Letters

Cluster headache and chronic paroxysmal hemicrania: How to classify borderline cases?

Sir: A 51 year old male clerk had suffered from severe paroxysmal headache for more than 25 years, when he was referred to the Migraine Centre, Department of Neurology, Medical Academy in Łódź. The headache was strictly unilateral and it began in the left temple. It spread to the left aural concha, maxilla and orbit and it radiated to the vertex. The pain was accompanied by slight tearing of the left eye and by obstruction of the left nasal duct. The patient had also the feeling of pulsation of vessels in the left temporal area. There were no other accompanying phenomena. He was symptom free between attacks. The headache attacks were of relatively short duration, lasting 15–20 minutes.

The bouts of disease appeared once or twice per year and lasted from 1 to 7 weeks, and they were separated by periods of full remission. The number of attacks ranged between 4 and 17 per day and was highest in the middle of the bout. The headaches occurred regularly at all times of the day and night. The intensity of pain reached the maximum when the headaches were most frequent. There were no known factors that either precipitated or ameliorated the condition and alcohol did not worsen it. Apart from his headaches the patient was in good health. His family history revealed no chronic headaches. The patient was seen by a number of physicians. The previous medications included ergot preparations, carbamazepine, pizotifen, oxynphenbutazon and various non-narcotic analgesics, all without benefit. Only large amounts of aspirin (up to 10 (400 mg) tablets per day) alleviated, but did not stop, the headache.

Physical and neurological examinations were within normal limits, when the patient was first seen by us 5 years ago. Blood pressure was 120/80 mm Hg. Routine laboratory tests, skull radiographs and EEG were normal, as were ophthalmological and laryngological examinations.

When the next bout developed the patient was admitted to the Department. Based on the report of the ameliorating effect of aspirin on his headache, indomethacin (100 mg per day) was administered to the patient. After 2 days of medication the attacks disappeared and re-appeared 2 days after replacing the drug by placebo. Indomethacin given again stopped the attacks. When the treatment was discon- tinued 7 weeks later, the patient was completely free of headache.

The pattern of the disease has not changed during the last 5 years and the patient has suffered from 1–2 clusters of attacks yearly. Each time he was successfully treated with indomethacin 100 mg per day. If the gradual decrease of the dose 3–4 weeks later caused recurrence of abortive attacks, the initial dose was re-administered for some weeks. He did not need indomethacin therapy longer than 8 weeks. During one of the exacerbations lithium was tried, but with no effect. During the last year the patient was systematically treated with propranolol for hypertension, which was diagnosed, but this drug did not prevent the last bout of illness.

Chronic paroxysmal hemicrania is a head-ache syndrome originally described by Sjaastad and Dale in 1974. The disease has some clinical features in common with cluster headache: localisation and unilaterality of pain, its severity and concomitant autonomic phenomena. On the other hand, when in 1980 first known cases of chronic paroxysmal hemicrania were carefully analysed, among "the specific traits that separate chronic paroxysmal hemicrania from Horton’s disease” the following were considered absolute for the diagnosis: (1) frequency of attacks (maximum = 15 in 24 h), (2) headache every day, and (3) good indomethacin effect. Since that time numerous new cases fitting diagnostic criteria for chronic paroxysmal hemicrania have been described and the disease appears to be an entity with rather a definite clinical picture separate from cluster headache and of unknown aetiology.

In our case the pattern of the disease was typical of episodic cluster headache, that is, the pain attacks appeared in clusters separated by periods of full remission. In chronic paroxysmal hemicrania the typical chronic stage may be preceded by atypical headaches lasting for years (pre-CPH stage), but in our case the pattern seems to have been well established. The frequency of attacks and their duration fit the criteria for chronic paroxysmal hemicrania rather than cluster headache. Dramatic improvement after indomethacin administration is characteristic of chronic paroxysmal hemicrania.

Our case is strikingly similar to the one described by Geaney, the only differences being that the pain in our patient was for 25 years on the same side, and no pupil abnormality was observed.

Responsiveness of chronic paroxysmal hemicrania to indomethacin is of great clinical value, as the drug can completely abolish the headache. Other non-steroidal anti-inflammatory drugs have been reported to ameliorate chronic paroxysmal hemicrania. The efficacy of these drugs strongly suggests the disturbed metabolism of arachidonic acid via cyclooxygenase pathway, as the pathogenetic factor of chronic paroxysmal hemicrania. On the other hand, it is well known that cluster headaches do not respond to indomethacin.

From the diagnostic criteria cited above “complete indomethacin effect” appears to be the most significant. Geaney classified the case described by him as “indomethacin-responsive episodic cluster headache.” We find the term inappropriate and misleading as it suggests that there are cases of cluster headache which may be treated successfully with indomethacin. We suggest the borderline cases should be classified using the criterion of responsiveness to indomethacin, not the clinical picture alone. It is our belief that our case should be diagnosed as chronic paroxysmal hemicrania with relatively low frequency of attacks and with occurrence of spontaneous remissions.

For some years one of us (AB) observed a male patient who suffered from short-lasting (max. 20 min.) and frequent (up to 30 per day) one sided headaches daily. Indomethacin was given without response, but lithium produced disappearance of attacks and the drug was effective in the next 3 years. In spite of the clinical picture, the diagnosis of chronic cluster headache was made.

One may expect that the spectrum of borderline cases is relatively wide and some patients diagnosed earlier as cluster headache were in fact suffering from chronic paroxysmal hemicrania.

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References
Chronic paroxysmal hemicrania, episodic cluster headache and classic migraine in one patient

Sir: Cluster headache has a prevalence of 2.4/1000. The reported male to female ratio is between 4.5 and 12:6:1 in different series. There is no increased incidence of migraine, either in the cluster headache patients or in their families.

Chronic paroxysmal hemicrania is a much rarer disorder, with less than 40 patients reported in the world literature in 1983. Recorded cases have been almost exclusively in women. A dubious “pre-chronic paroxysmal hemicrania stage” is described, where patients have rather atypical tension or migraine headaches; this occurred in five of eight patients reported by Sjaastad. Rapaport has recorded chronic paroxysmal hemicrania in a patient with classical migraine earlier in life, and there have been three possible cases of chronic paroxysmal hemicrania in patients with cluster headache.

Although chronic paroxysmal hemicrania is sometimes regarded as a variant of cluster headache, there is no evidence that cluster headache predisposes to chronic paroxysmal hemicrania, nor that chronic paroxysmal hemicrania occurs with increased frequency in migraine or cluster headache patients. We have found no recorded example of a single instance of the three syndromes in the same individual. We record the possibly unique instance of a patient with classical migraine, cluster headache and chronic paroxysmal hemicrania occurring independently over a 40 year period.

A 58 year old man presented in 1978 with a two year history of episodic headaches. They occurred every two weeks, but there were also bouts in which he had attacks daily for 3 to 4 days, notably more frequent in the evenings and occasionally at night. Alcohol would precipitate attacks. They started without aura as a severe localised throbbing pain in the left eye radiating to the left forehead which was tender and felt hot. During attacks the left eye watered and was visibly bloodshot. They lasted 30 to 120 minutes. There was no nausea or vomiting, or disturbance of vision. General health was good; he smoked cigars, and drank ¼ bottle whisky a day.

He gave a history of attacks of migraine from the age of 16 years until 20 in which he had expanding flashing lights in either visual field, followed in half an hour by throbbing unilateral headache, nausea and prostration for 18 to 24 hours. These occurred irregularly perhaps 3 or 4 times a year. His mother had suffered from “severe migraine”, but there was no other past illness or family history of note.

No abnormality in the CNS was found except for left sided ptosis suggesting a mild Horner’s syndrome. He had frequent extrasystoles, but no other abnormality in the heart, neck vessels, peripheral pulses, chest or abdomen. Blood pressure was 150/85 mm Hg.

Fig  Daily record of duration and intensity of episodic paroxysmal hemicrania. Attacks ceased on day 36.