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Hemicrania Continua

- A Case Report -

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= Abstract = **Probably due to rarity and unfamiliarity, hemicrania continua is not reported in Korea. We hereby report a woman who had a very rare unilateral headache syndrome, hemicrania continua. For about 20 days, a 58-year-old woman suffered from persistent unilateral non-throbbing headache which did not respond to analgesics and narcotics. Headache was maximal in the left occipital area intermittently radiating to the left temporofrontal and nuchal areas. There was no autonomic dysfunction, photophobia, phonophobia, nausea or vomiting. Extensive diagnostic evaluations, including cervical spine X-rays, cranial computed tomography and erythrocyte sedimentation rates, were normal. Her unilateral headache responded dramatically and specifically to indomethacin.**

Key words: *hemicrania continua, occipital area, indomethacin, tender point*

INTRODUCTION

Hemicrania continua (HC) is characterized by the continuous unilateral headache of moderate severity (Bordi *et al.*, 1991). In contrast to cluster headache, it seems to be more common (1.8:1) in female (Newman *et al.*, 1994). Indomethacin invariably causes complete remission and this is regarded as the sine qua non for diagnosis (Bordi *et al.*, 1991). To our knowledge, there is no report of HC in Korea. We herein report a woman with HC with maximal pain intensity in the left occiput.

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CASE HISTORY

A 58-year-old woman was admitted to the ospital due to moderate to severe unilateral (left) occipital headache intermittently radiating to the left posterior cervical and temporofrontal areas. Her headache developed rather suddenly about 15 days ago. She took several over-the-counter drugs but without symptomatic improvement. The headache was prickling and intermittently throbbing, and it fluctuated but continued all days long. It aggravated three days ago and always persisted except during sleep. However her headache did not disturb her sleep and there was no nocturnal exacerbations. There was a small tender point at the left retro-mastoid occipital area. Nuchal or sternocleidomastoid tenderness was absent. Also ab-

sent was shoulder or arm pain. There was no associated autonomic symptoms, photophobia, phonophobia, nausea or vomiting. There was no positional effect as well. Her headache was not aggravated by Valsalva maneuver or neck movements. The headache was exclusively unilateral (left) without side shift. She denied past history of significant headache, mastoiditis, otitis or head trauma.

She has had mild hypertension and asymptomatic diabetes for three months. Physical and neurological examination were normal. The scalp was not tender except for the left retro-mastoid area. Routine laboratory evaluations including erythrocyte sedimentation rate were normal. Skull X-rays and cervical spine series were normal as well. The contrast enhanced cranial computed tomography showed no abnormality as well. Acetaminophen, diazepam, amitriptyline and propranolol (120mg/day) were administered but failed to improve her pain. Even codein did not show significant improvement.

On the first day of indomethacin (25mg BID), her headache improved remarkably. On the second day of indomethacin (25mg TID), her headache disappeared nearly completely. She refused diagnostic withdrawal of indomethacin. For more than two months after discharge, she is still free from headache on indomethacin (25mg TID). She reported that skipping a dose of indomethacin disclosed the unilateral headache of the same nature.

DISCUSSION

Since the first description of HC (Sjaastad and Spierings, 1984), there have been more than 30 cases of unequivocal HC reported hitherto (Bordi *et al.*, 1991: Newman *et al.*, 1994). The strictly unilateral, almost continuous headache persistent for more than 20 days in our patient, which responded dramatically and specifically to indomethacin, is very compatible for the "clinical traits" of HC (Bordi *et al.*, 1991).

Although HC can locate in both anterior (orbital, frontal and temporal) or posterior (occipital, nuchal) area or both, the usual pain location is anterior area (Bordi *et al.*, 1991; Newman *et al.*, 1994). In the recent report of HC (Newman *et al.*, 1994), the headache location maximal in the occipital area, as in our patient, seems to be unusual. Of their ten patients, there was no HC with maximal intensity in the occipital area. Usually the headaches were maximal in the orbital area (Newman *et al.*, 1994). Among the 18 patients reviewed by Bordi *et al* (1991), five had occipital pain. Summing up these two reports, 18 percents (n = 5/28) of HC patients had occipital pain. Other cluster headache syndromes also usually locate in orbitotemporal area as their maximal intensity. This seemingly uncommon pain localization in our patient delayed the indomethacin trial for a week after admission.

The differential diagnosis includes cervicogenic headache and other cluster headache syndromes, especially chronic paroxysmal hemicrania. The cervicogenic headache may mimic HC because it is also strictly unilateral. The pain is usually initially felt in the nuchal and retroaural areas (Sjaastad, 1992a). The maximal pain is frequently in the temporal/ocular region and forehead. In our patient, both initial and maximal pain were in the left occiput. However the pain localization may not of definitely discriminatory value in differentiating this disorder from cluster headache (Sjaastad, 1992a). Headache of marked and constant intensity, as in our patient, is not characteristic of cervicogenic headache (Berger and Gerstenbrand, 1986). The manual examination of the cervical spine, which did not induce any pain in our patient, is the most important diagnostic method (Berger and Gerstenbrand, 1986). In addition, there was no nuchal tenderness, no radiating pain to the shoulder or arm, and no radiological abnormality of the cervical spine as well.

Chronic paroxysmal hemicrania is another headache syndrome that specifically responsive to indomethacin. It could be excluded because the pain was not paroxysmal and not too severe to disturb her sleep and not associated with autonomic dysfunction.

We do not think that the tender area in the left occiput is not compatible with HC. In HC and cluster headache tender spots can occur in the neck although not in the occiput as in our patient (Silberstein *et al.*, 1994; Sjaastad, 1992b).

Of the three temporal patterns of HC, our patient's headache belongs to a chronic, nonremitting headache from onset (Newman *et al.*, 1994). HC is

usually benign with moderate severity, but in two patients, headache was so severe and boring that they tried suicidal attempts (Bordi *et al.*, 1991). Thus early diagnosis seems to be very important to at least some patients.

Hitherto there is no reliable diagnostic method of HC except for the careful observation of the clinical profile and dramatic response to indomethacin. However indomethacin-resistant HC was described as well (Kuritzky, 1992). No gross abnormalities as regard the sympathetic function was found in HC (Antonaci *et al.*, 1992). Brain MRI studies were normal (Antonaci, 1994) except for a HC patient with a mesenchymal tumor in the right sphenoid bone involving the clinoid process and the base of the skull (Antonaci and Sjaastad, 1992).

The recently proposed diagnostic criteria of HC (Table 1) did not include any radiological or neurophysiological test (Silberstein *et al.*, 1994). The headache in our patient meets this criteria except for the shorter duration of headache. We do not think that the headache more than one month in this criteria is absolute for the diagnosis. Even though restricted by the short period of headache, we believe that our patient had HC with maximal intensity in the left occiput. High index of suspicion helped us early diagnosis and treatment.

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Table 1. Proposed Criteria for hemicrania Continua(HC)*

A. Headache present for at least one month
B. Strictly unilateral headache
C. Absolute response to indomethacin
D. Pain has all 3 of the following present:
   1. Continuous but fluctuating
   2. Moderate severity
   3. Lack of precipitating mechanisms
E. May have associated "jabs and jolts"
F. At least one of the following
   1. There is no suggestion of one of the disorders listed in groups 5-11**
   2. Such a disorder is suggested, but it is ruled out by appropriate investigations
   3. Such a disorder is present, but first headache attacks do not occur in close temporal relation to the disorder.

* HC is usually non-remitting, but rare cases of remission have been reported.

** groups 5-11: secondary headaches with underlying disorders

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