

Remitting Chronic Paroxysmal Hemicrania or Episodic Paroxysmal Hemicrania?

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Recurrent, short-lasting, nocturnal headaches can be a diagnostic challenge as illustrated by this patient's story.

CLINICAL HISTORY

This 75-year-old woman has a 2-year history of recurring headaches. The first cycle of headaches lasted for about 1 month. Since then, the headaches recur every few months and last about 3 weeks. This recurrence started 3 weeks ago. The headaches almost always occur during the night awakening her from sleep. She describes a left retro-orbital and peri-orbital severe pressure and stabbing associated with tearing of the left eye, swelling of the left eyelid, and either clear drainage or congestion of the left nares. During the headache, she has to get up and pace and cannot lie or sit still. The headaches last 1 to 2 minutes and can recur about every 2 hours. During the headache periods, the headaches occur zero to four times daily, with an average of about three daily. She had seen other physicians and was tried on lithium as a preventive and butorphanol tartrate nasal spray as symptomatic treatment without benefit. A magnetic resonance imaging scan of the brain was normal. She has a past medical history of essential tremor and a postganglionic Horner syndrome present for 20 years with a negative evaluation. There is also a history of

hypertension but no diabetes, ischemic heart disease, or cerebrovascular disease. Neurological examination was normal except for the left Horner syndrome and a mild action and postural tremor.

She was started on indomethacin sustained release, 75 mg daily. For a few days, she had two to three typical headaches occurring during the day while awake lasting 1 to 2 minutes, but less intense than previously and no nocturnal headaches. The headaches then completely resolved. She discontinued the indomethacin after about 1 month and has had no headaches for 6 weeks.

Questions.—Is this case consistent with remitting chronic paroxysmal hemicrania or episodic paroxysmal hemicrania? Are they distinct entities or different names for the same disorder? If she was not able to tolerate indomethacin, are there any other medications which might be of benefit?

EXPERT COMMENTARY

The International Headache Society (IHS) classification defines cluster headache (CH), episodic and chronic, and chronic paroxysmal hemicrania (CPH). Around these core entities are a number of less well-defined syndromes such as cluster-tic, cluster migraine, episodic paroxysmal hemicrania (EPH), short-lasting unilateral neuralgiform pain with conjunctival injection and tearing (SUNCT), and hypnic headache. In addition to these proposed diagnostic entities are many individual cases, each presenting with somewhat variant symptomatology. In specialized centers, many such cases are seen, for example, patients ful-

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filling all diagnostic criteria for CH but who have attacks lasting 6 or more hours.

The present case is impossible to fit into one particular set of IHS diagnostic criteria. She has very short-lasting attacks fitting CPH or SUNCT, but the attacks occur periodically and are too few for a typical CPH or SUNCT. They are not sufficiently localized to the eye for SUNCT, but in their quality, typical of CH or CPH. Clinical conclusion: most compatible with EPH, which, by now, is fairly well described.

Another clinical question—could there be a connection to the Horner syndrome of long standing but on the same side as the present attacks? I am unaware of any association between Horner syndrome and subsequent CH or CPH, but maybe one of our readers has seen it? The last comment relates to the

therapeutic response. The patient had no benefit from lithium, which is also more known for its effect in chronic cluster. Also, butorphanol tartrate nasal spray was without effect. This treatment is poorly substantiated, and the negative result is not helpful in the diagnostic considerations. There was an excellent response to indomethacin, a drug that is effective only in CPH and EPH. This certainly supports the latter diagnosis, but it is also possible that the recovery was spontaneous and not related to the treatment because indomethacin was started at about the same time; based on the usual duration of the patient's symptomatic period, a remission might have been expected. If efficacy of indomethacin can be proven during the next period, then a diagnosis of EPH must be almost certain.

INVITATION TO SUBMIT CLINICAL DILEMMAS

The "Expert Opinion" section of *Headache* is edited by Randolph W. Evans, MD who invites readers to submit clinical problems, case reports, or other headache-related questions for commentary by one or more experts. Comments on the expert's opinion as letters to the editor are also welcome.

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