

CASE REPORT

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Chiari type 1.5 malformation as a cause of secondary trigeminal neuralgia: case report and literature discussion

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Abstract

Background Chiari syndrome is a very rare cause of secondary trigeminal neuralgia (TN). There are a few cases of TN associated with Chiari syndrome in the literature, and all of these cases were reported as Chiari type 1. In this report, we present a case of secondary trigeminal neuralgia caused by Chiari type 1.5 for the first time unlike the literature.

Case presentation A 38-year-old male patient, who had frequent and severe attacks of pain under the orbit, at the chin and rim of the mouth, was evaluated with craniocervical Magnetic Resonance Imaging (MRI) and revealed Chiari type 1.5 malformation and syringomyelia.

Conclusions The mechanism of TN associated with Chiari malformation is unclear. However, the main concepts emphasized in cases presented in the literature are compression and stretching. The rapid relief of pain after decompression surgery in these cases also supports this situation. It can be predicted that compression and tension forces will become more pronounced in Chiari type 1.5 malformation, in which the brainstem elongation and ventral pressure are more prominent. Decompression of the foramen magnum provides rapid and effective pain control in the treatment of TN accompanying Chiari 1.5 syndrome that does not respond to medical treatment.

Keywords Trigeminal neuralgia, Chiari tip 1.5 malformation, Syringomyelia, Suboccipital decompression

Background

Trigeminal neuralgia (TN) is defined as pain attacks in the form of electrical shoot-like occurring in the facial area innervated by one or more branches of the trigeminal nerve. These neuralgias are classified as classical TN due to vascular compression in the transitional zone of the trigeminal nerve, and secondary TN caused by causes such as tumor, aneurysm, and multiple sclerosis. A very rare cause of secondary TN is the Chiari malformation [1, 2]. In Chiari malformation, there is herniation of the cerebellar tonsils from the foramen magnum into the

spinal canal. Chiari malformation is divided into different types according to the degree of herniation (Table 1). In these patients, in addition to herniation, these patients also have elongation and sagging of the medulla and kink formation, angulation of the odontoid, and stenosis of the posterior fossa. These components distinguish Chiari malformation from the simple tonsillar herniation due to increased intracranial pressure. In addition to the types identified, some patients presented with caudal herniation of the cerebellar tonsils, the brainstem and obex without herniation of the fourth ventricle or vermis. These patients, who are neither CM I nor CM II, were previously considered part of CM I but were later defined as CM type 1.5 [3]. There are few cases of TN associated with Chiari syndrome in the literature, and all these cases have been reported as Chiari type 1. However, the mechanism of formation of trigeminal neuralgia in these cases is not clear. In this report, we presented a case of

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Table 1 Chiari Malformation subtypes [13]

Type I	Herniation of the cerebellar tonsils more than 5 mm below the foramen magnum Usually not accompanied by brain stem prolapse Hydrocephalus is rare
Type II	Displacement of the cerebellar vermis, brain stem, fourth ventricle caudally Accompanied by other intracranial anomalies Almost all have myelomeningocele and hydrocephalus Most have syringomyelia
Type III	Occipital encephalocele associated with intracranial anomalies seen in type 2
Type 1,5	Chiari I + Elongated brain stem and fourth ventricle
Type 0	Syringomyelia without brainstem prolapse

secondary trigeminal neuralgia caused by Chiari type 1.5 for the first time unlike the literature. We discussed the possible mechanisms in the development of trigeminal neuralgia and the treatment of pain.

Case presentation

A 38-year-old male patient was evaluated for pain in the form of attacks in the left half of the face, under the orbit, at the chin and on the rim of the mouth. Firstly, he was admitted to the dentist because of the pain and received treatment. However, there was limited improvement in pain. He also could not touch his face and his pain was triggered by even drinking water. Deep tendon reflexes were normal, and there were no pathological reflexes. Cerebellar tests were sufficient except for straight-line stepping. He had neuralgia triggered by sensory stimulation in the area consistent with the 2nd and 3rd branch of the trigeminal nerve. In the Cranial MRI, the cerebellar tonsils were herniated below the 16 mm of foramen magnum. The medulla oblongata extended downward from the foramen magnum and the obex was located approximately 2.5 cm below the foramen magnum. The 4th ventricle was elongated. Significant ventral compression was observed due to narrow posterior fossa and odontoid disclosure. There was also a syringomyelia extending from cervical to thoracic and wider than 5 mm due to impaired cerebrospinal fluid (CSF) dynamics (Fig. 1). The patient was operated in prone position using a spiked headgear. In the operation, decompression of the foramen magnum and C1 laminectomy was performed. The dura was opened with a 'Y' incision. It was observed that the obex was located under the foramen magnum. Arachnoidal bands were released. Tonsils were reduced with the help of bipolar cautery. Duraplasty was performed using a fascia lata. Pain attacks decreased significantly in the early postoperative period and completely disappeared on the 3rd postoperative day. In his follow-up 3 months later, he was free of pain without any medical treatment. Postoperative 6th month imaging showed decompression of the foramen magnum, restoration of the cerebellar

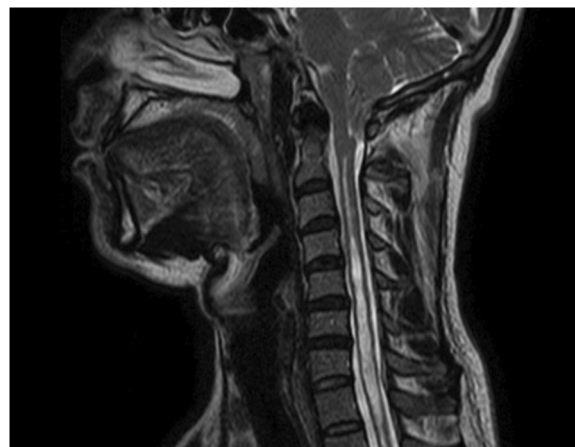


Fig. 1 Chiari type 1.5 is observed in the T2-weighted sagittal cervical MR image of the case. Obex is located below the foramen magnum. Elongation in the brainstem and syringomyelia extending to the upper medulla is seen

tonsils and regression of syringomyelia (Fig. 2). Ethics approval and consent to participate were obtained from the patient.

Discussion

The Chiari type 1 is a very rare cause of secondary TN. There are different theories about the mechanism of formation. The main theories have been reported as ischemia of the trigeminal nerve due to compression of the vascular structures, stretching of the trigeminal nerve due to the inferior location of the medulla, ventral compression, and close contact of the trigeminal nerve with the vascular loop [4–6]. The approaches and results in the treatment of case reports in the literature provide important clues in the evaluation of possible hypotheses. Vince et al. they reported a case whose pain was relieved by microvascular decompression. In this case, compression of the trigeminal nerve with the vascular loop was demonstrated both on MRI and during surgery. The authors discussed the role of compression by the small posterior fossa in Chiari type 1



Fig. 2 The T2-weighted cervical MR sagittal section shows decompression of the foramen magnum, restoration of the cerebellar tonsils and regression of syringomyelia

cases [1]. In fact, this case presented by Vince et al. also suggests that there may be an incidental association between Chiari type 1 and primary TN. When the frequency of Chiari's disease is compared with the number of cases with trigeminal neuralgia, the fact that trigeminal neuralgia is very rare and supports this possibility.

Some studies have reported that hydrocephalus leads to changes in CSF dynamics and causes traction of the trigeminal nerve in the basal cisterns and preponine segment, thus TN occurs [7]. The improvement of pain in patients treated with VP shunt and third ventriculostomy without foramen magnum decompression in cases with hydrocephalus indicates the role of CSF dynamics in the formation of secondary TN. It has been suggested that the shifting of neurovascular structures in the posterior fossa, which expands following CSF drainage, is effective in the improvement of pain [2, 7, 8]. However, the existence of chiari cases without hydrocephalus, such as our case, suggests that hydrocephalus is not the main factor in the development of trigeminal neuralgia. However, it is known that CSF dynamics are disturbed and there is a pressure difference between compartments in chiari cases even if hydrocephalus is not accompanied. Therefore, the absence of hydrocephalus does not exclude the idea that foramen magnum decompression improves pain by restoring CSF dynamics.

The syringomyelia extending to the medulla oblongata has been reported as another rare cause of trigeminal neuralgia [4, 9]. Syringomyelia is frequently seen in Chiari cases. The syringomyelia extending into the superior medullary area may affect the nucleus of the trigeminal nerve, leading to trigeminal neuralgia [9]. The presence of syringomyelia extending to the upper medulla was remarkable in our case. Although there was no regression of syringomyelia in the very early post-op period in our case, the improvement of pain limits the effect of syringomyelia in our case.

Holanda et al. reported the role of asymmetrical petrous bone development, very narrow posterior fossa, ventral compression and stretching of the medulla oblongata due to abnormal position of the odontoid in cases of secondary TN caused by basilar invagination [5]. Chiari cases have similar anatomical changes. Accordingly, ventral compression and stretching may be observed.

The Chiari type 1 cases which the brainstem is more elongated and the obex is located below the foramen magnum classified as Chiari type 1.5 in the last decades. Some authors suggest that this anatomical difference, described as Chiari type 1.5, influences the onset of symptoms and recovery [3, 10]. In our case, the obex was located quite below the foramen magnum, consistent with Chiari 1.5, and the brainstem was significantly elongated. When all the mechanisms reported above are evaluated together, the main factors emphasized in the development of chiari-induced trigeminal neuralgia are compression and stretching. It can be easily predicted that the compression and tensile forces will become more pronounced if the brain stem is more significantly elongated, as in Chiari 1.5. Our case strengthens the idea that compression and stretching are the main factors in the development of trigeminal neuralgia in chiari cases. In addition, the rapid improvement of pain after decompression surgery in our case supports this hypothesis. The presence of syringomyelia in our case clearly shows abnormal CSF dynamics between compartments. Therefore, the role of disturbed CSF dynamics in the occurrence of pain should not be ignored.

The decompression of the foramen magnum is the most preferred approach in the treatment of Chiari malformation. In the treatment of trigeminal neuralgia associated with Chiari malformation, a rapid and significant improvement in pain after decompression of the foramen magnum have been reported [2, 4–8, 12]. In our case, after the decompression of the foramen magnum (craniectomy, reduction of tonsils with bipolar cautery and duraplasty), the patient's pain improved rapidly and completely.

Conclusion

Trigeminal neuralgia may develop rarely in cases with Chiari malformation. The compression and stretching are common concepts emphasized in all possible mechanisms in the formation of trigeminal neuralgia. From this point of view, Chiari type 1.5 creates a more facilitating situation. Decompression of the foramen magnum provides rapid and effective pain control in TN with Chiari type 1.5 malformation.

Abbreviations

CSF	Cerebrospinal fluid
MRI	Magnetic resonance imaging
TN	Trigeminal neuralgia

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Not applicable.

Author contributions

H.E. helped in the idea of the research and collecting material. M.S. collected material and wrote the manuscript. All authors read and approved the final manuscript.

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Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate

Ethics approval and consent to participate were obtained from the patient.

Consent for publication

Consent for publication was taken from the patient.

Competing interests

There were no competing interests.

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