Trigeminal Neuralgia Due to Meckel's Cave Crowding in the Setting of Possible Idiopathic Intracranial Hypertension without Papilledema

Sir,

Trigeminal neuralgia (TN) can be a debilitating condition characterized by recurrent neuralgic painful paroxysms in the distribution(s) of one or more divisions of the trigeminal nerve, most often the maxillary and mandibular divisions.[1] The usual goal of neuroimaging is to identify neurovascular compression of the dorsal root entry zone of the trigeminal nerve ("classic" TN) and also to exclude the wide gamut of neoplastic, inflammatory, infective, demyelinating, vascular, traumatic, dental, and toxic causes of "symptomatic" TN or trigeminal neuropathy.[2] Herein, we describe the case of a female with symptoms of TN due to crowding of Meckel's caves by expanded CSF in idiopathic intracranial hypertension (IIH); specifically, in the setting of Possible Idiopathic Intracranial Hypertension Without Papilledema (PIIHWOP).[3,4]

A 63-year-old female presented with more than a year history of facial pain. The pain was initially intraoral but later converted to intermittent, paroxysmal "electric-shock like pain" in the left upper and lower gums/teeth and occasionally in the left temporal region. Each episode lasted 1–2 min and was triggered by the cold wind, cold temperature, brushing teeth, or touching her face. Her

symptoms were partially controlled by oral baclofen and oxcarbazepine but she was unable to tolerate higher dose of medications. There was no facial or intraoral numbness or tingling sensation. The previous history comprised of remote thyroidectomy for a benign thyroid mass, asthma, and sinusitis. There was no history of migraines, headaches, or visual symptoms. Neurologic and dental examinations were normal. Physical examination revealed a morbidly obese body habitus with a body mass index (BMI) of 49.21 kg/m². The clinical diagnosis was established as left TN per ICHD-3 diagnostic criteria,[5] involving mandibular and maxillary divisions with a presumptive etiology of neurovascular compression. Subsequently, contrast-enhanced MRI of the brainstem and face and a dedicated heavily including T2-weighted (T2w) imaging of the trigeminal nerves was performed [Figure 1]. CT venogram revealed tapering of bilateral distal transverse sinuses near the transverse-sigmoid sinus junctions also suggestive of IIH. There was no cerebral venous sinus thrombosis. Lumbar puncture revealed elevated opening pressure of 30.8 cm of water (measured in left lateral decubitus) with unremarkable cerebrospinal fluid analysis (glucose 51 mg/dL, protein 29 mg/dL, 1/mcL total nucleated cell, and 3/mcL red cells). The diagnosis of possible idiopathic intracranial hypertension without papilledema (PIIHWOP) was made using recently published consensus and guidelines,[3,4] and the patient was started on oral acetazolamide and weight loss regimen. At 2-months follow-up, the patient had a weight loss of 7 kg, near-complete resolution of her left-sided face pain and lowered intake of her pain medications. At 4-months follow-up, the left-sided pain had been completely resolved. She subsequently maintained the treatment regimen without the return of the left TN.

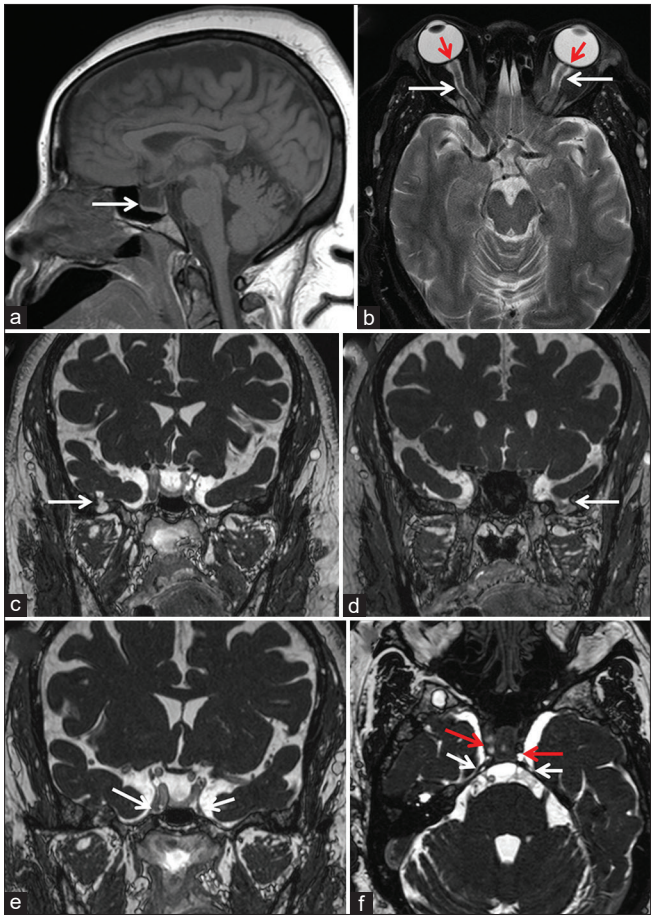

Figure 1: T1-w eighted image shows partially empty sella morphology (white arrow, a). T2-weighted image (b) shows mildly tortuous optic nerves with prominent perioptic CSF spaces (white arrows, b) and slight posterior flattening of globes (red arrows, b). Heavily T2-weighted constructive interference steady state (CISS) images (c and d) show lateral sphenoid meningoceles (white arrows, c, d). Findings suggest IIH. T2-weighted CISS images show maxillary divisions in the cavernous sinuses (white arrows, e) and Meckel's caves (white arrows, f) are crowded by medial temporal meningoceles (red arrows, f) and Meckel's caves have a slit-like configuration

The modified Dandy Criteria, the basis for a definite idiopathic intracranial hypertension (IIH) diagnosis mandates intracranial hypertension symptoms (e.g., headache, pulsatile tinnitus) and signs (e.g., papilledema and/or sixth cranial nerve palsy) are present in the setting of intracranial hypertension of unclear cause.[6] More recent consensus diagnostic criteria specifically label those meeting modified Dandy criteria who have no papilledema but have sixth cranial nerve palsies as suffering from idiopathic intracranial hypertension without papilledema (IIHWOP).[3] Although not specific, neuroimaging findings can suggest IIH [Table 1].[7] There is a subset of patients, however, in whom an IIH diagnosis is "suggested" based on neuroimaging findings who have documented intracranial hypertension without known cause ('idiopathic"), and have no papilledema or other physical exam signs of intracranial hypertension. This latter subset has been classified in the recent consensus diagnostic criteria as "Possible Idiopathic Intracranial Hypertension Without Papilledema (PIIHWOP)"[3,4] and is the group where our reported case belongs to [Table 1]. Important to note is the fact that PIIHWOP has not been well studied yet and whether its

Table 1: Possible Idiopathic Intracranial Hypertension without Papilledema (PIIHWOP). Adapted from[3]

1.	Normal neurologic exam
2.	Neuroimaging shows normal brain parenchyma and no cerebral venous sinus thrombosis
3.	Normal CSF constituents
4.	Elevated lumbar puncture pressure>25 cm CSF
Plus:	
5.	Three neuroimaging findings suggestive of raised intracranial pressure
o	Empty sella
o	Flattening of the posterior globe
o	Distention of perioptic subarachnoid space +/- tortuous optic nerve
o	Transverse cerebral venous sinus stenosis

(3). Mollan SP, Davies B, Silver NC, Shaw S, Mallucci CL, Wakerley BR, *et al*. Idiopathic intracranial hypertension: Consensus guidelines on management. J Neurol Neurosurg Psychiatry 2018;89(10):1088-100

presentation could differ from that of definite IIH or IIHWOP remains unclear.

This case demonstrates an association of IIH with TN and imaging evidence consistent with Meckel's caves crowding by meningoceles. Previous older clinical vignettes have described both TN and trigeminal neuropathy associated with IIH that was resolved by lumbar puncture and conservative management; however, no further details or neuroimaging was available.[8,9] Another report described electrophysiological evidence of trigeminal neuropathy in a patient with IIH which improved after conservative management but did not have neuroimaging correlate.[10] While TN may be incidentally seen in IIH, it is important to note that treatment directed towards IIH may lead to symptomatic resolution of TN. Contrary to the uncommon association of TN with IIH, small meningoceles/encephaloceles in the temporal bone and sphenoid wing are common in IIH. This is presumably a compensatory mechanism wherein chronically elevated CSF pressure remodels the thinner areas of the skull base, with subsequent bulging of meninges through skull base foramina and arachnoid pits.[11]

While IIH can be associated with Meckel's caves enlargement,[12] crowding of Meckel's caves appears to be unusual. A few prior reports have also described TN due to Meckel's caves meningoceles that were treated surgically but an underlying etiology of IIH seems to be rare.[13-15] In this clinical setting, recognition of the imaging signs of IIH and the appropriate clinical diagnosis of IIH and its variants is important to facilitate appropriate medical and lifestyle, rather than surgical, treatment of TN.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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Submitted: 05-Jan-2020 **Revised:** 21-Jan-2020 **Accepted:** 08-Feb-2020
Published: 06-May-2020

DOI: 10.4103/aian.AIAN_690_19