CLINICAL CORRESPONDENCE

Occipital nerve stimulation for chronic cluster headache and hemicrania continua: pain relief and persistence of autonomic features

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The trigeminal autonomic cephalgias (TACs) and hemicrania continua are primary headache disorders characterized by pain in a unilateral trigeminal distribution that occurs in conjunction with prominent ipsilateral cranial autonomic features (1). Cluster headache, paroxysmal hemicrania and short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) are classified as TACs, while hemicrania continua is considered a separate primary headache disorder. The autonomic features and first (ophthalmic) division pain have been assumed to be interdependent. Autonomic features have been considered the result of activation of first-division nociceptive afferents, which, through a functional brainstem connection between the trigeminal nucleus caudalis and superior salivatory nucleus, leads to activation of parasympathetic efferents (2). However, this interdependence has been challenged by reports of patients with typical cluster headache without autonomic features, patients with autonomic features without cephalgia, patients with continuing autonomic features without pain after trigeminal root section and patients who report the appearance of autonomic features prior to onset of pain (3–6). Herein we report one patient with chronic cluster and one with hemicrania continua who experienced persistence of autonomic features in the absence of head pain after placement of an occipital nerve stimulator.

Case reports

Case 1

A 35-year-old man was referred to our institution for evaluation of a 5-year history of chronic cluster headaches. The patient reported episodes of severe, exclusively right-sided, retroorbital and frontal pain associated with agitated behaviour and prominent autonomic features including periorbital oedema, ptosis, lacrimation, conjunctival injection, rhinorrhoea and nasal stuffiness. The pain was said to peak rapidly and last between 30 min and 120 min. Attacks occurred with a variable frequency, but up to three times per day. Episodes most often occurred between 12.00 h and 02.00 h. Headaches and medication side-effects had resulted in disability from work, financial difficulties and deficits in concentration and memory. He had no family history of cluster or migraine headaches.

He had tried multiple preventative medications including indomethacin, verapamil, topiramate, gabapentin, lithium, prednisone and valproic acid. Abortive medications had included high-flow oxygen, triptans and dihydroergotamine. Subcutaneous sumatriptan had caused intolerable side-effects and high-flow oxygen was ineffective. Intramuscular injections of dihydroergotamine were effective in reducing severe pain. At the time of evaluation, he was taking daily oxycodone, as well as benzodiazepines and dihydroergotamine as needed. None of these medications had altered his headache frequency. The patient’s neurological and imaging evaluations, including magnetic resonance imaging of the brain and magnetic resonance angiography, were normal.

A right-sided greater occipital nerve stimulator (Bion; Advanced Bionics, Sylmar, CA, USA) was implanted in an attempt to ameliorate this patient’s long-standing disabling and medically refractory cluster headaches. He was placed on continuous mode stimulation with a pulse width of 200 µs and frequency of 45 Hz. The patient stimulated at an
average of 3.6 mA, although he would increase it to 4.6 mA with severe headaches. He reported a 100% improvement in quality of life, a 70% improvement in headache frequency, duration and intensity, and a significant reduction in usage of acute pain medications. However, he was frequently suffering from episodes of right-sided lacrimation, conjunctival injection and rhinorrhea in the absence of both head pain and agitated behaviour. He considered these symptoms and signs to be identical to those that had occurred with his usual preoperative cluster headaches.

Case 2

A 44-year-old woman was referred to our institution for evaluation of medically intractable hemicrania continua. Twelve years previously she fell to the ground suffering head trauma and brief loss of consciousness. When she awoke, she instantaneously noted head pain in the left occiput with radiation up and over the left ear, terminating in the preauricular region. The pain, which she described as ‘drill-like’, was constant and continuous. Four to five times per week she developed painful exacerbations lasting 30 min to 4 days with an average duration of several hours. With severe pain she developed ipsilateral ptosis, lacrimation and nasal congestion. Severe pain was often preceded by a feeling of sand in her left eye. The patient’s past medical history was significant for infrequent non-disabling headaches since the age of 19. There was no family history of headache.

Previous preventative medications had included indomethacin, verapamil, nortriptyline, lamotrigine, duloxetine, fluoxetine and bupropion. Indomethacin 225 mg/day decreased the intensity of her background headache from a 7/10 to a 2/10 and the frequency of severe exacerbations. However, indomethacin was discontinued secondary to abdominal pain, dizziness and nausea. Otherwise, she was unresponsive to preventative medications. Her current medications included daily gabapentin, as well as p.r.n. tramadol, oxycodone/acetaminophen and frovatriptan. Neurological examination and brain imaging studies were normal.

A left occipital nerve stimulator was placed (Bion; Advanced Bionics). The stimulator was set to continuous mode, pulse width of 300 µs and frequency of 45 Hz. She stimulated over an amplitude range from 3 mA to 7 mA. The patient had significant improvement postoperatively in terms of the frequency and severity of her headaches. She became pain free at baseline with superimposed severe headaches occurring only five times over a 3-month period. She was able to discontinue gabapentin and significantly decrease the use of acute pain medications. However, on six occasions she had episodes of lacrimation, nasal stuffiness and a feeling of sand in her left eye without the development of head pain. These symptoms were identical to those associated with her usual prestimulator pain exacerbations.

Discussion

These cases illustrate and reinforce two important points. Occipital nerve stimulation (ONS) may be an effective treatment modality for a variety of refractory primary headache disorders. In addition, the expression of cranial autonomic features is likely to be under the control of a rostral diencephalic pacemaker and, therefore, may be an accompanying rather than simply a secondary feature of ophthalmic-division pain.

ONS has been reported to be an effective treatment modality for chronic migraine (7). In a previous report, it was suggested that ONS may also be effective for a variety of other refractory primary headache disorders, including hemicrania continua and chronic cluster headache (8). While the mechanism underlying the analgesic effect of ONS is not clear, mobilization of central pain-modulating centres has been suggested by functional imaging studies (7). In addition, ONS may also have a direct effect on activity of nociceptive neurons within the trigeminal nucleus caudalis. Stimulation of the greater occipital nerve results in increased metabolic activity in the trigeminal nucleus caudalis and cervical dorsal horn at the C1 and C2 levels (9). There is clear evidence of coupling between meningeal nociceptive afferents and sensory afferents in the greater occipital nerve (10). Individual stimulation of both the dura and the greater occipital nerve results in activation of neurons in the C2 spinal dorsal horn. These neurons have receptive fields corresponding to the ophthalmic division of the trigeminal nerve as well as skin and muscle from C2 (10).

The pathophysiological mechanisms underlying the genesis, pain and cranial autonomic features of cluster headache and hemicrania continua may be similar (11) The hypothalamus has been implicated as the central generator of both cluster headache and hemicrania continua, while the pain and autonomic features are felt to be secondary to activation of cranial parasympathetic efferents and first-division nociceptors (12–14). However, dependence on nociceptive activation of the trigeminal nerve for parasympathetic activation has been questioned by
reports of typical autonomic features without head pain, autonomic features that precede pain, and continuation of autonomic features after trigeminal root sectioning. The patients presented in this report serve as further evidence for dysfunctional control of superior salivatory nucleus (SSN) activity from a diencephalic pacemaker. The SSN, located dorsolateral to the facial nucleus in the brain stem, participates in the control of a number of physiological functions including lacrimation, cerebrovascular tone, nasal blood flow and nasal secretion. Activity of SSN neurons themselves is regulated by afferent inputs from many areas of the brain, including the dorsomedial and paraventricular hypothalamic area (15, 16).

The relief of pain in these cases illustrates the potential for ONS as an effective treatment modality for medically recalcitrant cluster headache and hemicrania continua. The persistence of cranial autonomic features in the absence of pain also reinforces previous suggestions that the underlying generator for the TACs and hemicrania continua may be similar.

References