Introduction

Nummular headache (NH) is a well-defined clinical condition characterized by local pain that occurs in a small (typically 1–6 cm in diameter), rounded or elliptical area of the head. It was the first described by Pareja et al.\(^1,2\). It was also included in the appendix of the International Classification of Headache Disorders (ICHD) in 2004.\(^3\) However, in the ICHD 3 beta (2013),\(^4\) it is well-placed in group 4 in Part 1 among other primary headaches. NH is characterized by circumscribed areas of mild to moderate head pain which is usually chronic and continuous but which can also be remitting and relapsing. Superimposed on the continuous pain may be associated lancinating jabs of pain which may vary from seconds to hours. The affected area may also have paresthesias, dysesthesias, and allodynia even during periods of remission. The most characteristic feature of the NH is the presence of a well-defined area of pain that is typically circular or oval in shape. The size of the affected area is usually 2-6 cm in diameter. The pain is often described as burning or tingling and may be associated with numbness or tingling sensations in the affected area. The pain may be chronic and continuous, or it may fluctuate in intensity and duration. The location of the pain is typically in the parietal or occipital region of the head, but it can also occur in other areas such as the temple or forehead. The pain may be triggered by a variety of factors, including stress, fatigue, or exposure to certain environmental stimuli. The pain is often relieved by application of heat or cold to the affected area. The diagnosis of NH is based on the presence of a well-defined area of pain that meets the criteria for a primary headache disorder, as defined by the ICHD. Treatment of NH typically involves the use of nonsteroidal anti-inflammatory drugs (NSAIDs) or other analgesic medications, as well as the use of preventive treatment strategies to reduce the frequency and severity of attacks. The use of preventive medications is often necessary to control symptoms, as the pain of NH can be severe and disabling. In some cases, surgery may be considered as a treatment option, depending on the severity and duration of symptoms. Overall, the prognosis for NH is good, with most patients experiencing significant improvement with appropriate treatment.
of NH is its precise localization over a localized area over the scalp. The most common area described is the parietal area although other areas as well as bilateral and multifocal involvement have been described. It is a rare entity and only a limited number of patients have been described worldwide. Even though it is considered as a primary headache syndrome, NH has also been described in patients with intracranial lesions. Hence, we have attempted to describe patients with NH in a population in South India to further elucidate the epidemiological aspects of this uncommon entity and throw light on the different variations in its clinical presentation.

Materials and Methods

Patients attending the medicine and neurology outpatient departments of a tertiary referral hospital (Government T. D. Medical College Hospital, Alappuzha) in South India were studied over 30 months from January 1, 2013, to June 30, 2015. Those patients diagnosed to have NH were identified as per the criteria laid down in the ICHD 3 beta version. A detailed history was taken about headache emphasizing the time of onset and course, whether it was chronic, persistent, progressive, or remitting-relapsing as well as the nature of pain. The specific details about headaches included exact location of headache, side of headache, size and shape of the affected area, number of foci, quality of headache, and intensity of headache as noted by the visual analog scale (VAS). The presence of any other type of headaches was noted meticulously. A history of any other neurological disease was also taken, and a detailed neurological examination was performed. In addition, a meticulous inspection and palpation of the scalp were done specifically to look for autonomic changes such as color, vasomotor, and sudomotor changes as well any local area of tenderness. Scalp arteries including temporal and frontal arteries were palpated to assess pulsation. After this, the patient was asked to outline the affected area, and special emphasis was given to inspection and palpation of the symptomatic area. The Hamilton Depression Scale was completed by all patients. X-ray studies of the cervical spine were performed in selected patients. Neuroimaging studies with either computed tomography scan (14 patients) or magnetic resonance imaging (15 patients) of the head were completed in all patients. Patients detected to have either local structural lesion of the skull or scalp or intracranial lesions which could have clinically caused a secondary headache were excluded from the study. Complete blood counts, erythrocyte sedimentation rates, standard biochemical tests, thyroid functions tests, and a baseline vasculitic workup consisting of rheumatoid factor, antinuclear antibody, and C-reactive protein were done.

Results

A total of 29 patients identified as per the ICHD 3 beta criteria were obtained in our study. There were 19 females (65.52%) and 10 males (34.48%). The mean age of our patients at presentation was 47.62 years (standard deviation [SD]: 11.90). Sixteen (55.17%) cases were above 50 years of age while 7 (24.14%) were above 40 years of age. The ages varied from a minimum of 32 years to a maximum of 69 years. The duration of headache varied from 3 months to 5 years with a mean of 24.17 months (SD - 15.69). The site of headache was predominantly in the parietal area 15 (51.72%), followed by the occipital area 7 (24.13%). No definite side preponderance could be noted between the right and left sides (52% vs. 48%). All of the patients in our series proved to have unifocal areas of headache. The mean diameter of the affected area was 3.23 cm (SD - 0.26). In the majority of cases, i.e. 22 (75.86%), the area was described as circular, being elliptical in the remainder. In most of our patients, the intensity of headache proved to be mild to moderate with a mean VAS score of 5.03 (SD - 0.790). The quality of pain was most commonly felt as burning dysesthesia 12 (41.38%), followed by a feeling of pressure 10 (34.48%). Superimposed lancinating jabs of pain in the same area were reported by 10 (36%) of patients. In the majority of patients, i.e. 21 (72.41%), pain was chronic and continuous; however, in more than half, i.e. 15 (51.72%) of these patients, there were fluctuations in the severity of pain, sometimes such as to superficially resemble a relapsing-relapsing pattern. None of the patients had any significant trophic change even though paresthesias, dysesthesias, and allodynia were reported by nearly one-third of patients, i.e. 9 (31.03%). Only one (3.45%) patient proved to have depression as per the Hamilton Depression Rating Scale (Score-24). Regarding the clinical history of other illnesses, only one patient (3.45%) gave a history of head injury. Ten out of 29 patients (34.48%) had other types of concurrent headaches, the majority of which proved to be migrainous, i.e. 7 (24.14%), 2 patients (6.89%) with tension headache, and 1 patient (3.45%) with trigeminal neuralgia. Neurological examination was normal in all patients. None of the cases in our series showed any neuroradiological abnormality, except for the one patient with a history of head injury who showed a chronic subdural hematoma. Laboratory findings were also normal in all patients.
Discussion

NH is defined as per the ICHD, 3rd edition (beta version).[4] a. Continuous or intermittent head pain fulfilling criterion B b. Felt exclusively in an area of the scalp, with all of the following four characteristics: 1. Sharply contoured 2. Fixed in size and shape 3. Round or elliptical 4. 1–6 cm in diameter. c. Not better accounted for by another ICHD-3 diagnosis.

NH is an uncommon entity. Only a limited number of cases have been described in the international literature. Pareja et al. in a prospective study calculated the incidence to be 6.4/100,000/year and concluded that it is a noninfrequent primary headache.[5] Even though infrequently described in the literature, the true incidence likely exceeds this as the condition is mostly unrecognized as many patients with NH may not seek medical help due to the mild/moderate pain intensity. In addition, the condition may have been either ignored or shown only to general practitioners without neurology referral due to its relatively benign nature. Further detailed epidemiological studies to assess the prevalence and incidence in the general population are required as ours was not a community study. Moreover, being a noncomputerized government hospital in a resource-poor country, the total number of cases of headaches due to all causes attending the hospital outpatient department during this period is not available. In view of its relative rarity, for this study, we compared the epidemiological and clinical features of our patients with those appearing in the literature review of all published cases of NH by Dai et al.[6] [Table 1]. Those authors studied a total of 238 cases from the 30 published English studies from 2002 to 2012 and had comprehensively described their clinical and demographic characteristics.

Our study confirmed the female preponderance of NH as found in the previous studies. Similarly, NH proved to be an affliction of the middle-aged as found in all previous studies. The mean duration of headache in our study was 24.17 months which is shorter than the figure of 6.4 years available from the published English literature.[6] We postulate that a larger study extending for a longer period might well reduce this discrepancy. Regarding the clinical features, the location (parietal) and diameter and shape (circular) proved to be similar to that described in the international literature. The severity of pain as assessed by the VAS was also comparable being mild to moderate. In all our patients, pain was strictly unifocal. Similar to this in the above literature survey, headache was unifocal in the vast majority of patients (93%), being bifocal in 6% and multifocal in 0.5% only. None of our cases showed trophic changes in the affected area. However, this appeared to be rare even in the cases reviewed worldwide, being only 5%. In 72.41% patients, pain was chronic and continuous since its development in comparison to the 65% reported in the literature review. This is not surprising as NH is essentially a chronic disease. In fact, the ICDH 3 beta version[4] itself states that “in up to 75% of published cases, the disorder has been chronic (present for longer than 3 months), but cases have also been described with durations of seconds, minutes, hours, or days.” However, the nature and quality of pain proved to be different in our study; the most common type was a burning type of paresthesia, in contrast to the nearly half of patients in the literature review having a sense of pressure as their painful symptom. This pressing type of pain proved to be the second most common type of pain in our series. Another difference noted was in the presence of concurrent headache types. Concurrent headaches were present in 10 (34.48%) of our cases as opposed to a lower rates of 13% in the literature review. Moreover, in our series, the concurrent headaches proved to be

| Variable | Wei Dai et al. | Rammohan et al. |
|----------|---------------|-----------------|
| Female-male ratio | 1.5:1 | 1.9:1 |
| Mean age at reporting (years) | 47.5 | 47.62 |
| Mean headache duration (months) | 76.8 | 24.17 |
| Longest duration reported (years) | 50 | 5 |
| Localization-parietal area (%) | 44 | 52 |
| Right-left occurrence (%) | 56 (right) - 44 (left) | 52 (right) - 48 (left) |
| Bifocal involvement (%) | 6 | 0 |
| Multifocal involvement (%) | 0.5 | 0 |
| Shape of affected area (circular) (%) | 81 | 75.86 |
| Mean diameter of affected area (cm) | 3.6 | 3.1 |
| Quality of pain (%) | Pressing type | Burning paresthesia |
| Mean visual analog score | 5.53 | 5.23 |
| Trophic changes (%) | 4 | 0 |
| History of prior head injury (%) | 4 | 3.44 |
| Concurrent headaches (%) | 13 | 34.48 |
| Concurrent migraine headaches (%) | 11 | 24.14 |
| Chronic continuous pattern (%) | 65 | 72.41 |
| Exacerbations (%) | 43 | 34.48 |
| Sensory disturbances (dysesthesia, allodynia) (%) | 56 | 31.03 |
divided between migraine and tension headaches in an approximate ratio of 4:1. Migraine headaches constituted only 24% of concurrent headaches in our study unlike the overwhelming preponderance of migraine headaches; 11% out of 13% in the literature review. This aspect needs further study to elucidate whether it is only an epidemiological difference between different populations or could be accounted for etiopathological causes such as different trigeminovascular activation patterns in different patients.

Some authors have considered the possibility of psychogenic factors in the etiology of NH; however, we could not find any evidence for the same. There was no significant relation between the Hamilton Depression Rating Scale and the occurrence of NH in our patients. This is consistent with the previous studies which have also not found any psychological deficit in patients with NH. Moreover, it is usually considered that strictly unilateral head pain has an organic cause. However, the possibility that episodes of preexisting NH may be triggered by stress or psychological factors is likely and requires evaluation by further studies.

Only one patient (3.45%) in our series had a history of head injury similar to the previously described rates of 4% in the literature. In our study, depression and prior head injuries did not have any significant role in the pathogenesis of NHs. Another topic of contemporary interest regarding the etiology of NH is whether the origin is central or peripheral. Pain of NH was classically considered to be originating peripherally from epicranial tissues such as terminal branches of cutaneous nerves. In favor of the peripheral etiology, theory is the fact that NH is localized to a precise area of the scalp with local tenderness and sensory disturbances such as paresthesias and allodynia. Moreover, the pressure pain threshold is reduced in the affected area and botulinum toxin type A gives effective relief of pain. Finally, trophic changes in the affected area of the skin have been described in some patients. Against, these are several points that favor a central etiology. For instance, patients have been described with bifocal and multifocal NHs as have patients in whom the painful area extends over the midline of the scalp, which would not be expected in cases with peripheral etiology. Similarly, infiltration of local anesthetics did not relieve pain in the majority of cases. A few interesting case descriptions such as large diameter NHs with bitrigeminal hyperalgesia and a case of NH in which pain recurred even after focal scalp resection of the affected area have also favored a central causation of NH. In this regard, it is interesting that one of our patients also had preexisting trigeminal neuralgia. Cupka and Login have described a case of trigeminal neuralgia evolving into NH over the same area, but this has not been described in other literature to date. With this said, we are of the view that the majority of clinical features in our series favor a peripheral etiology as we could not obtain any bifocal case or case in which pain extended across the midline of the scalp or demonstrated trophic changes in the skin of the affected area. Similarly, we could not find any evidence to suggest a cervicogenic headache even when the painful area extended to the occipital area. This could indicate that central pain sensitization due to diseases such as trigeminal neuralgia or migraine could facilitate the development of NHs in some way. This could explain the higher occurrence of other primary headaches in our studies as compared to the international literature. This is also in agreement with authors such as Rocha-Filho who suggest that even though NH is a peripheral epicranial neuropathy, its expression is facilitated by other central or peripheral predisposing factors. Authors including Chen et al. have also proposed that a minority of trigeminal afferents arise ipsilaterally to the thalamus and sensory cortex regardless of nociceptive or nonnociceptive signals and that this neuroanatomic minority is responsible for pain of NH.

Conclusion

Our study proves the existence of the newly described primary headache syndrome called NH in South Indian population. To the best of our knowledge, there have been no other systematic studies investigating NHs in India. In comparing our results with the international literature, the number of similarities is much greater than the differences. The differences included a significantly increased occurrence of concurrent headaches, a different type of pain character (burning paresthesia), and a lesser overall duration of the disease in our study. In our study, depression and prior head injuries did not have any significant role in the pathogenesis of NHs unlike that seen in the international literature. The etiology of pain in our series appeared to be primarily peripheral with a role for central pain sensitization in some cases due to a variety of concurrent central causes of head pain with possible trigeminovascular activation such as migraine, tension headaches, and trigeminal neuralgia.

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Conflicts of interest
There are no conflicts of interest.
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