

Hemifacial Spasm as an Atypical Presentation of Idiopathic Intracranial Hypertension - A Case Report

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SUMMARY:

Idiopathic intracranial hypertension (IIH) is a challenging disorder of intracranial hypertension in the absence of an intracranial mass, hydrocephalus, or other identifiable etiology with normal cerebrospinal fluid content. The manifestation of the disease varies from typical headache, transient visual obscuration to less common features as 6th nerve palsy and rarely facial palsy and hemifacial spasm. There are a few reports of hemifacial spasm as a manifestation of the IIH in the literature. Also, there is no report of IIH with only hemifacial spasm. Here in, we describe a 39 year old woman with refractory hemifacial spasm who was eventually diagnosed with IIH to highlight the importance of complete neurologic examination in all patients and an more exploratory view in approach to hemifacial spasm.

Keywords: Hemifacial spasm, hypertension, idiopathic intracranial hypertension

INTRODUCTION

Idiopathic intracranial hypertension (IIH) is a challenging disorder of intracranial hypertension in the absence of an intracranial mass, hydrocephalus, or other identifiable etiology with normal cerebrospinal fluid content. The incidence is approximately 0.9/100.000/year rising to 13/100.000/year in overweight women between 20 and 44 years of age^{1,2,3,4,5,6}.

The most common manifestation of the disease include of sub-acute headache accompanied by transient visual obscuration, visual field defect, blurred vision in advanced stages and tinnitus. However less common presentation as diplopia due to 6th nerve palsy and rarely peripheral facial palsy and hemifacial spasm has been reported mostly in the pediatrics^{7,8,9}.

On the other hand, hemifacial spasm should always raise the suspicion of a microvascular compression at the facial nerve root exit zone from the brainstem or less common at it's entry zone into the internal auditory meatus¹⁰.

There are only a few prior cases of IIH associated with HFS in the published literature. Furthermore, there is no report of this case in Iran. Here in we aim to describe a case of refractory left hemifacial spasm who was eventually diagnosed with IIH.

CASE PRESENTATION

Patient's demographic, History, physical exam: The patient was a 39-year-old right handed woman who was presented in referral for a one year history of refractory hemifacial spasm. Since last year, she had developed intermittent twitching and "drawing-up" of the left side of her face with sustained closure of the left eye lasting for 1-2 minutes which was unassociated with other involuntary movement or trigger factors.

In her previous neurologic work up she had been under brain magnetic resonance imaging (MRI) which was not suggestive of vertebral-basilar artery structural lesion.

Subsequently, the patient was started on medical treatment including of Carbamazepine 200 mg TDS, Gabapentine 300 QHS, Baclofen 10 mg BD. Despite of medical treatment, no improvement was observed and the frequency of spasms has gradually increased resulting to significant discomfort for the patient.

On our visit, the patient did not notice any complaint of headache, blurred vision, visual field defect or tinnitus. The past medical history was unremarkable.

On examination, frequent left hemifacial spasm resulting to eye closure was seen. There was no facial weakness. Visual acuity was 20/20. The pupils were midsize and reactive to light. The confrontation test did not show any visual field restriction. Funduscopy revealed moderate bilateral papilledema. The remainder of the ophthalmologic and neurologic examination was normal.

Patient's laboratory and diagnostic data: The brain MRI was re-evaluated which showed marked empty sella. The routine biochemistry tests were normal. Once the diagnosis of bilateral optic disc swelling was made, the patient underwent lumbar puncture to assess the intracranial pressure. The opening CSF pressure was 42 cm H₂O and the analysis was normal.

Based on the findings, the patient was diagnosed with idiopathic intracranial hypertension (IIH). Carbamazepine discontinued and upon treatment with oral Acetazolamide 250 mg two times a day, the patient noted spontaneous resolution of her hemifacial spasm.

Ophthalmologic examination by Goldman perimetry showed mild bilateral blind spot enlargement. Three days later, lumbar puncture was re-performed showing decrease of CSF pressure by 32. At 6 months follow-up, the patient remained asymptomatic. She was maintained on acetazolamide 250 mg twice daily. Her neuro-ophthalmologic examination was normal. Several months later, acetazolamide was tapered without evidence of recurrence.

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DISCUSSION

The characteristic features of IIH include sub-acute headache, visual loss, which can be irreversible if not treated promptly. Less common 6th nerve palsy and rarely other cranial nerve involvement might be reported^{7,8,9}. The mechanism of cranial nerve palsies in the context of IIH still remains controversial. Nevertheless, an acceptable hypothesis for any cranial nerve palsy associated invokes pressure-induced intracranial structural shifts. The coincidence of hemifacial spasm and IIH is even more speculative¹¹.

The first report of hemifacial spasm as a manifestation of IIH was reported on 1994 by Aki K. Selky. He reported a 46 year old woman with a 2 months history of headache and right hemifacial spasm with papilledema on her examination who with were eventually diagnosed with IIH¹².

In the following, on 2010 Charles Druckman reported a 47 year old obese woman who presented with right hemifacial spasm and blurred vision with superimposed headache who were treated upon typical IIH treatment¹³. The last available of coincidence of IIH and hemifacial spasm was reported by S Muzerengi on 2012 who reported a 33 year old obese woman with sub-acute headache and subsequent hemifacial spasm¹⁴.

Despite to previous cases, in our patient the sole complaint was a chronic hemifacial spasm without any prodromal sign of IIH. The IIH suspicion has raised based on the incidental ophthalmologic findings.

CONCLUSION

Idiopathic intracranial hypertension (IIH) is a challenging disorder of intracranial hypertension in the absence of an intracranial mass, hydrocephalus, or other identifiable etiology with normal cerebrospinal fluid content. The final diagnosis of this illness is by Dandy's modified criteria.

The increase CSF pressure may lead to a vast majority of manifestations even the compression of facial never resulting to hemifacial spasm which occurred in our patient.

Conflicts of interest Disclosure: The authors declare that they have no conflict of interests.

Ethical statement: Informed consent was obtained by the patient for publication of the case report

REFERENCES:

1. Paybast S, Assarzagdegan F, Lima BS, Koosha M.

- Relationship of Opening CSF Pressure and Visual Field Defect in Idiopathic Intracranial Hypertension. *Int Clin Neurosci J.* 2018;5(2):51–4.
2. Digre KB, Nakamoto BK, Warner JEA, Langeberg WJ, Baggaley SK, Katz BJ. A comparison of idiopathic intracranial hypertension with and without papilledema. *Headache.* 2009 Feb;49(2):185–93.
 3. Friedman DI, Liu GT, Digre KB. Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children. *Neurology.* 2013 Sep;81(13):1159–65.
 4. Durcan FJ, Corbett JJ, Wall M. The incidence of pseudotumor cerebri. Population studies in Iowa and Louisiana. *Arch Neurol.* 1988 Aug;45(8):875–7.
 5. Sorensen PS, Krogsaa B, Gjerris F. Clinical course and prognosis of pseudotumor cerebri. A prospective study of 24 patients. *Acta Neurol Scand.* 1988 Feb;77(2):164–72.
 6. Baheti NN, Nair M, Thomas S V. Long-term visual outcome in idiopathic intracranial hypertension. *Ann Indian Acad Neurol [Internet].* 2011;14(1):19–22. Available from: <https://www.ncbi.nlm.nih.gov/pubmed/21633609>
 7. Wall M, Kupersmith MJ, Kieburz KD, Corbett JJ, Feldon SE, Friedman DI, et al. The idiopathic intracranial hypertension treatment trial: clinical profile at baseline. *JAMA Neurol.* 2014 Jun;71(6):693–701.
 8. Digre KB. Idiopathic intracranial hypertension. *BMJ [Internet].* 2010 Jul 7;341:c2836. Available from: <http://www.bmj.com/content/341/bmj.c2836.abstract>
 9. Jensen RH, Radojicic A, Yri H. The diagnosis and management of idiopathic intracranial hypertension and the associated headache. *Ther Adv Neurol Disord.* 2016 Jul;9(4):317–26.
 10. Czyz CN. Botulinum Toxin Treatment of Benign Essential Blepharospasm (BEB) and Hemifacial Spasm. In: *Operative Dictations in Ophthalmology [Internet].* Cham, Switzerland: Springer; 2017. p. 549–51. Available from: <https://books.google.com/books?id=bWi8DgAAQBAJ>
 11. Soiberman U, Stolovitch C, Balcer LJ, Regenbogen M, Constantini S, Kesler A. Idiopathic intracranial hypertension in children: visual outcome and risk of recurrence. *Childs Nerv Syst.* 2011 Nov;27(11):1913–8.
 12. Selky AK, Purvin VA. Hemifacial Spasm An Unusual Manifestation of Idiopathic Intracranial Hypertension. *J Neuro-Ophthalmology [Internet].* 1994;14(4). Available from: https://journals.lww.com/jneuro-ophthalmology/Fulltext/1994/12000/Hemifacial_Spasm_An_Unusual_Manifestation_of.3.aspx
 13. Druckman C, Nguyen T-V. Hemifacial Spasm as an Unusual Manifestation of Pseudotumor Cerebri. *Optom Am Optom Assoc [Internet].* 2010 Jun 1;81(6):288. Available from: <https://doi.org/10.1016/j.optm.2010.04.044>
 14. Muzerengi S, Moor C, Davies M. Facial spasm and headaches: should we call it "IIH-Spasm syndrome?" *J Headache Pain [Internet].* 2013/02/21. 2013;14(Suppl 1):P225–P225. Available from: <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3620183>.