Case Studies of Uncommon and Rare Headache Disorders

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KEYWORDS

• Status migrainosus • Cervical artery dissection
• Spontaneous intracranial hypotension • Hemicrania continua • Migraine with aura
• Limb pain and migraine • Cluster headache • Trigeminal neuralgia

KEY POINTS

- Status migrainosus often lasts a few weeks and may not respond to emergency department treatment.
- Cervical artery dissection can mimic migraine and present with headache and/or neck pain only.
- Spontaneous intracranial hypotension can present with a variety of headaches and be difficult to diagnose.
- Hemicrania continua and cluster headaches are commonly misdiagnosed.
- There are a diverse number of migraine auras.
- Migraine can cause limb pain with or without headache.

About 20% of the practice of the general neurologist consists of seeing patients with headaches, most of whom have primary headache disorders, mainly migraine. These case studies may be of benefit when the uncommon or rare headache disorder presents.

CASE 1. A 12-DAY MIGRAINE WITH RECURRING AURA?

Case 1 is a 60-year-old man with a history of migraine with and without aura since his 40s up to 25 days per month, decreased to about 5 per month on gabapentin 600 mg 3 times a day previously followed for 3 years. The headaches could be at the back of the head or generalized or occasionally with bitemporal pressure with an intensity of 3 to 10/10 associated with light and noise sensitivity but no vomiting relieved by eletriptan

Disclosures: Speaker's bureau, Allergan, Depomed, Avanir and Pernix, Teva; advisory board, Lilly; royalties or editorial fees from Elsevier, Lippincott Williams & Wilkins; Medlink Neurology; Oxford; Practical Neurology, and UptoDate.
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Neurol Clin 34 (2016) 631–650
http://dx.doi.org/10.1016/j.ncl.2016.04.006 **neurologic.theclinics.com**
0733-8619/16/$ – see front matter

in 2 hours. He would occasionally have a visual aura of flashing lights in both eyes for 20 minutes before the headache.

He presented with a 12-day history of a right temporal, behind the right eye and occasionally right back of the head, sharp pain with an intensity of 4/10 at onset and 7 to 9/10 since associated with nausea, light, and noise sensitivity but no vomiting on a daily basis lasting 2 to 8 hours with ibuprofen or eletriptan. He reported blurred vision with things missing in the right eye intermittently for 3 to 5 hours daily from the day of onset of the headache for 10 days. He had no fever. His primary care physician had placed him on a methylprednisolone dose pack without help. Examination by an ophthalmologist was normal. Neurologic examination was normal.

Question: What is your diagnosis?

Perhaps he has migraine status, which is migraine lasting longer than 72 hours. The headache is different than prior migraines but seems to fit migraine criteria: the headache is unilateral associated with nausea, light, and noise sensitivity. In addition, he has been having daily episodes of blurred vision in the right eye, which may be a migraine aura.

The prevalence of migraine status is uncertain but is probably rare. In a retrospective French study of 25 patients with status migrainosus seen in a tertiary care center (of 8821 migraineurs over 11 years), the demographics were as follows: mean age at first episode, 39 years; male:female ratio, 4:21; duration 4.8 weeks (3–10); relapse of status migrainosus, 32%; and delay of relapse, 61.5 months.[1] Precipitating factors included the following: stress/anxiety, 69%; menses, 31%; and lack of sleep, 6.3%. The great majority of patients had the same attack frequency before and after the status, and most cases occurred in those with low-frequency migraine attacks.

Question: What treatments are effective for migraine status in the emergency department?

Adequate IV hydration is important as indicated, and nonopioids are preferred because the emergency department (ED) visit may be longer than with use of nonopioids[2] and risk of habituation and abuse. Medication use may be guided by comorbidities, which are contraindications to use (such as cardiovascular, cerebrovascular or cerebrovascular disease, history of gastrointestinal bleeding, and pregnancy), prior response to medications, and medications taken before the ED visit. There are a variety of potential adverse events with the medications, including movement disorders with dopamine receptor antagonists.[3]

Little has been published on treatment of migraine status in the ED. Rozen[4] reviewed ED and inpatient management of migraine status.

The Canadian Headache Society performed a systemic review of 44 studies of acute treatment of migraine in the ED and found many were of poor quality and lacking in comparator trials.[5] The Society strongly recommended the use of prochlorperazine IV, metoclopramide IV, sumatriptan subcutaneous, and ketorolac intramuscular. The Society recommended strongly against the use of dexamethasone IV and haloperidol IV.

Persistent or recurrence of migraine without status is common after the ED visit. In a study of 186 migraineurs who presented for treatment to the ED with headaches present with a median duration of 24 hours (12–72 hours), 31% had moderate or severe headache present within 24 hours of ED discharge.[6] Although no data are available, ED treatment of migraine status may not be adequate, and some patients may need to be admitted for longer treatment. In the French study of 25 patients with status migrainosus, 60% required hospitalization for a mean of 6 days and were given IV amitriptyline (not available in the United States). Dihydroergotamine