Sphenopalatine Neuralgia Should be Retained as an Independent Diagnostic Entity

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DESCRIPTION

The term “Sphenopalatine Neuralgia (SN)" first named by Sluder, defines a clinical disorder characterized by pain in one side of the head and face, with ipsilateral eyelid and nasal mucosa congestion, runny nose and tears and other autonomic nervous symptoms [1]. Then, cases were reported one after another, and most of them were reported by otolaryngologists [2-3]. However, due to the characteristic manifestations which has many overlaps with the characteristics of cluster headache (CH), the use of this diagnosis has been in a confused state. Some views, especially those from neurologists, consider SN to be a subtype of CH, or even that the diagnosis is simply a different name for the same symptom used by otolaryngologists and neurologists [1-3].

SN is not classified as an independent disease in the International Classification of Headache (ICHD). There is no mention of SN in the second edition of ICHD, but there are some neuralgias which have not been fully verified clinically in classification 13.19, and SN may be classified into this category [4]. In the third edition, SN was mentioned as the “previously used term” of CH [5]. However, if we use the current ICHD classification method to consider SN as a CH, it can be noted that in the published cases, although there is some overlap between the clinical manifestations of SN and CH, it does not completely fit the diagnostic criteria, and the corresponding treatment effect and prognosis are different [6].

Oomen KP had published an article in cephalalgia that summarized the differences between SN and CH, they thought that SN is an independent diagnostic entity, and they believed that SN should be another Trigeminal Autonomic Cephalalgia (TAC) that different from CH [7]. Our study also supported that SN and CH have different clinical manifestations and therapeutic effects. What’s more, we prefer SN to be a neuralgia entity that involving the sphenopalatine ganglion rather than as a TAC.

We reported seven patients, among them six cases were secondary to structural lesions surrounding the ipsilateral sphenopalatine ganglion, and all of them experienced significantly clinical improvements after removing their primary causes. In addition, in our literature review, sixty-seven percent of patients with SN had structural lesions around the sphenopalatine ganglion [8]. According to the anatomical characteristics of the sphenopalatine ganglion, we are more inclined to believe that SN is a neuralgia. This further suggests that this entity is different from CH. CH is widely known to be the activation of hypothalamus, which then secondary activates the trigeminal autonomic reflex, possibly through the trigeminal-hypothalamic pathway [9]. Hence, CH is central encephalopathy. SN is regard as peripheral origin and could be caused by potentially damaged of sphenopalatine ganglion [10]. According to the precise positioning of the anatomical structure and the pathogenesis, Sluder’s proposed “sphenopalatine neuralgia” is more rigorous and scientific. Therefore, we proposed that SN should be considered as a neuralgia entity, and it should be classified as primary and secondary, like other neuropathic pain in the Classification 13 of ICHD-III.

The primary SN is a type of headache that cannot be classified as TAC but which cannot find the cause. As confirmed above, it is different from CH and does not fully fit with other current diagnoses of TACs, such as responding to specific treatments such as high flow oxygen and indomethacin. Nevertheless, it responds to medications for neuralgia such as carbamazepine and pregabaline. Its pathological mechanism may be similar to that of trigeminal neuralgia, which is related to demyelination sphenopalatine ganglion.

The secondary SN is caused by peripheral structural lesions that affecting the sphenopalatine ganglion, which may include secondary or symptomatic TACs that mentioned in the previous literature. There is no uniform nomenclature before. Regarding secondary headaches in the International Headache Classification, that provides such a diagnostic classification: 11.9 Headache attributed to other disorder of cranium, neck, eyes, ears, nose, paranasal sinuses, teeth, mouth or other facial or cranial structures. This classification is considered to be the causal link between the potential obstacles and head or facial pain has been clearly established. However, it’s difficult in clinical practice. Many patients had no other symptoms before...
the onset of headache, and headache was the only clinical manifestation. Therefore, in clinical reasoning, we cannot make a diagnosis at the beginning according to 11.9. Instead, we could make a final diagnosis only after we find the underlying etiology in the development of the disease and the treatment is effective according to the etiology. But how do we make diagnosis when the patient visits?

Hence, as clinicians, we need a clinic reasoning picture. A diagnosis of sphenopalatine neuralgia may be considered in patients with lateral locking headache with unilateral autonomic nervous symptoms. Then secondary lesions need to be excluded, especially the structures around the sphenopalatine ganglion, and brain MRI and sinus CT and dental fragments are extremely necessary. If such headache does not fully meet the criteria for TACs after the exclusion of secondary causes, a diagnosis of primary SN may be considered. However, it is worth mentioning that strictly unilateral headache is always a red flag. Therefore, for such patients, whether the diagnosis is TACs or SN, the thinking is the same, we first need to rule out secondary causes. TACs are primary headache, and it is possible to ignore potential causes when it is diagnosed as possible TACs. We once met a patient who fulfilled criteria A to D of hemicrania continua, and the imaging examination was normal at the first visit, at follow-up one month later, revealed a neoplasm in the nasopharynx by the second brain magnetic resonance imaging [11].

Therefore, SN should not be classified as the former name of CH or as another TAC. Its clinical manifestations do not conform to the attack time and attack pattern of CH or other TACs. In addition, the clinical incidence of SN does not appear to be less than that of CH. We believe that the diagnosis of retained SN is helpful for clinicians’ clinical reasoning.

REFERENCES
1. Sluder G. Role of the sphenopalatine (Meckel’s) ganglion in nasal headaches. N Y State J Med. 1908; 87:989-990