**Occipital Neuralgia Secondary to C2 Spinal Cord Infarction**

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**Abstract:**
We herein report the first case of occipital neuralgia secondary to spinal cord infarction. A 74-year-old woman suddenly developed numbness and dysmetria in her right arm. Two days later, she developed a paroxysmal shooting pain in the right posterior part of the scalp three to five times per day. Magnetic resonance imaging revealed a hyperintense lesion in the right posterior column and dorsal root entry zone at the C2 level. The patient was subsequently diagnosed with occipital neuralgia secondary to spinal cord infarction. Diverse etiologies need to be considered in occipital neuralgia secondary to spinal cord lesions.

**Key words:** occipital neuralgia, spinal cord infarction, C2 spinal cord, dorsal root entry zone

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**Introduction**

Occipital neuralgia is characterized by unilateral or bilateral paroxysmal shooting pain in the posterior scalp and is distributed in the region of the greater, lesser, or third occipital nerves. Although the detailed pathophysiology remains uncertain, it is presumed to be caused by injury to the C2-3 nerve roots or occipital nerves; however, several cases of occipital neuralgia secondary to an upper cervical cord lesion, including inflammatory myelitis, multiple sclerosis, neuromyelitis optica, neurosyphilis, and cavernous angioma, have been reported (1-5).

We herein report a case of occipital neuralgia secondary to C2 spinal cord infarction. We propose that diverse etiologies should be considered in occipital neuralgia secondary to spinal cord lesions.

**Case Report**

A 74-year-old woman with a history of hypertension and hyperlipidemia suddenly presented with numbness and dysmetria in the right upper extremity. Two days later, she developed a paroxysmal shooting pain in the right posterior scalp. Each pain attack lasted a few seconds and occurred up to three to five times per day. Touching of the occipital region triggered pain. A neurological examination revealed reduced vibration sensation in the right hemi-body below the neck, allodynia in the right C2 dermatome, and right upper limb ataxia. When the patient was asked to grasp her left thumb with her right hand, with her eyes closed, she attempted the grasping movement up to 10 cm away from her left thumb. Therefore, the cause of her right upper limb ataxia was presumed to be proprioceptive impairment. Tenderness in the right occipital region was absent.

Three days later, although the paroxysmal occipital pain and allodynia in the right C2 region had disappeared, a similar pain attack occurred in the right retroauricular region. Touching of the right retroauricular region triggered pain. As the pain did not appear in the right auricle or angle of the mandible, great auricular neuralgia seemed unlikely. This retroauricular pain attack persisted for four days and then spontaneously disappeared. Magnetic resonance imaging (MRI) of the brain revealed no causative lesion. On MRI of the cervical spine, diffusion-weighted imaging showed a high signal at the spinal cord level of C2 (Figure A). An apparent diffusion coefficient map showed a low-intensity signal in the same region (Figure B). T2-weighted imaging showed a hyperintense lesion in the right posterior column, right dorsal root entry zone, and part of the right lateral column at the spinal cord level of C2 (Figure C, D). Magnetic resonance angiography revealed a normal appearance of the right vertebral artery and hypoplasia of the left vertebral artery (Figure E).

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Blood test results were as follows: anti-nuclear antibody, lupus anticoagulant, anti-cardiolipin antibody, anti-aquaporin-4 antibody, and syphilis antibody were negative; vitamin B12, folic acid, and angiotensin-converting enzyme (ACE) levels were normal. Cerebrospinal fluid test results were also normal. The IgG index was 0.42 (<0.7), while the oligoclonal band and cytology were both negative. Although local anesthetic block was not performed for the short-term episodes of pain, the patient was diagnosed with occipital neuralgia secondary to C2 spinal cord infarction.

Occipital neuralgia originating from the C2 spinal cord has been suggested as a complication of right retroauricular pain. Great auricular neuralgia is caused by a peripheral lesion in the great auricular nerve, which originates from the C2 and C3 nerve roots. This condition is characterized by auricular and mandibular pain attacks. As our patient presented with only retroauricular pain, the origin of the pain attack was presumed to be the C2 spinal cord.

The cause of spinal cord infarction appeared to be atherosclerosis of the posterior spinal artery. As the patient had vascular risk factors, clopidogrel was started to prevent the occurrence of other atherothrombotic events. Three-month follow-up MRI revealed a T2 hyperintense lesion localized to the right posterior column (Figure F, G). A causative relationship between occipital neuralgia and T2 hyperintensity in the right dorsal root entry zone has previously been suggested. Although mild hand weakness gradually disappeared within three months, right upper limb ataxia and sensory impairment in the right hemi-body persisted for over one year.

**Discussion**

While several cases of occipital neuralgia secondary to non-ischemic spinal cord disorders including inflammatory myelitis, neurosyphilis, and cavernous angioma have been reported, a case of occipital neuralgia secondary to spinal cord infarction has not yet been reported. The most frequent cause of occipital neuralgia is a posterolateral spinal cord lesion at the C2 spinal cord level. The low prevalence of ocipital neuralgia secondary to spinal cord infarction may be attributable to the rarity of spinal cord infarction in the posterior column at the C2 level (6). The collateral network might be more extensive and blood flow to the spinal cord...
occipital neuralgia and subsequent retroauricular pain. As vasogenic edema is a reversible change, it could also explain transient episodes of spinal cord infarction, cytotoxic edema, infection, inflammation, and compression, in the dorsal root entry zone may play a crucial role in the development of occipital neuralgia.

It is well known that dorsal root entry zone lesions are associated with the onset of neuropathic pain in occipital neuralgia and trigeminal neuralgia (9, 10). Direct impairment of the sensory afferent pathway or the impaired descending inhibitory pathway may be related to neuropathic pain with spinal cord origin (11). In the present case, although both the right dorsal root entry zone and right posterior column presented with T2 hyperintensity, only T2 hyperintensity in the right posterior column persisted. In the acute phase of spinal cord infarction, cytotoxic edema is concomitant with vasogenic edema. While T2 hyperintensity in the right posterior column reflects cytotoxic edema, T2 hyperintensity in the right dorsal root entry zone reflects vasogenic edema. Vasogenic edema peaks roughly one to two days after cerebral ischemia (12). Delayed vasogenic edema may explain the discrepancy between the onset of numbness and ataxia in the right upper extremity and that of occipital neuralgia and subsequent retroauricular pain. As vasogenic edema is a reversible change, it could also explain transient episodes of occipital neuralgia and subsequent retroauricular pain.

Conclusion

Although spinal cord infarction rarely occurs in the C2 posterior column, this unusual disorder should be considered in the differential diagnosis of acute-onset occipital neuralgia.

The authors state that they have no Conflict of Interest (COI).

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