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Trigeminal Neuralgia and Associated Hydrocephalus: Not a Mere Incidental Finding? A Case Report and Literature Review

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Background: Trigeminal neuralgia (TN) is most commonly related to vascular compression of the trigeminal root entry zone into the brainstem. Secondary causes of compression are much less common. TN as a sole manifestation of hydrocephalus has rarely been reported in the literature.

Case Report: We describe a young woman with medically resistant TN, who was discovered to have a tectal plate glioma causing secondary obstructive hydrocephalus. In addition, she had an incidental finding of ecchordosis physaliphora. Cerebrospinal fluid diversion by an endoscopic third ventriculostomy resulted in TN resolution.

Conclusion: Hydrocephalus is a rare diagnosis to consider as a secondary cause for TN. Since the literature on this is sparse, it can be only suggested that in very select cases, treating the hydrocephalus may also treat the TN. Additional studies and reports are required.

Key words: Trigeminal neuralgia, hydrocephalus, ecchordosis physaliphora, tectal plate glioma

BACKGROUND

Trigeminal neuralgia (TN), is a chronic facial pain disorder commonly characterized by unilateral, recurrent, brief bouts of severe, electric shock-like pains appearing in any or all of the dermatomal distributions of the 5th cranial nerve (CNV, trigeminal) (1-3). The pain usually lasts seconds to minutes and can occur spontaneously, or more commonly be triggered by a nonnoxious, innocuous stimulus to the affected CNV dermatome (4,5). TN is a rare condition, with a peak age of onset typically between 50 and 70 years of age, having a prevalence of 20 to 200 per 100,000; women are generally affected twice as much as men (1,2,5,6). The most commonly affected branches of CNV are the maxillary (V2) and/or the mandibular (V3), with a higher prevalence on the right side, where the pain is usually more paroxysmal and sharper in nature (2,5,7).

TN's etiology is unknown. There are many theories regarding the pathophysiology of this disorder, but it is generally believed that TN results from complex neurophysiological nociceptive interactions, resulting in hyper-excitability of the nerve (5,8–10). The most common cause of TN is a neurovascular conflict, with trigeminal nerve root compression by the superior cerebellar artery or a petrosal vein, which results in focal demyelination of the nerve root as it exits the brainstem at the prepontine cistern (11).

We describe here a rare case of a young woman with medically resistant TN, as well as hydrocephalus secondary to a tectal tumor. She also had an incidental

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finding of ecchordosis physaliphora. We also provide a detailed literature review.

CASE PRESENTATION

A 27-year-old healthy woman presented to our department with an 18-month history of TN involving the left side of her face, in dermatomal distribution of the maxillary and mandibular divisions of the trigeminal nerve. The pain was intermittent in nature. She had a pain-free period until a month before her admission. At that time her symptoms had spontaneously worsened. During that period, a trial with carbamazepine failed and was discontinued because she suffered from medicinal side effects.

A magnetic resonance imaging (MRI) scan of the brain was performed prior to her hospital admission. It revealed marked enlargement of the lateral and third ventricles (Fig. 1) resulting from obstructive hydrocephalus caused by a tectal glioma, compressing the cerebral aqueduct (Fig. 1). The tectal plate (i.e., tectum), is located at the dorsal part of the midbrain, posterior to the cerebral aqueduct of Sylvius. A mass lesion in this location can therefore compress the cerebral aqueduct of Sylvius leading to obstructive hydrocephalus.

Our patient's tectal tumor was well-defined, hypointense nonenhancing on T1 weighted images (T1WI), hyperintense on T2 weighted images (T2WI), and fluid-attenuated inversion recovery (FLAIR). Notably, Chiari malformation was not demonstrated. In addition, there was an incidental, extra-axial, small rounded mass located along the left side of the midline in the prepontine cistern, not touching the surface of the pons or the trigeminal nerve (Fig. 2). No neurovascular conflict explaining the TN was evident on the constructive interference in steady-state sequence. The lesion appeared hypointense on T1WI and hyperintense on T2WI, and did not enhance after gadolinium administration.

Interestingly, our patient did not present with any increased intracranial pressure symptoms such as headaches, nausea, vomiting, or visual concerns. The patient underwent an endoscopic third ventriculostomy procedure, during which the incidental mass was identified and resected (Fig. 3). Postoperatively, the patient recovered well and had significant relief from her symptoms, that began occurring at a much lower frequency and intensity. She was discharged home on postoperative day 3. A postoperative MRI performed at 3 months demonstrated a reduction in the volume of the ventricles (Fig. 4). Furthermore, at a follow-up clinic visit 4 months later, she reported being symptom-free. A pathological evaluation of the incidental mass diagnosed ecchordosis physaliphora, a benign lesion, derived from notochordal remnants (12), having morphological characters similar to well-differentiated chordoma.

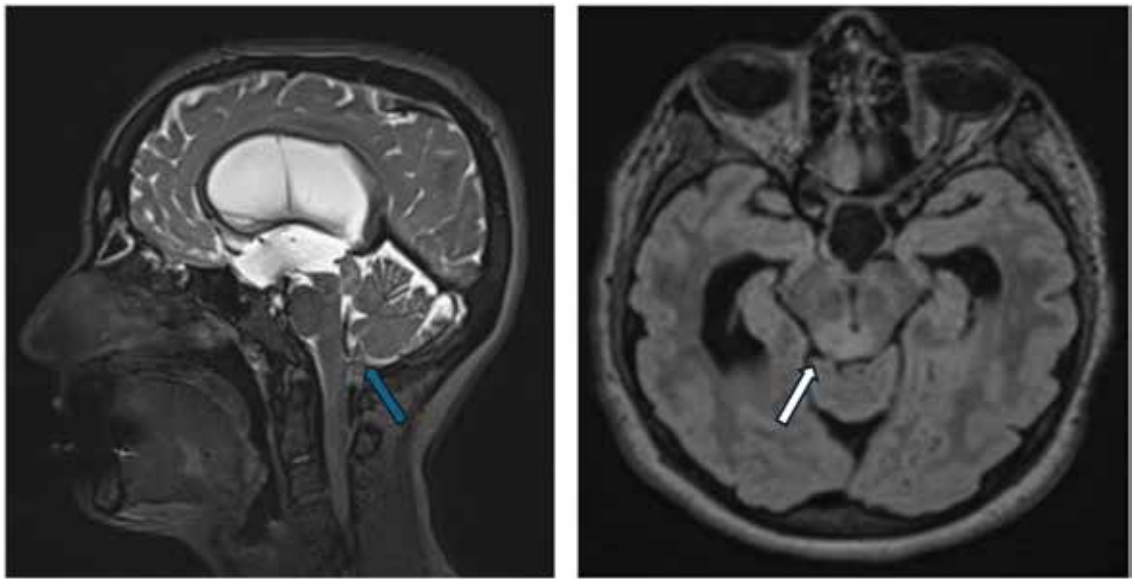

Fig. 1. Sagittal T2WI scan shows marked enlargement of the lateral and third ventricles, resulting from obstructive hydrocephalus caused by a tectal tumor. Chiari malformation is not demonstrated (blue arrow). Axial FLAIR scan at the midbrain level, shows a well-defined hyperintense tectal plate glioma (white arrow).

DISCUSSION

There are many international guidelines classifying TN (13). Generally, TN is subclassified into primary (classic), secondary, or idiopathic (6,14). The primary type, the most common, is caused by trigeminal root vascular compression resulting in root atrophy and/or displacement, as seen on preoperative imaging or revealed during surgery (6).

The most common cause is the superior cerebellar artery. It is responsible for about 50% of vascular compression cases (2,5,8,15), while the anterior inferior cerebellar artery, basilar artery, and pontine veins are responsible for the majority of the rest (5,16,17).

The secondary type, which accounts for about 15% of cases, is diagnosed when other identifiable disease

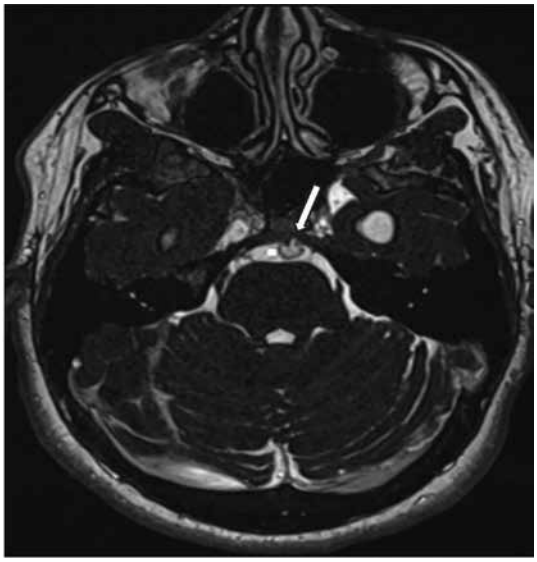

Fig. 2. Axial constructive interference in steady-state (CISS) T2WI sequence, shows a hyperintense, extra-axial, stalk-like structure projecting from the clivus (white arrow), in the prepontine cistern, with no contact with the pons or the trigeminal nerve.

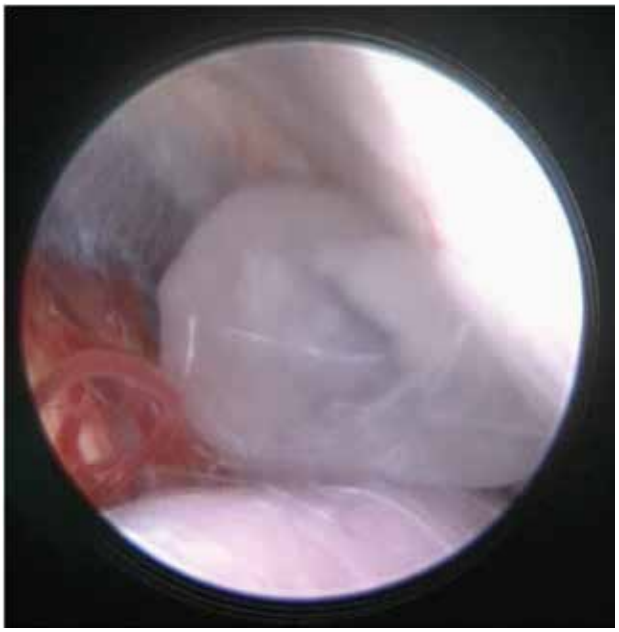

Fig. 3. Intraoperative endoscopic image demonstrating a cystic gelatinous nodule located ventral to the basilary artery. The mass was easily removed.

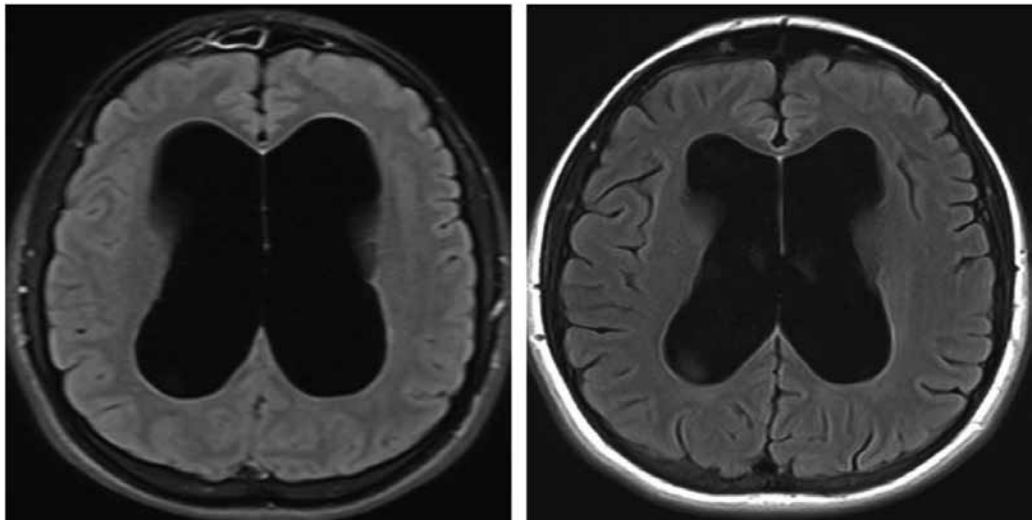

Fig. 4. Axial constructive T1W1 MRI demonstrating a reduction in the volume of the lateral ventricles at postoperative MRI (right) compared to preoperative MRI (left).

or mass, except vascular compression, can be attributed to TN, including arteriovenous malformation, vestibular schwannoma, skull base meningioma, pontine infarction, intracranial hypotension, autoimmune connective tissue diseases, Chiari malformation type I, epidermoid cyst, and multiple sclerosis, the last often resulting in a bilateral TN presentation (2,6,18-28).

The idiopathic type accounts for the rest and, after adequate investigation, is diagnosed by exclusion when there is no apparent cause (6,14).

The association of hydrocephalus, as a secondary cause of TN, has rarely been reported (Table 1) (29-37). There are several proposed mechanisms by which hydrocephalus may cause TN, including speculation that hydrocephalus causes downward displacement of the brainstem and as a result, stretches the trigeminal sensory nerve root, altering the normal anatomical relationship between the vascular loop and the trigeminal nerve, predisposing its compression (26,31,33). Therefore, cerebrospinal fluid drainage relieves the vascular impingement on the trigeminal nerve, leading to resolution of the symptoms.

While ecchordosis physaliphora has been rarely reported in the literature as a cause of trigeminal symptoms (12,38), we believe that the coincidental finding of ecchordosis physaliphora in our patient is only by chance alone; it did not represent the etiology of TN in our case, since the lesion was small and did not have direct contact with the brainstem or trigeminal nerve.

CONCLUSION

Hydrocephalus is a rare diagnosis to consider as a cause of secondary TN, and should be kept in mind while looking for secondary causes. Since the literature on this is sparse, it can be suggested that in very select cases, the treating the hydrocephalus may also treat the TN.

Table 1. Literature review, reporting trigeminal neuralgia associated with hydrocephalus

Author	No. of patients (n = 15)	Age, Gender	Hydrocephalus cause	Hydrocephalus treatment	TN symptoms resolved
Seeger, 1963 (32)	3	Unknown	Obstructive hydrocephalus	Yes (unknown method)	Yes
Maurice-Williams and Pilling, 1977 (31)	2	47, W	Communicating hydrocephalus	Ventriculo-atrial shunt	Yes
		22, W	Post-meningitis obstruction	Ventriculo-atrial shunt	
Tucker et al, 1978 (29)	2	29, W	Aqueductal stenosis	Ventriculo-cisternal shunt	Yes
		24, M		VPS	
Findler and Feinsod, 1982 (30)	1	42, W	Aqueductal stenosis	VPS	Yes
Gnanalingham et al, 2005 (33)	1	31, M	Chiari 1 malformation	VPS	Yes
Teo et al, 2005 (34)	1	38, W	Chiari 1 malformation	ETV	Yes
Vince et al, 2010 (37)	1	50, W	Chiari 1 malformation	Craniocervical decompression and VPS	Yes
Liu et al, 2014 (35)	1	24, M	Chiari 1 malformation	VPS	Yes
Na et al, 2017 (36)	3	31, M	Aqueductal stenosis	ETV	Yes
		45 M	Aqueductal stenosis	ETV	
		50 W	Communicating hydrocephalus	VPS	

VPS, ventriculo-peritoneal shunt; ETV, endoscopic third ventriculostomy; W, women; M, men.

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