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【 CASE REPORT 】

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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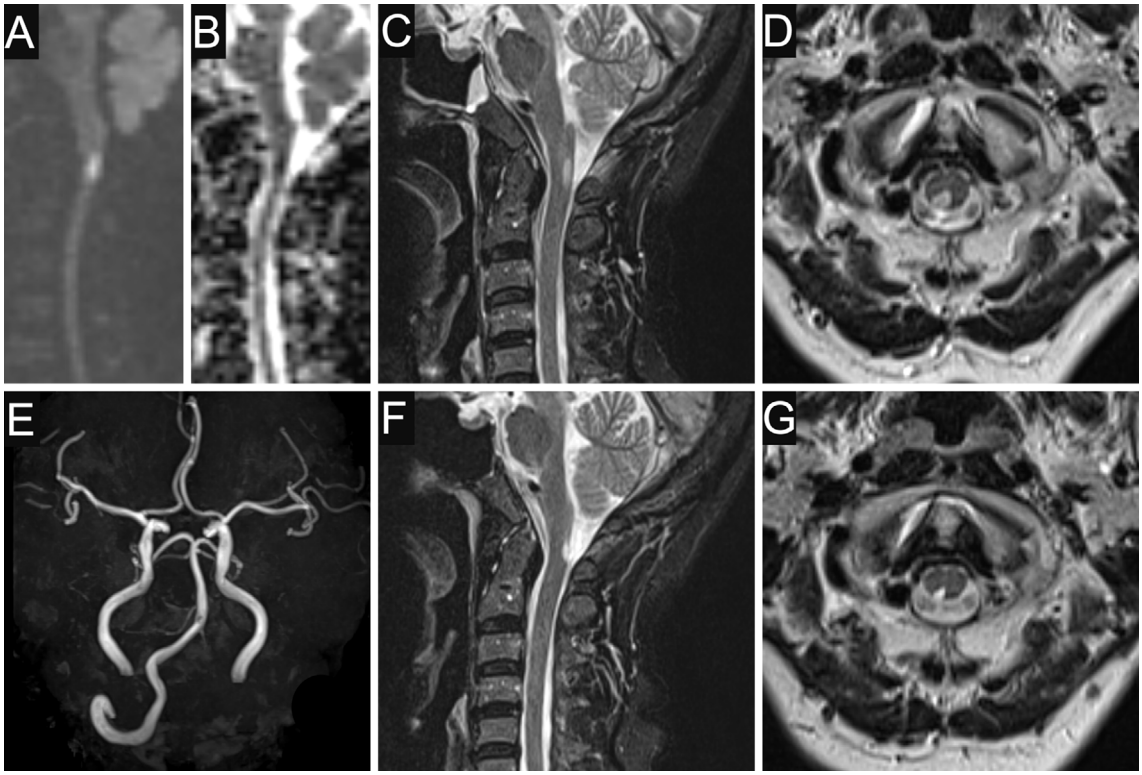

Figure. **Magnetic resonance imaging of the brain and cervical spinal cord. (A) Sagittal diffusion-weighted imaging showing a hyperintense lesion at the C2 spinal cord level. (B) A sagittal apparent diffusion coefficient map showing a hypointense lesion at the spinal cord level C2. (C) Sagittal T2-weighted imaging of the cervical spine showing a hyperintense lesion in the posterior part of the C2 spinal cord. (D) Axial T2-weighted imaging shows a hyperintense lesion in the right posterior column and dorsal root entry zone at the C2 spinal cord level. (E) Magnetic resonance angiography showing hypoplasia of the left vertebral artery. (F) On three-month follow-up T2-weighted imaging, the size of the T2 hyperintense lesion had decreased. (G) In the axial slice, T2 hyperintensity was strictly localized to the right posterior column.**

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 columns (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 occipital 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

more abundant in the region of the posterior spinal artery at the C2 level than at other spinal cord levels. Weidauer et al. reported 16 cases of spinal cord infarction, including 5 of spinal cord infarction involving the C2 cord (6). In all five of those cases, the infarction was localized to the anterior spinal artery territory, so the posterior column and dorsal root entry zone were spared from ischemia. None of the five patients presented with occipital neuralgia. Although two cases of spinal cord infarction involving the C2 posterior column have been reported, neither patient presented with occipital neuralgia (7, 8). One patient had an ischemic lesion in the unilateral C2 posterolateral column, and another had an ischemic lesion that was strictly localized to the unilateral C2 posterior column. Considering both the previously reported cases and the present case, less severe damage than occurs during post-ischemic cytotoxic edema, such as vasogenic 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).

References

1. Kissoon NR, Watson JC, Boes CJ, Kantarci OH. Occipital neuralgia associates with high cervical spinal cord lesions in idiopathic inflammatory demyelinating disease. Cephalalgia **39**: 21-28, 2019.
2. Cerrato P, Bergui M, Imperiale D, et al. Occipital neuralgia as isolated symptom of an upper cervical cavernous angioma. J Neurol **249**: 1464-1465, 2002.
3. Smith DL, Lucas LM, Kumar KL. Greater occipital neuralgia: an unusual presenting feature of neurosyphilis. Headache **27**: 552-554, 1987.
4. Boes CJ. C2 myelitis presenting with neuralgiform occipital pain. Neurology **64**: 1093-1094, 2005.
5. Hayashi Y, Koumura A, Yamada M, Kimura A, Shibata T, Inuzuka T. Acute-onset severe occipital neuralgia associated with high cervical lesion in patients with neuromyelitis optica spectrum disorder. Headache **57**: 1145-1151, 2017.
6. Weidauer S, Nichtweiss M, Lanfermann H, Zanella FE. Spinal cord infarction: MR imaging and clinical features in 16 cases. Neuroradiology **44**: 851-857, 2002.
7. Elzamly K, Nobleza C, Parker E, Sugg R. Unilateral upper cervical posterior spinal cord infarction after a neuroendovascular intervention: a case report. Case Rep Neurol Med **2018**: 5070712, 2018.
8. Wang CM, Tsai WL, Lo YL, Chen JY, Wong AM. Unilateral right occipital condyle to C2 level spinal cord infarction associated with ipsilateral vertebral artery stenosis and contralateral vertebral artery dissection: a case report. J Spinal Cord Med **34**: 118-121, 2011.
9. Nakashima I, Fujihara K, Kimpara T, Okita N, Takase S, Itoyama Y. Linear pontine trigeminal root lesions in multiple sclerosis: clinical and magnetic resonance imaging studies in 5 cases. Arch Neurol **58**: 101-104, 2001.
10. De Santi L, Monti L, Menci E, Bellini M, Annunziata P. Clinical-radiologic heterogeneity of occipital neuralgiform pain as multiple sclerosis relapse. Headache **49**: 304-307, 2009.
11. Svendsen KB, Sørensen L, Jensen TS, Hansen HJ, Bach FW. MRI of the central nervous system in MS patients with and without pain. Eur J Pain **15**: 395-401, 2011.
12. Dostovic Z, Dostovic E, Smajlovic D, Ibrahimagic OC, Avdic L. Brain edema after ischaemic stroke. Med Arch **70**: 339-341, 2016.