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Case Report

Trigeminal Neuralgia as An Atypical Manifestation of Intracranial Hypertension. Case Report and Literature Review

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Abstract

Trigeminal neuralgia, a chronic neuropathic pain, is characterized by frequent episodes of stabbing facial pain, in the territory of the trigeminal nerve, which can severely impair patient's quality of life. It is primarily caused by vascular compression of the trigeminal nerve root, but it is sometimes secondary to other identifiable neurological conditions, such as multiple sclerosis or a cerebellopontine angle tumor. Uncommonly, it may be caused by the presence of idiopathic intracranial hypertension, a condition most likely identified by headache, visual disturbances, and sixth nerve palsy, among other manifestations. Secondary causes of trigeminal neuralgia are usually detected after an atypical clinical manifestation, such as ophthalmic rather than maxillary or mandibular nerve involvement, more than one branch affected, or bilateral symptoms. We present a patient who presented with trigeminal neuralgia as a primary manifestation, and after thorough evaluation she was diagnosed with intracranial hypertension as the underlying mechanism responsible of the trigeminal neuralgia.

Keywords: *Trigeminal Neuralgia, Idiopathic Intracranial Hypertension, Meckel's Cave, Intracranial Pressure.*

Abbreviations

IIH: Idiopathic Intracranial Hypertension; IH: Intracranial Hypertension; CSF: Cerebrospinal fluid; ICP: Intracranial pressure; TN: Trigeminal neuralgia; NSAIDs: Non-steroidal anti-inflammatory drugs; CTA: Computed tomography angiography; MRI: Magnetic resonance imaging; MC: Meckel's Cave; CVST: Cerebral venous sinus thrombosis.

Introduction

Idiopathic Intracranial Hypertension (IIH), previously known as "Pseudotumor cerebri", is a disorder characterized by increased intracranial pressure with no known, apparent or evident cause. Patients present with clinical signs and symptoms of increased intracranial pressure, such as headache, visual disturbances, papilledema, and sixth nerve palsy (1), but are otherwise alerted and oriented (2). Lumbar puncture and cerebrospinal fluid (CSF) analysis are normal, except for elevated intracranial pressure (ICP), and neuroimaging findings are unremarkable except for findings attributable to chronic increased intracranial pressure, in the absence of intracranial mass or ventricular dilatation (1).

The nomenclature of IIH has changed over the years, first named by Quincke in the late 19th century "Serous meningitis", later "Pseudotumor cerebri" and more recently "Benign intracranial hypertension" (3).

The latter has been termed an inappropriate concept, considering that if left untreated, the condition may progress with visual loss and chronic headache (4), which may severely impair quality of life by deteriorating functional independence, producing medication overuse headache, among other complications.

Albeit IIH can be sometimes easily diagnosed when the full clinical picture is recognized, it may present a challenge when atypical manifestations are present. Some of them include asymmetric or unilateral papilledema, IIH without papilledema, third or fourth nerve palsy, internuclear ophthalmoplegia, olfactory disfunction, hearing and vestibular disfunction, lower cranial nerve disfunction, facial nerve disfunction, and trigeminal neuralgia (5).

Regarding the latter, trigeminal neuralgia (TN) is a chronic neuropathic pain characterized by spontaneous frequent paroxysms of stabbing pain in a region innervated by one or more of the trigeminal nerve branches (6), which has been associated with poor quality of life, and in severe cases even suicide has been attributed to the condition (7). Its lifetime prevalence is estimated between 0.1-0.3% (8), being more prevalent in women than in men, with a W:M ratio of 3:2 (3). The TN incidence increases with age, ranging in adults between 24-93 years, with a mean age of onset of 55 years (9).

It has been classified traditionally in classical, secondary and idiopathic types, being the classical the most common, comprising around 75% of all cases, and it is usually caused by intracranial vascular compression of the trigeminal root, usually the superior cerebellar artery, inducing morphologic changes in the trigeminal nerve root (6). Secondary TN, accounting for about 15% of the cases, is attributed to an underlying neurological condition (except for the classical neurovascular compression), such as a cerebellopontine angle tumor, arteriovenous malformation, or multiple sclerosis, the latter specially in young adults, intracranial hypertension, among others (10). Multiple sclerosis related TN has been attributed to a demyelinating plaque in the fascicle of the trigeminal nerve as it courses through the central pons (11), and it can be sometimes be the isolated manifestation of a patient presenting with multiple sclerosis (12). Cerebellopontine angle tumors can often be the underlying condition responsible for TN, whether is a neurinoma, meningioma or epidermoid cyst (13). In a series of four studies, comprising 243 people involved, a tumor was the cause of 8% of the cases of TN (10), generating compression and focal demyelination of the trigeminal nerve root, triggering the same high frequency discharges that occur in classical neurovascular compression (13). Trigeminal neuralgia as a manifestation of IIH has been rarely reported (5,14). In some cases, the TN presentation was resolved after treatment with acetazolamide after lumbar puncture demonstration of increased intracranial pressure (15), suggesting a pressure related phenomenon that might be responsible of the overlapping condition (16,17).

Patient and Methods

Patient is a 24 y/o white Caucasian female with a normal corporal mass index, and a past medical history of cerebral venous thrombosis (CVST), diagnosed and treated when she was 18, and involved the left transverse sinus, left sigmoid sinus, and part of the superior sagittal sinus, without secondary venous stroke, and attributed at the time to the use of hormonal contraceptive medication. She completed a 6-month anticoagulation therapy with warfarin, and a thrombophilia test pack was performed, and eventually it came with negative results. On follow up she persisted with episodic unilateral headache, treated with NSAIDs and amitriptyline with partial response. 5 years after the described event she started with a progressive, sporadic, intense, stabbing pain lasting only a few seconds in the left side of the face. Two weeks before hospital admission she presented with exacerbation of the clinical condition, with an increase in the frequency of the episodes, appearing on a daily basis. She was then admitted for a complete workup and treatment in the neurology wards of our hospital. Neurological physical examination revealed paresthesia and trigger points in the V1 and V2 territories of the left trigeminal nerve with the rest of the cranial nerves unaffected. Rest of the neurological and general physical examination was otherwise unremarkable.

Due to the atypical presentation of TN with two nerves involved, a complete workup was done. Computed tomography of the brain was normal, CTA was as well unremarkable, especially the venous sinus which were completely permeable. Brain MRI showed lower amplitude of the ventricular system, sellar arachnoidocele (Figure 1 and 2), small basal bilateral encephalocele (Figure 3), and optic nerve tortuosity with fluid surrounding them (Figure 4) with significant compression of the left MC, with a lesser degree of involvement of the right one (Figures 5 and 6). Fundoscopic examination also showed a small excavation of the optic papilla, with slightly blurred edges, raised half diopter, with a healthy macula.

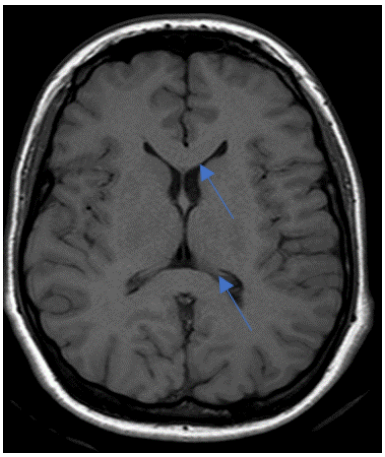

Figure 1: *Axial T1 sequence, lower amplitude of the ventricular system (blue arrows).*

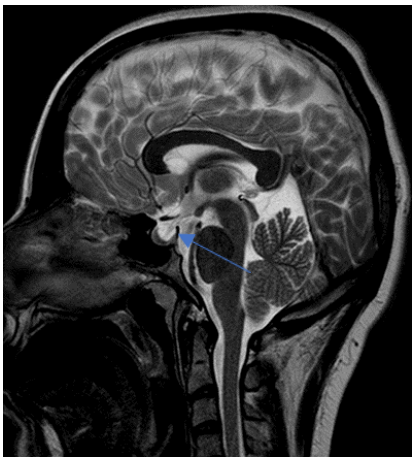

Figure 2: *Sagittal T2 sequence, sellar arachnoidocele (blue arrow).*

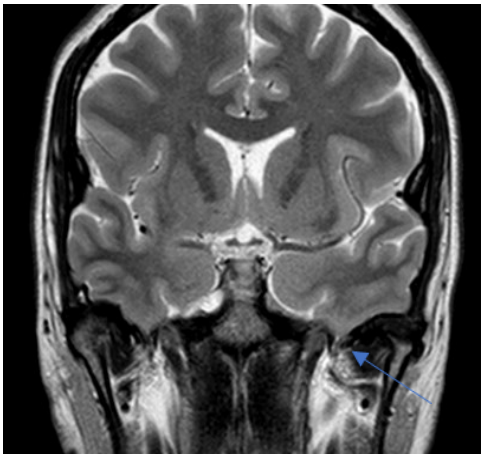

Figure 3: *Coronal T2 sequence, small temporal encephaloceles (blue arrow).*

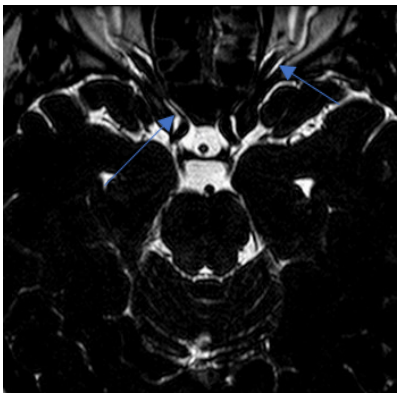

Figure 4: *Axial T2 drive sequence, enlarged optic nerve sheath diameter (blue arrows).*

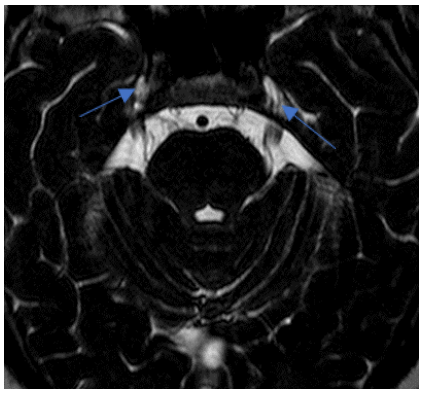

Figure 5: *Axial T2 drive sequence, example of normal MC (blue arrows).*

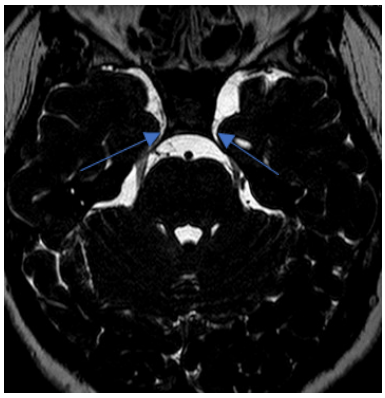

Figure 6: *Axial T2 drive sequence 3 mm, compression of both MC, most notably the left one (blue arrows).*

Due to the described findings, a lumbar puncture with pressure analysis was performed, which revealed a 39 cmH20 opening pressure, with normal laboratory results. Due to the clinical picture, fundoscopic examination and radiologic tests performed, alongside the pressure findings, the diagnosis of intracranial hypertension with MC compression and secondary TN involvement was made, as an atypical manifestation of IIH. The patient was treated with carbamazepine and acetazolamide with great clinical response to both the IIH and TN symptoms, with a 3-month follow up that showed complete resolution of the clinical symptoms and normalization of the intracranial pressure. Patient maintained regularly clinical follow-up to date.

Discussion

TN as a manifestation of IIH is a rare, but a feasible condition that can hide the underlying condition (17). It is not clear whether the trigeminal nerve involvement is directly affected due to raised ICP or as a consequence of the morphological changes visible in MC, becoming enlarged or indented (18). A plausible theory explains that the morphological changes previously described predisposes the trigeminal nerve to compression or distension as it traverses MC (18). Alas, these alterations have been identified in patients with TN without IIH and have been implicated as a cause of TN in patients in whom neurovascular compression has been discarded (19). In our patient the clinical presentation of TN was clearly atypical, especially regarding the simultaneous V1 and V2 involvement, which warranted a completely a thorough work up. In classical TN, the nerve involvement is usually more frequent in V3, less frequent in V2 and rarely in V1 (3,7,11), hence the need in our patient to a complete study in order to exclude secondary conditions. The past medical history of CVST prompted immediate search for radiological elements of persistent thrombosis with absence of recanalization, but both MRI and CTA failed to prove that.

In a retrospective cohort of 102 patients with CVST, the authors reported that up to 10% of the patients developed intracranial hypertension (20), and described a relationship between the latter and the absence or incomplete recanalization of the venous system on follow up (20). Although the relationship between CVST and IH has been widely recognized in the acute setting (21), the manifestation of IH after complete remission of CSVT has not been fully studied.

There is however, an undeniable anatomic relationship between the CSF and the venous system, since the arachnoid villi represent the main pathway of resorption of CSF from the subarachnoid space into the lumen of the venous sinuses, and from there to the jugular vein and the general venous circulation (22). This connection, although anatomically feasible, can be expanded in several physiological hypotheses linking the CSF and interstitial fluid with the glymphatic, lymphatic and venous pathways of drainage with the development of IH (23), so it is theoretically possible that albeit no anatomic irregularities were found on MRI or CTA of the previous CSVT, there could be interstitial or venous alterations at the cellular level, warranting further research to understand this physiological relationship.

Conclusion

Our case report visualizes the need to extend the knowledge of atypical presentations of typical conditions, in this case the presence of TN in the setting of underlying IIH. Intracranial hypertension, although a rare condition mimicking as trigeminal neuralgia, should be searched in patients in whom atypical or secondary TN is suspected, especially in women or in patients with prior CSVT. This case prompts the need to further research the cellular and physiological implications of CSVT in the potential development of IH in patients, even if total recanalization is achieved. In this setting, a directed analysis towards the MC in neuroimaging is warranted when this association is suspected.

Declaration of Patient Consent

The authors certify that an informed consent was obtained from the patient in order to access medial records regarding the use of clinical information and images. The patient understands that the use of this information is classified, and all due efforts are made to ensure that no personal information is revealed in the process.

Conflicts of Interest

The authors declare no conflict of interest.

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