Improvement of Trigeminal Neuralgia Following Endoscopic Third Ventriculostomy in a Patient With Normal Pressure Hydrocephalus: A Case Report and Review of the Literature

Mohamadreza Hajjabadi1,2, Babak Babakhani3,4, Mostafa Farzin1, Sara Tavakolizadeh2

1 Department of Neurosurgery, Shariati Hospital, Tehran University of Medical Sciences, Tehran, Iran
2 Brain and Spinal Cord Injury Research Center, Neuroscience Institute, Tehran University of Medical Sciences, Tehran, Iran
3 Brain and Spinal Cord Injury Research Center, Iranian International Neuroscience Institute, Tehran University of Medical Sciences, Tehran, Iran
4 Imam Khomeini Hospital Complex, Tehran University of Medical Sciences, Tehran, Iran

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Abstract - Normal-pressure hydrocephalus (NPH) has rarely been reported to be associated with trigeminal neuralgia (TN). We report a case of NPH with trigeminal neuralgia, whose symptoms completely resolved after endoscopic third ventriculostomy. We will notify another surgical management of trigeminal neuralgia, considering the different pathophysiology of TN and a brief review of the literature.

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Introduction

Trigeminal neuralgia (TN) or “Tic Doloureux” is characterized usually with sever unilateral brief electrical- stabbing pain in one or more branches of the fifth nerve. This pain has been known since ancient times; there are descriptions of facial pain by one of the most popular Iranian physicians, Avicenna (980-1073 AD) (1). TN is a common orofacial pain and has a significant impact on the quality of life and the socio-economic functioning of the patient (1).

The pathophysiology of TN is usually in the trigeminal root entry zone. Traction or compression, demyelination or inflammation of this region can cause TN. Although the most common cause is vascular compression, but tumors, hydrocephalus, Chiari malformation, and Dandy-Walker syndrome, cranial base settling (2) have also been recently reported in the literature. Dr. Iseki reported a case of NPH with trigeminal neuralgia without focusing on the pathophysiology.

Improvement of TN after treatment of hydrocephalus has been reported in limited publications (2). In this report, we present a patient with normal pressure hydrocephalus and TN with a discussion about the pathophysiology of TN associated hydrocephalus.

Case Report

A 45-year-old woman who suffered from gait disturbance, urinary incontinence since 3 months ago. She had no papilledema. She complained of episodes of unilateral neuralgiform pain in the distribution of left V2 and V3. Bilateral temporomandibular joints and teeth were evaluated by the dentist. No sign and symptoms of temporomandibular disorder (TMD) was recorded. Carbamazepine did not afford any significant improvement of symptoms. Brain neuroimaging revealed ventriculomegaly without neurovascular compression at the trigeminal root entry zone (Figure 1). Regarding three ventricular ventriculomegaly, a lumbar puncture was not done. The patient underwent endoscopic third ventriculostomy. Urinary incontinence ceased on the second day after surgery. The patient was able to walk with no assistance one week after the surgery. The neuralgiform symptoms gradually improved, and the patient was pain-free with preoperative doses of carbamazepine 3 months postoperatively. Six months after surgery, she reported no episodes of facial pain despite discontinuing carbamazepine. The follow-up...
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imaging did not show any change in the size of the ventricles (Figure 2). After 3 years, she is still free of any complaint.

Discussion

Hydrocephalus associated TN has been reported rarely. Dr. Maurice-Williams and Pilling reported three cases with hydrocephalus and trigeminal neuralgia in 1977.

One year later, Dr. Tucker reported two patients with TN who suffered from raised intracranial pressure. The theory behind was a shift of brain stem due to intracranial pressure and traction of trigeminal nerve sensory root. Treatment of the hydrocephalus resulted in the remission of pain in these patients (3). Displacement, shift, or stretching of the brainstem may cause traction or compression of trigeminal root entry zone and promoting TN. TN with or without hemifacial spasm in association with Chiari malformation, Dandy-Walker syndrome, and cranial base settling have been recently reported (2,4).

Classically, (NPH) has been characterized by a syndromic triad of gait disturbance, urinary incontinence, and cognitive decline. The ventricles are dilated, but the CSF pressure is usually in the normal range. The standard treatment of NPH is CSF diversion with ventriculoperitoneal shunt or endoscopic third ventriculostomy. The actual pathophysiology of NPH is ambiguous. The old theory of the mechanism of hydrocephalus associated TN was the pressure gradient between supratentorial and infratentorial fossae. Theoretically, in most cases of NPH, regarding normal CSF pressure, rotation or traction of trigeminal nerve does not happen, and the suggested mechanism for neuralgia seems to be under debate. On the other hand, like secondary Parkinsonism in NPH due to basal ganglia compression, central cause like as reduced basal ganglia μ-opioid receptor (5) and dysfunction of multiple modulatory mechanisms may play a role not only in the chronicity of pain but also in the early pathogenesis. In this case, regarding three ventriculomegaly, trigeminal nerve displacement with probable central mechanism should be considered as the probable mechanism of neuralgia. We think that different underlying mechanisms should be considered in the upcoming studies to better understand and management of these patients.

We reported a case of trigeminal neuralgia associated with ventriculomegaly whose symptoms were completely resolved after CSF diversion by endoscopic third ventriculostomy. It seems that other mechanisms beyond the direct effect on the trigeminal nerve entry zone may play a role in the creation of trigeminal neuralgia.

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