

Bifocal pain in nummular headache: A clinical analysis and literature review

¹Yi-Ting Chen MD, ³Chiu-Hsien Lin MD, ²Tzu-Hui Li MD, ²Lian-Hui Lee MD, ²Wei-Hsi Chen MD MSc LLM

¹Departments of Family Medicine and ²Neurology, Kaohsiung Chang Gung Memorial Hospital, and College of Medicine, Chang Gung University, Kaohsiung; ³Department of Family Medicine, Antai Tian-Sheng Memorial Hospital, Pingtung, Taiwan

Abstract

Background: Nummular headache is a new category of primary headache disorder characterized by consistent location, size, and shape of painful areas. The pathogenesis is uncertain. Bifocal painful areas are rare manifestations but may expand the clinical diversity of nummular headache. **Methods:** The clinical characteristics of 5 bifocal nummular headache patients were reported and those of 11 patients in previous studies were reviewed. Bifocal nummular headache was classified into two types. Type I was defined as a simultaneous activation of two painful areas while type II was defined as two painful areas occurring in different times. **Results:** All 16 patients were female, with mean age of onset and initial presentation of 54.7 years and 58.2 years, respectively. There were seven type I and nine type II patients. The parietal area, especially the tuber parietale, was the leading site of involvement in both types of patients. The shape and size of painful areas were also similar between these two groups. There was an equal frequency of ipsilateral and contralateral painful areas. The pain intensity was similar in both types of patients but was milder in new painful areas than in previous painful areas in type II patients.

Conclusions: Bifocal nummular headache suggests a central role of nummular headache but does not debunk the peripheral theory of nummular headache. The accumulated findings in bifocal NH patients do not support a generalization of pain occurrence or a reproduction of local process of epicranial neuralgia at multiple sites in nummular headache.

INTRODUCTION

Nummular headache (NH), or coin-shaped cephalgia, was first reported in 2002 by Pareja *et al.*¹ It is characterized by repetitive or continuous pain with a consistent location, shape, and size of delineated painful areas.^{1,2} The precise pathogenesis remains uncertain. In previous studies, bifocal involvement is occasionally mentioned. Although the clinical significance is unclear, this topographic presentation may expand the clinical diversity of NH. This study reports bifocal NH patients and compares them with other patients in previous studies.

METHODS

Between 2007 and 2012, 30 primary NH patients were identified based on the diagnostic criteria of the second edition of the International Classification of Headache Disorder.² They include in-patient and out-patient individuals, and those

referred from local hospitals or private clinics for headache or medication consultation. The patients had no cranio-facial trauma, systemic or focal infection, or treatment for co-morbid headache within the last three months. Each patient underwent a detailed review of their medical and headache history, and a battery of laboratory tests that included biochemistry, hematology, serology, and cranial computerized tomography (CT) or head magnetic resonance imaging (MRI) to exclude secondary disease.³

Bifocal NH was defined as two painful areas that fulfilled the diagnostic criteria of NH² present in the same patient. These two painful areas were either simultaneously activated in the same attack or alternatively in different attacks.

RESULTS

The results were summarized in Table 1 and Figure 1.

Subjects

The 30 patients included 28 females (93.3%) and 2 males, with mean age of initial presentation of 59.8 years (range, 38-77 years). Five had bifocal NH whereas 25 had unilateral focal NH. The mean age of initial onset and initial presentation was 57.04 ± 10.9 years and 61.36 ± 10.7 years, respectively, in focal NH patients and 48.5 ± 3.2 years and 52.0 ± 1.6 years, respectively, in bifocal NH patients (Table 1). Bifocal NH occurred earlier than unilateral focal NH. The patients had no previous diagnosis of NH.

Characteristics of painful areas

Case 1 was a 53-year-old woman who suffered pain on the left parietal area for 10 years and had additional pain at the right parietal area 1 year later. The shape of the painful areas was elliptical and the size was 3.5 x 3.0 cm. The intensity of pain was milder in the newer painful areas.

Case 2 was a 54-year-old woman who suffered simultaneous pain on the left parietal and right fronto-parietal areas for 3 years. The shape of the painful areas was elliptical and the size was 3.0 x 2.5 cm. The intensity of pain was similar in both painful areas.

Case 3 was a 51-year-old woman who suffered simultaneous pain on the right temporal and right parietal areas for 1 year. The shape of the painful areas was elliptical and the size was 3.0-3.5 x 2.5 cm. Pain intensity was similar in the painful areas.

Case 4 was a 54-year-old woman who suffered pain on the right parietal area for 3 years and had additional pain on the left parietal area 2 months later. The shape of the painful areas was circular and the size was 3.0 x 3.0 cm. Pain intensity was similar between the initial and the additional painful areas.

Case 5 was a 50-year-old woman who suffered simultaneous pain on the left parietal and left posterior temporal area for half a year. The shape of the painful areas was elliptical and the size was 3.0 x 2.5 cm. The intensity of pain was similar between the painful areas (Table 1; Figure 1).

Overall, the painful areas were ipsilateral in two patients (right and left side in one patient) and bilateral in the other three. In the bilateral bifocal NH patients, the painful areas were symmetrical in two and counter-coup in one.

Pain characteristics

Pain was described as "on" or "beneath" the scalp in all patients. No one reported pain deep

inside or experienced pain extending to other cranio-facial areas. The pain was relapsing and remitting (episodic), with the relapsing period usually ranging between 2 to 6 weeks.

The nature of pain in the bifocal painful areas was identical in each patient. Electrical pain was present in two patients, lancinating pain in two, and electrical and stabbing pain in one.

The severity of pain in each attack was also similar in the painful areas in each patient. During the attack, allodynia occurred in 3 patients (Cases 2, 4, and 5). The magnitude of allodynia was also similar in the painful areas in each patient. There were no trophic changes, redness, or edema in the painful areas.

Treatment and response

Gabapentin was prescribed as prophylaxis in Case 2 and Case 5. Case 1, Case 3, and Case 4 preferred abortive treatment with non-steroidal anti-inflammatory drugs. According to the patient's description, gabapentin, ranging from 100 to 300 mg/day, rapidly abolished their headache within two weeks. The pain did not recur within one year after treatment. The abortive treatment in these patients also reduced the frequency and intensity of painful attacks but did not completely abolish the pain.

DISCUSSION

In the current series, the painful areas showed a predilection for the parietal area (tuber parietale), a 2.5-3.5 cm diameter size, and elliptical or circular shape. Some patients had allodynia. The clinico-graphic and sensory algometry of painful areas are similar to those of focal NH patients or previously reported NH patients.^{4,5} The mean ages at initial onset and at presentation are younger in bifocal NH patients than in focal NH patients.

In reviewing literature, bifocal NH has been previously reported in 11 patients.⁶⁻⁹ Together with five patients in the current series, the 13 patients can be categorized into two different types (Table 1). Seven patients,⁸⁻¹⁰ including three in the current series (Case 2, 3, and 5), have Type I bifocal NH, which is defined as a simultaneous attack of two painful areas. The painful areas are ipsilateral in three patients, contralateral in three, and rostro-caudal in one (Figure 1). The parietal area (tuber parietale) is the leading site of involvement. In terms of painful areas, the shape is similar in each patient and the diameter ranges from 1.5 to 4.0 cm. The intensity of pain is also similar in both painful areas.

Table 1. The summary of bifocal nummular headache in our series and previously reported patients in literature

Case	Author(s) Year	Age (years)	Gender	Type	Location	Shifting period	Trophic change	Outcome
1	Chen <i>et al</i> , present series	53	F	II	Left parietal and then right parietal area	1 year	No	Favorable to analgesics
2		54	F	I	Left parietal and right frontoparietal area	-	No	Favorable to gabapentin
3		51	F	I	Right temporal and right parietal area	-	No	Favorable to analgesics
4		52	F	II	Right parietal and then left parietal area	2 months	No	Favorable to analgesics
5		50	F	I	left parietal area and posterior temporal area	-	No	Favorable to gabapentin
6	Rocha-Filho PA, 2011 ⁷	52	M	I	Right parietal area	-	No	Favorable to lidocaine injection
7	Guerrero <i>et al</i> , 2011 ⁹	61	M	II	Right parietal and then right temporal area	6 months	No	Favorable to analgesics
8		71	F	I	Bioccipital area	-	No	Favorable to gabapentin
9		80	F	II	Left parietal and then right parietal area	20 years	No	Favorable to carbamazepine
10		30	F	I	Paraemedian occiput and sagittal frontal area	-	No	Not mentioned
11		30	M	II	Right frontal and then right occipital area	2 months	No	Favorable to gabapentin
12		32	F	II	Left para-sagittal parietal and then right para-sagittal parietal area	5 years	No	Favorable to gabapentin
13	Ruscheweyh <i>et al</i> , 2010 ⁶	57	F	II	right parieto-occipital and then left temporal area	1 month	No	Not mentioned
14	Cuadrado <i>et al</i> , 2009 ⁸	28	M	II	Right parietal area and then left parietal area	10 months	No	Not mentioned
15		67	F	II	Right frontal and then right occipital area	2 years	No	Not mentioned
16		51	M	I	Bitemporal area	-	No	Not mentioned

Nine patients have Type II focal NH, which is defined as a new painful area occurring after a time period from the occurrence of the first painful area.^{5,8-9} The time period for the additional painful area to occur ranges from 1 month to 20 years after the onset of the initial painful area. The additional painful area is ipsilateral to the first in three patients and contralateral in six (Figure 1). Interestingly, the parietal area (tuber parietale) is also the leading site of the initial and the additional painful areas in type II patients. The shape is similar between the additional and initial painful areas in each patient, and the diameter ranges from 2.0 to 4.0 cm. In type II patients, the intensity of pain in the additional painful area is

either similar to or milder than that of the initial painful area.

Cuadrado *et al.*⁸ and Guerrero *et al.*⁹ proposed that a bifocal involvement in NH may represent a reproduction of the local process of epicranial neuralgia at multiple sites in the scalp. Guerrero *et al.*⁹ further presumed that central pain sensitization or local skin disorder may facilitate a multi-focal appearance of NH. Rocha-Filho⁷ also suggested a generalized disorder for NH, concluding that NH is a peripheral epicranial neuropathy and its expression is facilitated by other central or peripheral predisposing factors.

Based on the current series and previously reported patients, there are two points that

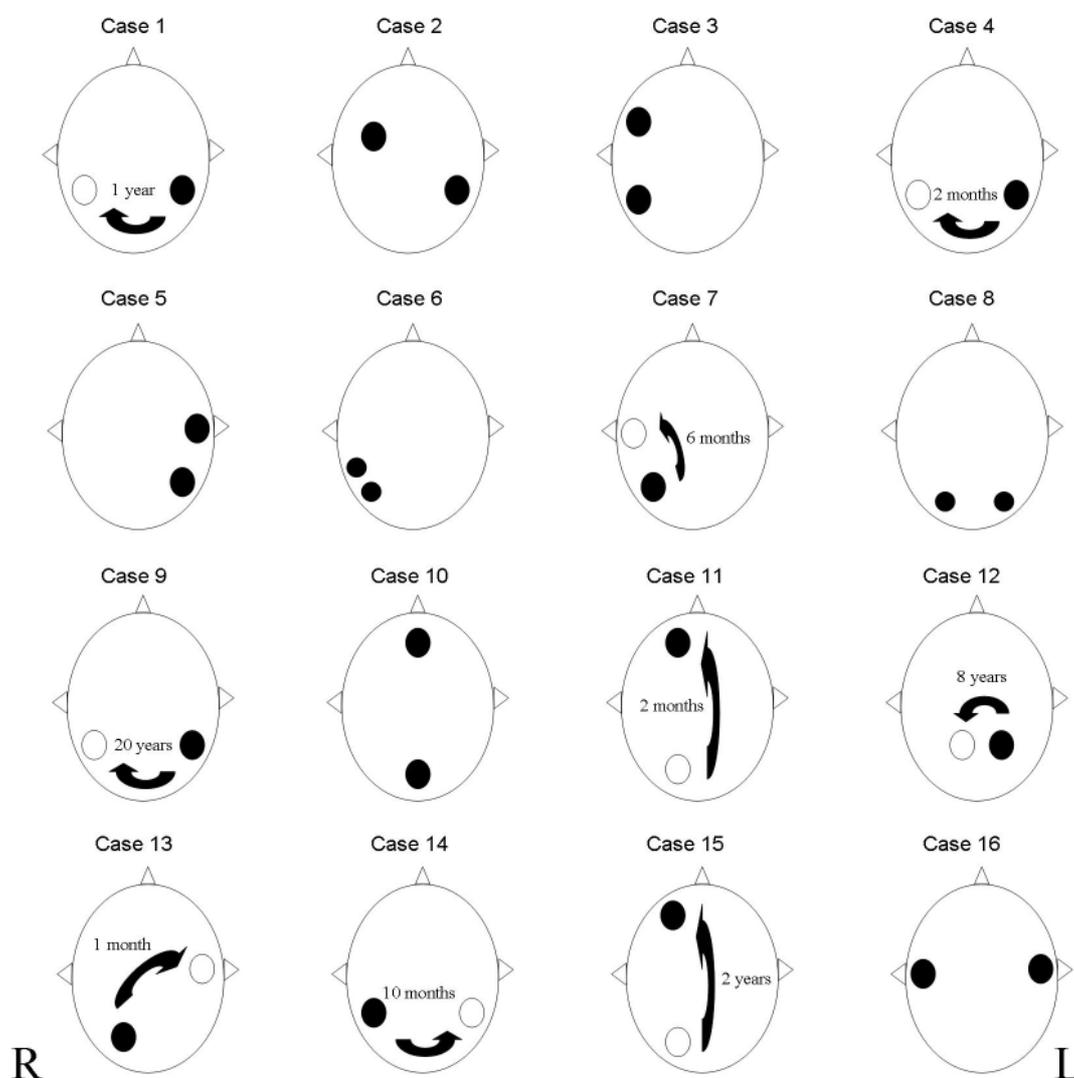


Figure 1. Distribution of bifocal painful areas in the current series and in previously reported patients in literature. R, right side; L, left side

challenge the previous proposals. First, a symmetrical distribution of painful areas is frequently seen in both type I and type II patients. Second, the shape and size of painful areas in type I patients, and between initial and additional painful areas in type II patients are consistent and similar. These findings not only argue for a generalized disorder for NH but also posit a reproduction of local process of epicranial neuralgia at multiple sites in the scalp.

Based on the current findings in bifocal NH, the proposal of this study is that a minority of trigeminal afferents ascend ipsilaterally to the thalamus and sensory cortex, regardless of nociceptive or non-nociceptive signal. This neuro-anatomic minority is responsible for the bilateral sensory disorder involving similar topography in a single lesion.^{10,11} The severity of sensory deficit provoked by the minority of afferents is usually equal to or milder than that of the majority. Thus, a direct involvement or central sensitization¹² in a single central site may be able to elicit a symmetric distribution of painful areas as seen in bifocal NH patients. This explanation is further supported by an unequal severity of pain in some type II bifocal NH patients. In fact, a central secondary sensitization can cause NH.^{5,13-15} The bifocal distribution of NH does not discredit the peripheral theory of NH⁵ but encourages a central role for NH.

Treatment guidelines for NH has not yet been established.⁵ In bifocal NH patients, abortive treatment like local lidocaine or analgesics, or preventive treatment, such as gabapentin, show favorable response. Heterogeneity of treatment strategies may suggest multiple cascades rather than a single pathway responsible for NH.

DISCLOSURE

Conflicts of interest: None

REFERENCES

1. Pareja JA, Caminero AB, Serra J, *et al.* Numular headache: a coin-shaped cephalgia. *Neurology* 2002; 58:1678-9.
2. Headache Classification Subcommittee of the International Headache Society: The International Classification of Headache Disorders, 2nd edition. *Cephalalgia* 2004; 24(Suppl 1):1-149.
3. Chen WH, Chen YT, Lin CS, Li TH, Lee LH, Chen CJ. A high prevalence of autoimmune indices and disorders in primary nummular headache. *J Neurol Sci* 2012; 320:127-30.
4. Pareja JA, Montojo T, Alvarez M: Nummular headache update. *Curr Neurol Neurosci Rep* 2012; 12:118-24.
5. Chen WH. Nummular headache - A review of world literature [Chin]. *TSMH Med Nurs J* (in press)
6. Ruscheweyh R, Buchheister A, Gregor N, Jung A, Evers S. Nummular headache: six new cases and lancinating pain attacks as possible manifestation. *Cephalalgia* 2010; 30:249-53.
7. Rocha-Filho PA. Nummular headache: two simultaneous areas of pain in the same patient. *Cephalalgia* 2011; 31:874.
8. Cuadrado ML, Valle B, Fernández-de-las-Peñas C, Barriga FJ, Pareja JA. Bifocal nummular headache: the first three cases. *Cephalalgia* 2009; 29:583-6.
9. Guerrero AL, Cuadrado ML, García-García ME, *et al.* Bifocal nummular headache: a series of 6 new cases. *Headache* 2011; 51:1161-6.
10. Chen WH, Lin HS, Chui C, Wu SS, Lui CC, Yin HL: Clinical analysis of cheiro-pedal syndrome. *J Clin Neurosci* 2012; 19:991-5..
11. Chen WH, Lan MY, Chang YY, Liu JS, Chou MS, Chen SS. Bilateral cheiro-oral syndrome. *Clin Neurol Neurosurg* 1997; 99:239-43.
12. Latremoliere A, Woolf CJ. Central sensitization: a generator of pain hypersensitivity by central neural plasticity. *J Pain* 2009;10:895-926.
13. Alvaro LC, García JM, Areitio E. Nummular headache: a series with symptomatic and primary cases. *Cephalalgia* 2009; 29:379-83.
14. Guillem A, Barriga FJ, Giménez-Roldán S. Nummular headache associated to arachnoid cysts. *J Headache Pain* 2009; 10:215-7.
15. Guillem A, Barriga FJ, Giménez-Roldán S. Nummular headache secondary to an intracranial mass lesion. *Cephalalgia* 2007; 27:943-4.