

JAMA Neurology Clinical Challenge

Keeping an Eye on This Headache

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Figure 1. The patient during the first pain episode, with a red left eye, lacrimation, and periorbital swelling.

A 24-year-old woman was referred to our clinic for evaluation and treatment of a first-ever episode of headaches that occurred 16 days before the consultation. She reported a strong unilateral headache located in her left cheek, eye, and forehead that evolved during the evening hours, peaked at night, and decreased again until the morning, without any remarkable trigger. This first episode went along with ipsilateral lacrimation, a red eye, and periorbital swelling but without ptosis, which she documented with her smartphone (**Figure 1**). During a painful night, she could not sleep. Pain remitted, but the swelling increased until the next morning and slowly decreased over approximately 36 hours. Recurrent but less intense episodes of several hours' duration occurred during the 2 subsequent nights with no symptoms during the following 14 days. The patient reported no nasal congestion, nausea, vomiting, or phonophobia or photophobia. The patient reported being an infrequent smoker, with about 2 cigarettes a month.

She had no history of impaired vision, diplopia, facial trauma, prior head or neck surgery, or allergies. Examination of her head and neck showed no particularities, periorbital edema or proptosis, or enlarged lymph nodes. The neurological examination was unremarkable, with normal visual acuity, symmetrically reactive pupils, and unimpaired ocular movements, as was a differential hemogram, which showed normal white blood cell count of 5500/ μL (to convert to $\times 10^9/\text{L}$, multiply by 0.001).

WHAT IS YOUR DIAGNOSIS?

- A.** The first episode of cluster headache episodes
- B.** Tolosa-Hunt syndrome, a retrobulbar inflammation
- C.** A space-occupying mucocele of the nasopharynx
- D.** An acute episode of angle-closure glaucoma



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Diagnosis

C. A space-occupying mucocele of the nasopharynx

Discussion

The patient reported diagnostic features suggestive of a cluster headache, including the episodic character (often at nighttime), strictly unilateral pain, restlessness, and trigemino-autonomic symptoms (eg, lacrimation, conjunctival injection, rhinorrhea or nasal congestion, eyelid edema, miosis, and ptosis) ipsilateral to the headache. These symptoms fulfilled core criteria of cluster headaches, although there were an insufficient number of episodes and a prolonged episode duration.

Nevertheless, after evaluating the self-made photograph and time course of the first episode, several indications against a primary headache disorder were revealed: no abrupt start and ending of the pain episodes, a duration more than 3 hours, female sex, no nasal autonomic symptoms, and periorbital instead of eyelid swelling that persisted after the episode. Furthermore, the duration of periorbital swelling exceeded by far what is usually seen in cluster headache episodes, as well as in Tolosa-Hunt syndrome and an acute glaucoma episode.

With regard to Tolosa-Hunt syndrome, at least 1 of the cranial nerves responsible for ocular movements should be paretic.¹

An acute episode of glaucoma could be ruled out by normal intraocular pressure and a normal ophthalmologic examination. The typical presentation is associated with conjunctival injection, yet the extent of autonomic symptoms and periorbital swelling, as well as the self-limiting character of the pain episodes and absent visual impairment, would be very unusual.²

Magnetic resonance imaging of the head revealed a mucocele with inflammatory changes (Figure 2) and a slim gadolinium-contrasted boundary as the underlying cause. Nasal endoscopy showed the bulging mucocele. Surgical removal was performed, and pathohistologic examination results confirmed clinical-radiologic diagnosis of a sinus mucocele. The episodes remitted, and the patient experienced no pain or visual impairment.

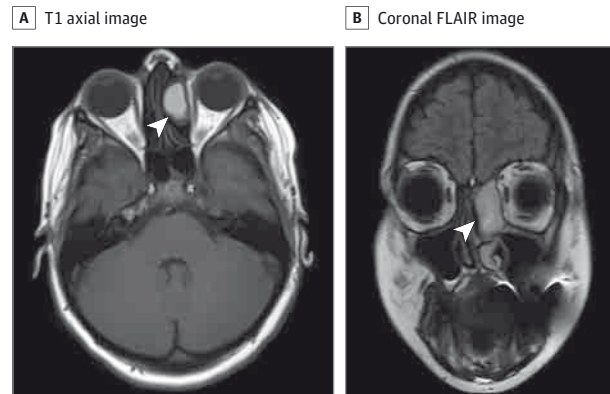


Figure 2. T1 axial (A) and coronal fluid-attenuated inversion recovery (FLAIR) (B) magnetic resonance imaging, showing a mucus-rich (with relative high intensity on T1) bulging mucocele of the left nasal cavity (arrowheads).

Mucoceles extending into the orbita are usually associated with proptosis, compromised visual acuity, and impaired ipsilateral bulb movements with diplopia. Spontaneous remittance of pain and swelling is rather unusual. In a retrospective study by Lee et al,³ 82 cases with intracranial or intraorbital extension of sinus mucoceles were reported; headache was documented in only 1 case and eye pain in 3 cases. Obeso et al⁴ reported craniofacial pain in 10 of 72 patients (14%) operated on for 81 mucoceles; 2 patients were diagnosed as having trigeminal neuralgia. Rapid recognition is mandatory owing to potentially devastating consequences for the eye and the optic nerve.^{5,6}

This case underscores the criteria¹ to diagnose cluster headache, especially with regard to number (>5 episodes) and duration (15-180 minutes) of the extremely stereotyped episodes. The International Headache Society criteria require that the symptoms not be better accounted for by another diagnosis. Cerebral magnetic resonance imaging is therefore recommended to exclude a lesional etiology of cluster-like episodes.

ARTICLE INFORMATION

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