Chronic paroxysmal hemicrania (CPH) is a rare, although well characterized, primary headache syndrome. The International Headache Society (IHS) classification criteria require at least 50 attacks of severe, unilateral, orbital, supra-orbital, or temporal pain lasting 2 to 180 minutes if untreated, and associated with at least one of the following autonomic features: conjunctival injection, lacrimation, nasal congestion, rhinorrhea, ptosis, or eyelid edema. The typical attack frequency is five or more attacks daily. The IHS criteria require that the attacks respond absolutely to treatment with indomethacin.

CPH and cluster headache are grouped together in the IHS classification since they share numerous clinical features. The IHS classification criteria for cluster headache require at least five attacks of severe, unilateral, orbital, supra-orbital, or temporal pain lasting 2 to 45 minutes if untreated, and associated with at least one of the following autonomic features: conjunctival injection, lacrimation, nasal congestion, rhinorrhea, forehead and facial sweating, miosis, ptosis, or eyelid edema. The typical attack frequency is from five or more attacks daily.

CASE REPORT

A 60-year-old, right-handed man without a prior headache history had a traumatic injury when a metal plate landed on his right supra-orbital ridge. There was no loss of consciousness or amnesia. The patient developed right periorbital bruising and swelling without a laceration. The initial moderately intense, constant, right orbital pain evolved into a right-sided intermittent headache after 3 days. He described excruciating, sharp, stabbing, retro-orbital, orbital, and forehead pain that frequently radiated in a straight line along the temporal and parietal region to the occiput. The pain occurred in discrete bouts lasting 20 to 40 minutes each, recurring three to five times daily. The attacks were associated with ipsilateral ptosis, lacrimation, and rhinorrhea. He experienced marked photophobia and preferred to lie still in a dark room during the attack. He denied nausea or vomiting, phonophobia, and osmophobia.

At the onset of the headache, the right side of his neck felt stiff for a couple of minutes. This then evolved into a tingling sensation like “pins and needles” which, over a period of 5 minutes, gradually moved down from the neck and shoulder, along the upper limb to the right hand. Once the tingling sensation reached the right hand, it continued unabated until the headache ceased. During most attacks, his right upper limb felt mildly weak, but this was not functionally significant. However, two attacks had been associated with marked right upper limb weakness to such a degree that he had difficulty lifting even light objects. The motor symptoms typically began a couple of minutes after the onset of the headache and continued throughout the headache. These sensory and motor symptoms never occurred without a headache.

He had tried paracetamol (acetaminophen), which had no effect. There was no past medical history of note. There was no family history of headaches. He ceased smoking 15 years before the onset of these headaches. He drank alcohol socially. Alcohol consumption did not trigger these headaches.

When we first saw him, he had been having these headaches for 5 months. Physical and neurologic examination was normal except for loss of all sensory modalities over the medial aspect of the right upper and lower eyelids, and medial aspect of the area above the right supra-orbital ridge. Routine hematologic and biochemical screening was normal. MRI of the brain did not reveal any abnormalities.

He was started on indomethacin 25 mg three times daily, and his headaches ceased completely after 2 days.
However, he infrequently had episodes of autonomic and aura symptoms without any accompanying pain. When present, these symptoms were exactly the same in character and duration as before the initiation of indomethacin except for a mild reduction in their intensity. Cessation of indomethacin led to recurrence of typical pain episodes within 2 days. While not taking indomethacin, the patient did not have any episodes of autonomic and aura symptoms without accompanying pain. Recommencing indomethacin at a higher dose of 25 mg four times daily led to the complete suppression of pain and autonomic and aura symptoms. He has now been asymptomatic for 3 months.

**Discussion.** The patient’s attacks were strictly unilateral, extremely painful, centered on the periorbital and forehead areas, and accompanied by ipsilateral autonomic phenomena. The intensity and character of pain, the duration and frequency of attacks, as well as the dramatic response to indomethacin are consistent with a diagnosis of CPH. However, our patient also satisfies the diagnostic criteria for cluster headaches. The male sex favors a diagnosis of cluster headache. The attack frequency and duration do not differentiate CPH and cluster headache, though the attack frequency is at the lower end of the range for CPH. The response to indomethacin strongly favors a diagnosis of CPH. There are rare case reports of so-called indomethacin-responsive cluster headache; we are unconvinced that many of these were not cases of CPH. As currently understood, the hallmark of CPH is the dramatic and complete response to indomethacin. We feel that cases of indomethacin-responsive headaches which satisfy diagnostic criteria of both CPH and cluster headache should be classified as CPH until the pathophysiological basis of these conditions is better understood.

We have described a case of CPH with an associated migrainous sensory and motor aura. A comprehensive review of CPH published in 1989 made no mention of associated aura symptomatology; and we have been unable to identify any case reports since. The occurrence of aura in CPH raises the possibility of coexistence of migraine and CPH. However, our patient denied suffering from migraine and, in fact, did not have a family history of migraine. He was never headache-prone. Aura symptoms have been described in association with cluster headache. A prospective physician interview study with patients from both referral clinics and patient groups has reported an aura rate of 14% with typical visual and sensory aura. In another recent series, aura symptoms were reported in 6% of cluster headache patients. In the former series, we raise the possibility that aura symptoms in cluster headache may be consequent to inheritance of aura genes. However, we noted that this is unlikely to be the entire explanation because one patient had both cluster headaches with aura, and migraine without aura. In this context, it is interesting to note that published case reports of coexistent migraine with aura and CPH do not allude to the presence of aura in association with CPH.

An interesting facet of this case report is that the headache occurred less than 14 days after the head injury, and had continued for more than 8 weeks after the injury, thus satisfying the IHS classification criteria for chronic post-traumatic headache. Again post-traumatic CPH must be very uncommon. The clear temporal relation between the head injury and onset of headache, and a relation of the location of the injury to the location of the pain, is highly suggestive of a causal or, more likely, a precipitous role for head injury in the onset of CPH in this patient.

Another remarkable observation in this patient was the dissociation of pain and autonomic features after administration of indomethacin 75 mg daily, but the abolition of all symptoms on indomethacin 100 mg daily. In one case report of CPH, there was a similar dissociation of pain and autonomic features, although this dissociation was only observed in the period after withdrawal of indomethacin. It has previously been demonstrated that some autonomic features may precede the pain, thus it is unlikely that the pain alone triggers the autonomic features, and pharmacologic inhibition of some autonomic features does not seem to influence pain. Our current concept is that CPH, like cluster headache, is essentially a brain disorder that entrains the trigeminal-autonomic system; it may do so independently, although when fully activated the trigeminal-autonomic reflex acts as a positive feedback loop to worsen the individual acute attack. Two great questions remain: where is the lesion in CPH, and how does indomethacin inhibit this syndrome? For the moment, practicing neurologists might think of paroxysmal hemicrania as being operationally defined as episodic headache responding to indomethacin; and remember to test therapeutically for such a response.

**References**

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NeuroImages

Figure. CT scan of the cervical-occipital hinge. Thin curvilinear, double band, periodontoid calcifications are detected in coronal and sagittal views (black arrows). Degenerative changes can be found at the articular surfaces (gray arrows).

Crowned dens syndrome in an elderly man

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An 89-year-old man presented to the emergency room with neck pain and fever (37.5°). Neurologic examination showed neck stiffness; general examination was normal. Erythrocyte sedimentation rate was 97 mm/1st h, fibrinogen 681 mg/dL, C-reactive protein 8.4 mg/dL. Other biochemistry, hematology, brain CT scan, CSF, and MR scan of the cervical spine were normal. Cervical CT scan revealed periodontoid calcifications. A clinical diagnosis of axial localization of calcium pyrophosphate dihydrate (CPPD) arthropathy was made. Therapy with a 50-mg oral dose of indomethacin provided complete recovery within 3 days and normalization of inflammatory markers.

CPPD arthropathy1 may involve any joint. Cervical localization is related to several clinical presentations: meningeal syndromes, compressive myelopathy, or an association of feverish acute cervical pain and calcifications in the periodontoid space, described as crowned dens syndrome.2

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