Trigeminal neuralgia in the presence of ectatic basilar artery and basilar invagination: treatment by foramen magnum decompression

Case report

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This 65-year-old woman presented with a 7-year history of classic trigeminal neuralgia. After a conservative therapeutic approach and because the pain had become unbearable, she was subjected to surgical treatment. Examinations revealed an ectatic basilar artery that indented deeply into the region of the root entry zone of cranial nerve V. Additionally, severe basilar invagination, the fusion of multiple cervical vertebrae, and partial agenesis of the clivus were observed. Foramen magnum decompression resulted in lasting relief from the pain. The authors discuss the pathogenesis of trigeminal neuralgia in a relatively rare clinical situation. (DOI: 10.3171/2009.6.JNS09469)

Key words • trigeminal neuralgia • basilar invagination • ectatic basilar artery • foramen magnum decompression

Fifth cranial nerve root entry zone compression due to an arterial or venous loop is an established cause of trigeminal neuralgia. Ectatic vertebral and basilar arteries have been frequently seen to cause vascular compression of the root entry zone of CN V.5,9,11 Microvascular decompression has been the most accepted treatment in this clinical situation. We describe the case of a patient with long-standing trigeminal neuralgia and an ectatic basilar artery and basilar invagination. Basilar invagination has rarely been associated with trigeminal neuralgia.3,7 We evaluated the relevant literature on the subject and the possible treatment strategy in such cases. The applied treatment for the trigeminal neuralgia in our case was foramen magnum decompression. Such a therapeutic modality for trigeminal neuralgia has not been reported in the literature.

Case Report

History and Examination. This 65-year-old woman, who was otherwise healthy and had no systemic illness, presented with symptoms of trigeminal neuralgia, which she had experienced for a period of 7 years. She had had a short neck since birth. A drug trial with carbamazepine was successful for a period of ~2 years but subsequently failed. The neuralgic pain became progressively unbearable. In the interim, all the molars and premolars in her upper jaw on the side ipsilateral to the pain were removed, as they were considered to be causing the pain.

When she was admitted to our institution, the pain was unbearable and she refused to remove a blanket covering her face or to talk. Even a whiff of air over her face resulted in severe neuralgic pain in the V2 and V3 divisions of the trigeminal nerve. There was no neurological deficit. Magnetic resonance imaging and CT scanning revealed an ectatic basilar artery that clearly indented into the root entry zone of CN V (Fig. 1A–C). Multiple vertebral segments of the cervical spine were fused, the cervical lordosis was obliterated, the clivus had significant agenesis, and the odontoid tip reached almost up to the sella.

Operation. Considering the presence of a marked craniovertebral anomaly and clear evidence of a reduction in the posterior fossa volume, foramen magnum decompression was performed. Bone decompression was difficult because of the small posterior fossa, with the external occipital protuberance reaching almost to the horizontal level of the foramen magnum. The posterior rim of the foramen magnum had an unusual inward deep curve, and the distance between the large and unusually shaped spinous process of the C-2 vertebra was markedly reduced.

Postoperative Course. Following surgery there was immediate relief from the neuralgic pain. At a follow-up
18 months after surgery, the patient had no recurrence of the neuralgia.

Discussion

Apart from vascular loops, a large number of etiological factors have been identified as causes of trigeminal neuralgia. Although rare, primary and secondary reductions of posterior cranial fossa volume have been identified as pathogenetic factors for trigeminal neuralgia. Ipsilateral posterior fossa tumors, contralateral posterior fossa tumors, and even supratentorial tumors have been reported to cause vascular compression syndromes. There are recorded instances in which achondroplasia, familial osteodysplasia, Chiari malformation, and basilar invagination have been identified as the cause of trigeminal neuralgia.

Microvascular decompression has been the accepted modality of treatment for this disease entity. Even in cases with local or remote tumors, it has been speculated that vascular compression at the root entry zone ultimately causes the pain. Resection of the tumor, with or without directly manipulating the vascular loop, has been associated with lasting pain relief. Trigeminal neuralgia has only rarely been identified in clinical situations that result in an obvious reduction in the posterior cranial fossa volume. It is unclear from the reported cases if the reduction in the posterior cranial fossa volume ultimately results in the vascular loop indentation at the root entry zone of CN V, or if the association between the 2 was an incidental observation. "Kinking" of normal-length arteries, which must adapt to a smaller posterior fossa in a case with basilar invagination associated with osteogenesis imperfecta, has been identified. Microvascular decompression has been successful in some of the reported cases in which there was trigeminal neuralgia in the presence of a small posterior cranial fossa.

In the presented case, basilar invagination was clearly associated with the presence of an ectatic basilar artery, which resulted in CN V root entry zone compression. The therapeutic surgical option of microvascular decompression versus posterior fossa decompression was a philosophical dilemma. Because the foramen magnum decompression resulted in lasting relief of the symptoms, it appears that the reduced posterior cranial fossa volume caused the ectatic basilar artery. We could not identify in the literature any combination of pathogenetic factors of trigeminal neuralgia such as those seen in our case. Successful treatment of trigeminal neuralgia by foramen magnum decompression has not been reported.

References


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