically has fewer adverse effects than most other prophylactic agents.

Cluster headache has anecdotally been associated with trauma, but this relationship remains speculative.[4] Although prominent masculine features have been noted in patients with cluster headache, paradoxically low levels of testosterone have been found and treated in small series.[5] The decreased response to pain and temperature over the left side of the

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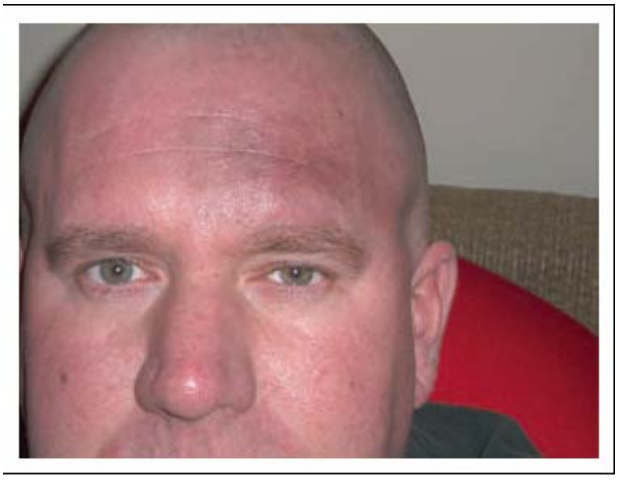
face and left arm is of uncertain importance. Curiously, allodynia was present over the same side of the face. The corneal reflexes were said to be “present,” but one wonders if the responses were equal. Sensory signs without corroborating features are often misleading.

Fig. 2



Bruises of the left side of the forehead after sneezing and only loosely associated with headache are an interesting feature in this patient, which may be unrelated to the headache pathophysiology. However, skin changes, including eyelid edema and periorbital echymosis, have been described in other cluster headache patients, and similar signs have also been noted in patients with migraine.[6-8] The pathogenesis of these phenomena is not understood, but autonomic vascular dysfunction may be responsible. Alternatively, some cluster headache sufferers habitually rub the forehead or face during pain episodes that might produce post-traumatic skin ecchymosis.

EPISODIC CLUSTER HEADACHE:
Description:
Cluster headache attacks occurring in periods lasting 7 days to 1 year, separated by pain-free periods lasting 1 month or longer.

CHRONIC CLUSTER HEADACHE:
Description:
Cluster headache attacks occurring for more than 1 year, without remission or with remissions lasting less than 1 month.

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QUESTIONS FOR DISCUSSION

1. What is the attack frequency range typically seen in Cluster Headache? Paroxysmal Hemicrania? SUNCT?
2. What is the proposed mechanism for Horner's syndrome in cluster headache?
3. List possible causes for the periorbital/frontal ecchymosis seen in cluster and other headaches.

This case presentation and discussion meets the ACGME requirements for residency training in the following core competency areas: Patient Care, Medical Knowledge, Practice-Based Learning and Improvement, and Systems-Based Practice.

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