

Can cluster headache occur in the occipital region?

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Abstract

Cluster headache is typically observed in the orbital, supraorbital or temporal regions, or in a combination of the above. The site of pain is one of the diagnostic criteria for cluster headache included in the International Classification of Headache Disorders. It has been reported that pain may also occur in the ear, nose, neck, shoulder, and other regions. In this report, we present a case who presented exclusively with occipital pain attributable to probable cluster headache. In the presence of other typical features of cluster headache, the diagnosis should still be considered even when headache pain occurs in extratrigeminal locations.

Keywords: Cluster headache, trigeminal autonomic cephalalgias, occipital region, treatment, greater occipital nerve block, pulsed radiofrequency.

INTRODUCTION

Cluster headache (CH) is the most common trigeminal autonomic cephalalgia, a group of primary headaches characterized by unilateral, short-term, excruciating attacks of pain with cranial autonomic symptoms and/or feelings of restlessness. It typically occurs in the orbital, supraorbital or temporal regions, or in a combination of these regions.¹ CH pain also may occur in atypical locations.² In this report, we present a case who presented with pain only in the occipital region that was best characterized as a probable CH.

CASE REPORT

A 62-year-old female was seen at our outpatient clinic with a complaint of pain in the right occipital region for 8 years. The patient described a stabbing pain with a numerical pain rating of 10/10. The pain occurred 3-4 times a day on average and usually lasted about 10 minutes. It occurred in periods lasting 2-3 weeks with pain-free periods of 3-4 months. There was no pain in between attacks. In addition to pain, she described miosis, lacrimation, eyelid oedema, and rhinorrhea on the same side as the pain. She described intense restlessness during most of the attacks and was awakened from sleep. Nausea, vomiting, photophobia, phonophobia,

and osmophobia were absent. She had a history of using paracetamol, various non-steroidal anti-inflammatory drugs (indomethacin, naproxen sodium and dexketoprofen) as well as eletriptan, but had responded to none of them. Verapamil 120 mg daily had been prescribed for her pain at another center, but she discontinued the drug due to low blood pressure. Pulsed radiofrequency (PRF) treatment had been performed on the right greater occipital nerve (GON) at another center, but without benefit. At presentation she had been using topiramate 100 mg daily for 6 months without improvement. The patient had hypothyroidism and hypertension and was using levothyroxine sodium 100 mcg/day, losartan 100 mg/day, and hydrochlorothiazide 12.5 mg/day. The family history and neurological examination were unremarkable. Biochemistry tests, the hemogram, the erythrocyte sedimentation rate, and the C-reactive protein value were within normal limits. Contrast-enhanced cranial MR examination was unremarkable. Ultrasound-guided nerve block of the right GON was applied at the C2 level, using 1 ml compound betamethasone (2 mg betamethasone sodium phosphate and 5 mg betamethasone dipropionate) and 4 cc lidocaine 1%. The patient became pain-free on follow-up for 9 months after the injection.

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DISCUSSION

According to the International Classification of Headache Disorders (ICHD-3), a typical cluster attack is observed in the orbital, supraorbital, temporal, or a combination of these regions.¹ CH pain may also differ from the typical frequency or duration, or present with atypical symptoms and signs.³ Although particular pain locations are part of the ICHD-3 diagnostic criteria, pain can also be experienced in other locations.^{1,2}

Bahra *et al.*² evaluated 230 patients with CH and reported pain in the retro-orbital and temporal regions in 92% and 70% of the cases respectively. Pain also occurred in the ear, nose, neck, shoulder, and other hemicranial regions in addition to the trigeminal nerve distribution. Another study reported that 8 of 29 CH patients experienced occipital pain. In most of these patients, the headache extended from typical sites to the frontal, cervical, carotid, and parietal regions.⁴

Sanin *et al.*⁵ reported three patients with atypical pain localization with possible CH. The patients had pain only in the neck or occipital regions, and not in the trigeminal nerve distribution. No other study has reported CH with pain occurring outside the orbital, supraorbital, or temporal regions. Our patient only had occipital pain which did not involve the trigeminal nerve distribution. Our patient's headache met ICHD-3 criteria for probable CH because it met all but one diagnostic criteria, that of localization combined with duration of pain.¹

Block of the greater occipital nerve, a C2 nerve branch, interrupts the trigeminal autonomic reflex pathway and is effective in CH treatment. According to the American Headache Society, despite level A evidence for GON injection, there is no consensus regarding the correct dosage, drug, and frequency of the procedure.⁶ Moreover, the duration of effect after nerve block may differ. In a double-blind placebo-controlled study, no attacks were observed 1 week after injection in 11 patients in the group treated with GON block. Eight of these patients were relapse-free for 4 weeks and remission persisted between 4 and 26 months in five of these patients.⁷

The current patient's occipital pain and improvement after GON block suggest occipital neuralgia as a differential diagnosis. Patients with occipital neuralgia may occasionally report concurrent autonomic symptoms; however, these symptoms are not prominent.⁸ Pain in occipital neuralgia lasts from a few seconds to minutes,

and dysaesthesia and/or allodynia occur with innocuous stimulations of the head. There is tenderness over the affected area and trigger points in the GON or C2 distribution. The absence of the aforementioned criteria excludes the diagnosis of occipital neuralgia.¹

PRF treatment can prolong the pain-free period in patients who benefit from GON injection.⁹ In our case, PRF for GON was performed under fluoroscopy in another center, but was ineffective. In PRF, the nerve is subjected to sensory stimulation, and the patient is asked if pain or paresthesia occurs in the area supplied by the nerve. Thus, patient cooperation is important. After the procedure, local steroid is injected around the nerve.⁹ That our patient benefited from GON block when the previous PRF failed suggests that the nerve was not well-localized by fluoroscopy in the latter procedure. Localization by ultrasound guidance may therefore be preferable.

Successful CH treatment depends on a clear diagnosis. The diagnosis of CH should still be considered in patients with pain outside the orbital, supraorbital, and temporal regions if other CH diagnostic criteria are met.

DISCLOSURE

Availability of data: Additional data that support the findings of this study are available on request from the corresponding author.

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