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■ Case report ■

Bilateral paroxysmal hemicrania with autonomic features in a child: A case report

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= Abstract =

Paroxysmal hemicrania (PH) is rare in children and not widely recognized. It is characterized by pain attacks and associated symptoms and signs similar to those experiencing cluster headaches, but the features have a shorter effect, are more frequent, and respond completely to indomethacin. Some patients with PH may experience slight pain across the midline. There are only four cases of bilateral PH in the literature and it is very rare in children. Here, I report the case of a 10-year-old female with bilateral PH diagnosed by the typical symptoms along with the favorable response to indomethacin therapy. (Korean J Pediatr 2009;52:619-621)

Key Words : Bilateral paroxysmal hemicrania, Indomethacin, Child

Introduction

Paroxysmal hemicrania (PH) is a rare syndrome characterized by repeated attacks of strictly unilateral, severe, short-lasting pain occurring with cranial autonomic features. The attacks in PH are shorter lasting and more frequent compared with cluster headache (CH) but otherwise PH and CH have similar characteristics. The hallmark of PH is absolute cessation of the headache with indomethacin in varing doses and hence distinguishable from the other trigeminal autonomic cephalalgias such as CH and short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT). In 1992, Sjaastad[1] reported that some patients with PH had pain that crossed the midline slightly. There were only four cases of bilateral PH in the literature[2-5]. Three cases had headaches without autonomic features[2-4]. The last was the first case of PH with autonomic symptoms[5]. I report a 10-year-old female with bilateral PH with autonomic symptoms, which was diagnosed because of the typical symptoms along with a favorable response to indomethacin therapy.

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Case report

A 10-year-old female was referred to the Department of Pediatrics of Chosun University Hospital due to uncontrolled headaches that did not respond to analgesics. The pain, which had begun 2 weeks previously, was characterized by abrupt onset and cessation. The attacks were usually triggered by lack of sleep. The headache usually lasted 20-30 minutes, occurred daily and 4 times per day. The headache was usually very severe and experienced as a feeling of squeezing. The headache was always bilateral in the forehead area, associated with vomiting, osmophobia, lightheadedness, bilateral lacrimation, paresthesia around eyes (sensation of heat).

She had a 3-year history of remitting headaches. The attacks had a throbbing quality and were associated with nausea, vomiting, osmophobia and lightheadedness. The frequency was once or two per week, the duration was about five minutes, the severity was moderate. The patien was taking over-the-counter analgesics intermittently. Sometimes there were not helpful. She had not been previously evaluated by a physician. Family history showed that her father, grandfather and an aunt suffered from migraine. Before coming to our center she was treated with antibiotics and analgesic for maxillary sinusitis and headache at the secondary hospital for 2 weeks, but there were no effect.

In the process of admission to the hospital, she underwent

Table 1. The International Classification of Headache Disorders II for Paroxysmal Hemicrania

Diagnostic Criteria
At least 20 attacks fulfilling criteria B–D
Attacks of severe unilateral orbital, supraorbital, or temporal pain lasting 2–30 minutes
Headache is accompanied by at least one of the following:
Ipsilateral conjuctival injection and/or lacrimation
Ipsilateral nasal congestion and/or rhinorrhea
Ipsilateral eyelid edema
Ipsilateral forehead and facial swelling
Ipsilateral miosis and/or ptosis
Attacks have a frequency of 5 per day for more than half of the time, although periods with lower frequency may occur
Attacks are prevented completely by therapeutic doses of indomethacin
Not attributed to another disorder

neurologic examination, blood tests and neuroimaging. Routine hematology and blood chemistry studies were unremarkable. Neurologic examination, her brain MRI, MRA, and EEG revealed no abnormalities.

As inpatient treatment, the patients was attempted hydration and metoclopromide 0.2 mg/kg/dose at the onset of headache for three days. This provided significant relief on the severity and the frequency of attacks. And then, she was started on flunarizine for short-lasting other primary headache and ibuprofen (10 mg/kg/dose) taken at the headache onset, but not more than three times per week. Two days after discharge, she visited the emergency room because she had severe headache similar to previous attacks during admission. Thus, she was started on indomethacin with slow titration to 50 mg two times per day. Maximum effectiveness was obtained five days after initiating the effective dose. On follow-up one month later, she reported three similar mild to moderate headaches per day for three days after discharge. Over the ensuring two months the patients no longer experienced attacks and there were no adverse effects from indomethacin. Thereafter, I attempted to slowly withdraw indomethacin without pain recurrence. She has no more headaches over the next three months. The clinical picture and response to indomethacin confirmed a diagnosis of PH, based on International Classification of Headache Disorders II (ICHD-II) criteria[6] (Table 1).

Discussion

PH is defined by the following International Classification of Headache Disorders, 2nd Edition : ICHD-II) criteria[6] (Table 1). There are two forms: the episodic, which occurs during a period lasting 7 days to 1 years separated by pain-free periods lasting 1 month or more; and the chronic, which has attacks that occur for more than 1 year without remission or with remissions lasting less than 1 month. PH is rare in children and has not been widely recognized. PH usually occurs during adulthood at a mean age of 34 years (range 6-81 years)[7]. The condition predominates in females by a sex ratio of 1.6-2.4:1.

There are four case reports of short-lasting, frequent, bilateral, indomethacin-responsive headaches that have been presented as bilateral PH[2-5]. Three cases differ from PH in lacking unilaterality and being without cranial autonomic features. In addition, there was a paucity of migrainous symptoms in the bilateral cases. These three cases report pose an interesting question with regard to their classification: do they represent a bilateral form of PH or are they cases of another novel indomethacin-responsive primary headache syndrome? Matharu and Goadsby[8] propose that these cases are not bilateral PH at all but may constitute a new category of primary headache and propose the term, "bilateral paroxysmal cephalalgia". However, there are rare reports of unilateral PH without cranial autonomic features. Recently, Evans[5] report the first case of bilateral hemicrania with autonomic symptoms. I add a case of bilateral PH with cranial autonomic features, fully relieved with indomethacin 50 mg per day. I suggest that this case is a variant of PH. I recommend that trial of indomethacin should be considered in all patients who present with short-lasting, frequent, unilateral or bilateral, with or without cranial autonomic features. However, the significance of this pattern is not certain, as the natural historye of PH is not well described. Additional case reports are needed to characterize and certainly classify these headaches.

한 글 요 약

소아에서 자율신경계의 증상을 동반한 양측 돌발 반두통 1예

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노 영 일

돌발 반두통은 일측성이고 짧은 시간동안 심한 통증이 있는 경우이며, 대부분 두개의 자율신경계 증상을 동반한다. 이는 군발 두통의 통증, 동반 증상과 징후와 비슷하지만, 돌발 반두통은 지속시간이 더 짧고, 더 빈번하며, 인도메싸신에 잘 반응한다. 1992년에 Sjaastad에 의해 돌발 반두통이 반대쪽으로도 이동한다고 처음 보고하였으며, 문헌에는 단지 4례가 보고되었다. 4례 중 3례는 자율신경계 증상을 동반하지 않았으며, 1례만 자율신경계 증상 동반을 하였다. 저자는 10세 소아에서 자율신경계 증상을 동반한 전형적인 증상과 인도메싸신으로 치료된 양측 돌발 반두통 1례를 경험하였기에 문헌 고찰과 함께 보고하는 바이다.

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