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Rare Cervical Intramedullary Cavernous Angioma with Trigeminal Neuralgia and Cervical Itch: Case Report and Review of the Literature

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Abstract

Introduction: Intramedullary cavernous angioma (ICA) is a rare lesion of the spinal cord, representing only 3% - 5% of central nervous system lesions. The coexistence of trigeminal neuralgia and refractory itch is very rarely encountered in clinical practice. To our knowledge, a report of an ICA with trigeminal neuralgia and local neuropathic itch has never been published to date. Thus, we present a very interesting case of a C2 ICA.
Case Presentation: A 61-year-old female presented with right facial pain for three years, which was exacerbated by accompanying cervical pain and itch for one month. The patient's symptoms were relieved after surgery, and there was no recurrence of lesions one year later.
Conclusions: ICA with trigeminal neuralgia and local neuropathic itch is very rarely encountered in clinical practice. As it is not always diagnosed at first, some patients miss the best treatment period. Therefore, we call for emphasis to be placed on early diagnosis and timely surgical treatment.

Keywords: Intramedullary Cavernous Angioma, Trigeminal Neuralgia, Neuropathic Itch, Surgical Treatment

1. Introduction

Neuropathic itch (NI) is of neuropathic origin and is produced by nerve system damage (1). Spinal itch has been associated with syringomyelia, tumor, spinal multiple sclerosis, and Brown-Sequard syndrome after traumatic injury (2). Intramedullary cavernous angioma (ICA)-caused itch is rarely encountered in clinical practice; hitherto, only a few cases have been reported (3). Trigeminal neuralgia (TN) is commonly caused by compression of the fifth cranial nerve in the pontocerebellar angle due to extra-axial pathologies (4, 5). Several studies (4, 6) have reported cases of TN caused by pontocerebellar angle cavernous angioma. However, only one case of ICA with TN has been reported (7). ICA with TN and NI, to the best of our knowledge, has never been reported to date. Here, we present the first case of ICA with TN and NI.

2. Case Presentation

A 61-year-old female was admitted to our department with complaints of right facial pain for three years with coexisting severe cervical pain and itch for one month. The onset of the right facial pain was June 10, 2010. The patient stated that the pain was not serious (verbal pain scale, VPS 2) and that she never needed any analgesics to relieve it. As the pain did not change, she paid no attention to it, and no imaging studies were performed until one month previously. New symptoms of right facial and cervical pain, as well as right cervical itch, developed on July 5, 2013. The pain (VPS 9) and itch were severe, and drugs (tramadol, carbamazepine, and antihistamines) did not relieve the symptoms. Physical examination showed only paresthesia of the right cervical area, with no abnormal observations of the limbs, the central nervous system, or the right cervical skin. Magnetic resonance imaging (MRI) of the head was performed one day after admission. Unfortunately, the head MRI showed that the right trigeminal nerve was compressed by the right posterior cerebellar artery (Figure 1). A trigeminal nerve microvascular decompression was planned; however, the right cervical pain and itch were not explained by the trigeminal nerve compression. After a group discussion involving the whole department, a cervical MRI was performed. The cervical MRI demonstrated a round intramedullary lesion located at the C2 level, exhibiting a core of increased signal intensity and surrounded by a low signal intensity rim on the T2-weighted images (Figure 2).

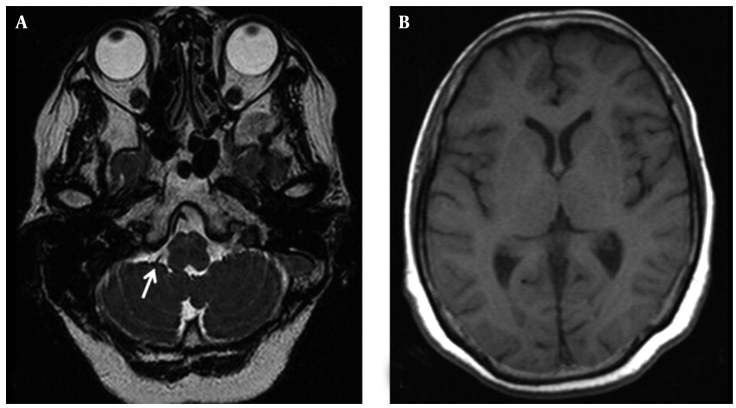

Figure 1. Head MRI showing the right trigeminal nerve compressed by the right posterior cerebellar artery (A), not by abnormal growth of the cerebral tissues (B).

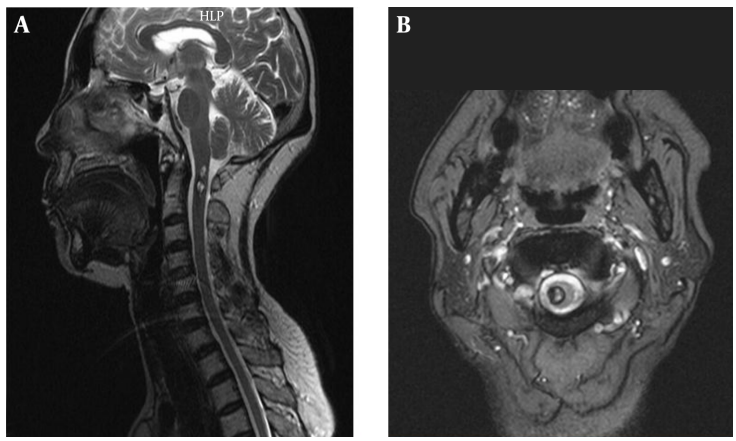

Figure 2. Cervical MRI showing a round intramedullary lesion located at the C2 level, exhibiting a core of increased signal intensity, surrounded by a low signal intensity rim on the sagittal T2-weighted images (A) and the axial T2-weighted images (B). These demonstrated that the lesion was located in the right dorsal horn.

A debate was held on which surgery should be performed first: microvascular decompression or intramedullary lesion resection. Considering that the right cervical pain and the itch were recent developments, we opted for the removal of the C2 intramedullary lesion first. After the preoperative examinations were completed, the patient was sent to the operating room. Microsurgical extirpation by posterior middle incision under general anesthesia was performed. After the skin and muscle were dissected, a laminectomy at the C2 level was performed under the microscope, assisted by neuroelectrophysiology. The dura was opened and an expanded cord a dark red lesion was clearly visible under the microscope (Figure 3). A myelotomy was performed directly above the lesion. The cavernous angioma was completely resected (Figure 4A and 4B), and was confirmed by a postoperative pathological examination. Interestingly, two weeks after the operation, the itch was effectively relieved and the patient stated that the right facial pain was also relieved, with her pain score decreased to 3 (VPS). Microvascular decompression was not performed because during the one-year follow-up period, the symptom of right facial pain was successfully relieved and no new symptoms occurred. No lesions recurred during the follow-up period (Figure 4C and 4D).

3. Discussion

Central nervous system lesions that affect the sensory pathways, including strokes, multiple sclerosis, and cavernous hemangiomas, can cause central itching (8). Animal experiments have corroborated that most nociceptive afferents terminate in the superficial region of the dorsal horn (3, 9). ICA always causes NI because the gli-

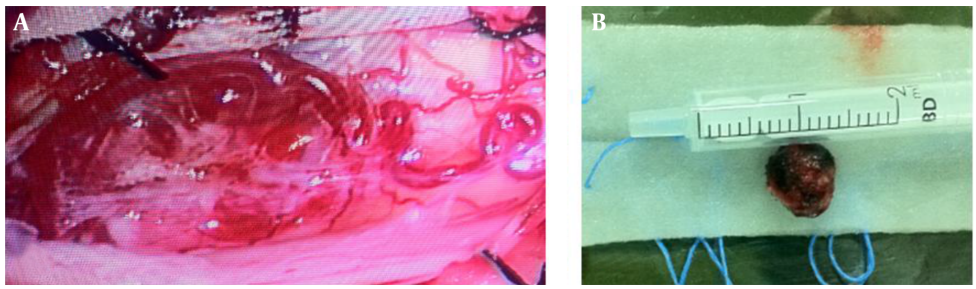

Figure 3. A dark red lesion was located in the dorsal spinal cord (A), with a diameter of 7 mm (B).

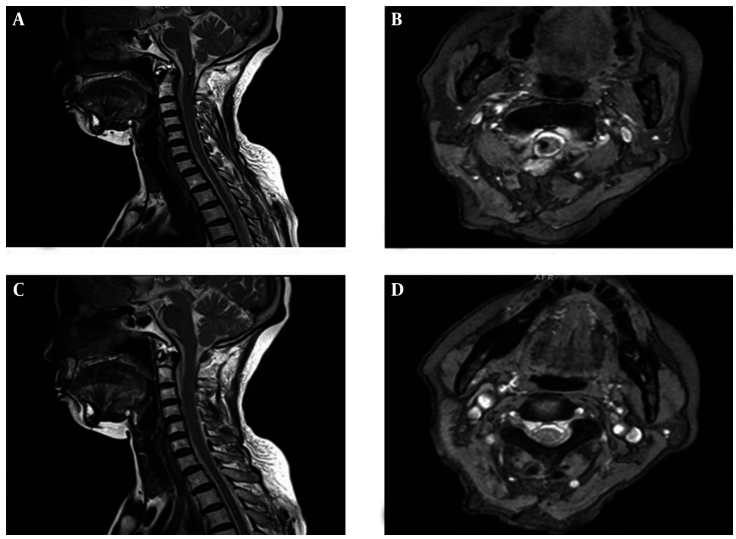

Figure 4. The area of the lesion, which was completely resected, is shown on the sagittal T2-weighted images (A) and the axial T2-weighted images (B) postoperatively. No lesion recurrence was observed on sagittal T2-weighted images (C) or axial T2-weighted images (D) after one year of follow-up.

otic rim containing hemosiderin-laden macrophages affects the neurons in the lamina I of the dorsal horn (3). TN is commonly encountered in clinical practice and is caused by compression of the fifth cranial nerve in the pontocerebellar angle by various lesions, especially due to vascular conflict with the trigeminal nerve (5, 8). Incidentally, TN has been effectively surgically treated by microvascular decompression.

Our ICA patient was unique in the sense that she presented with TN and NI together, and such a case has never before been published. Our case presented with a TN exactly like the one reported in 1989 by Saito et al. (7), and similar to published reports, her symptoms were relieved after surgery (3, 7). However, our case differs somewhat from the previously published case studies. For instance, in our case, the TN was found to be associated with a vascular conflict of the trigeminal nerve, which is different from the case reported by Saito et al. (7).

As in the published literature of human cases (3) and in the reports of animal experiments (3, 9), ICA is more likely to produce NI when the gliotic rim containing hemosiderin-laden phagocytes affects the lamina I of the dorsal horn (part of the itch pathway). Similar to the itch pathway, pain is commonly encountered in ICA. Indeed, it is challenging for neurosurgeons to deal with pain and itch. Antihistamines and anti-inflammatory drugs usually have no effect on NI (10). Thus, patients with NI require surgical treatment.

Our case presented with ICA and NI, as well as TN, with vascular conflict of the trigeminal nerve. Moreover, the TN, cervical pain, and itch were equally serious. Accordingly, it was important to decide at that point whether to first perform a microvascular decompression or an intramedullary lesion resection. A careful review the clinical history revealed that the right facial pain had not changed in two years, and was aggravated by the right cervical symptoms

during the last month. We hypothesized that the right cervical pain and the itch were caused by the ICA, while a remote hemorrhage could have affected the spinal tract of the trigeminal nerve, thus exacerbating the right facial pain. Intramedullary lesion resection was performed first, and the patient's symptoms were effectively relieved after the surgery, without the need for microvascular decompression (the TN recovered to the previous level), which proved our hypothesis. Sometimes, large ICAs can lead to facial pain due to affecting the spinal tract of the trigeminal nerve. However, when there is co-incidental vascular trigeminal nerve compression, the primary pathogenesis of the facial pain should be carefully identified. Our case demonstrated that the TN was mainly caused by the ICA. Hence, for an ICA with TN and NI, early diagnosis followed by timely and suitable surgical treatment is critical to relieve the symptoms.

The patient consented to submission of this case report to the journal.

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Footnotes

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