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Idiopathic Intracranial Hypertension without Papilloedema "Case Report"

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Abstract

Purpose: The purpose of this case report is to share and spread the awareness of diagnosing and managing idiopathic intracranial hypertension in atypical scenarios as in this case, who presented with multiple other medical conditions, and had no papilloedema. The presenting signs and symptoms of mere headache were misleading. The other medical conditions could have explained the patient's headache. These included cervical spine osteoporosis, and middle ear otolith disease. This case is unique because she was found to have IIH without papilloedema, and despite having multiple cancers in the uterus, breasts, and recently the thyroid, she had no malignant cells in the CSF. The patient was treated for IIH, and her malignancies were treated separately by the corresponding physicians. **Method:** The patient was assessed in 2019, and was followed up closely till 2021, in collaboration with her neurologist, oncologist, and orthopedist. **Result:** The patient had IIH after her neurologist attempted a lumbar puncture, which showed high CSF pressure. **Conclusion:** Headache is a common symptom presented by patients of different ages and backgrounds. The absence of papilloedema does not exclude the diagnosis of IIH. When possible etiologies for headache are ruled out (e.g. middle and inner ear infections, anemia, intracranial tumors, glaucoma, etc.), chronic headaches that do not respond to medications, and present with visual field changes may require a lumbar puncture.

Keywords

Idiopathic Intracranial Hypertension, Papilloedema, Lumbar Puncture, Case Report

1. Introduction

The Term Idiopathic intracranial hypertension (IIH or pseudotumor cerebri syndrome) is a simple and accurate term that describes a condition that most

commonly occurs in overweight women of childbearing age, where no other specific cause is evident [1]. Classically, patients with this condition suffer from an occipital headache that is associated with postural variability, valsalva maneuver, and classically occurs in mornings. Patients may also complain of tinnitis, nausea, vomiting, and, or diplopia. There is typically papilloedema, and a raised intracranial hypertension is evident on lumbar puncture. The rarity of this case is due to the absence of such papilloedema, atypical headache, and the presence of multiple other medical conditions.

This case report is about a 50 years old female patient who was referred to the neurophthalmology clinic in September 2019, to evaluate her headache. She had 5 days history of vortex and occipital headache, which was associated with dizziness and blurred vision, but was not related to changes in posture. It was not relieved by analgesics or sleep. She did not complain of diplopia, transient visual obscurations, nausea or vomiting. Regarding tinnitus, she mentioned a history of otoliths disease (canalithiasis) in her left ear, however, it was treated with Epplys maneuver and vestibular rehabilitation sessions.

The patient gave a past history of carcinoma of the uterus and breasts for which she was operated on twice in 2013 and 2015. She did not receive any chemotherapy or radiotherapy. She also has cervical disc prolapse, for which operation was offered by the treating orthopedic doctor, but the patient refused it.

2. Clinical Findings

On examination, her best corrected visual acuity (BCVA) was 20/25 (with +0.5 sph) in the right eye, and 20/25 (with +0.75/0.25/100˚) in the left eye. The Ishihara colour vision test showed 4/13 and 11/13 for the right and left eye, respectively. Both pupils were round, reactive and regular with no relative afferent pupillary defect (RAPD). The anterior segment exam was clear and did not show any evidence of uveitis in either eye. Her intra-ocular pressure (IOP) was 18 mmHg in both eyes. The posterior segment exam did not show evidence of vitritis, retinitis, or any other retinal pathology. Both optic discs had clear margins, present venous pulsations, normal vascular tortousity, with no signs of papilloedema, and both maculae were within normal limits.

3. Timeline

The patient had had an MRI brain done in 2018, indicating that her headache symptom is far more chronic that the history given, and it is still present despite treating her ear condition.

4. Diagnostic Assessment

The OCT of both optic nerves supported the absence of papilloedema. The C/D ratio was 0.48 and 0.61 in the right and left eye, respectively (**Figure 1** and **Figure 2**). The OCT of the macula was within normal limits in both eyes (**Figure 3** and **Figure 4**). The visual field test showed enlarged blind spot and nasal field

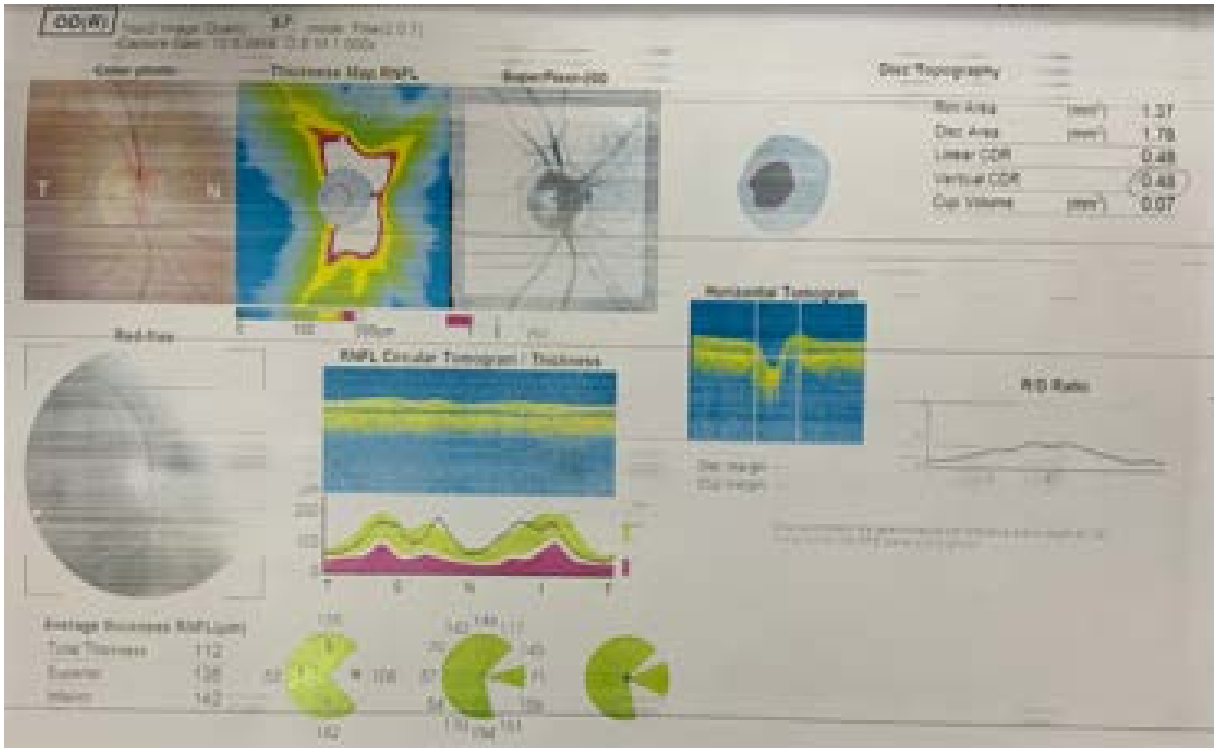

Figure 1. OCT of the right optic nerve, showing normal retinal nerve fiber layer thickness (RNFL) in both eyes.

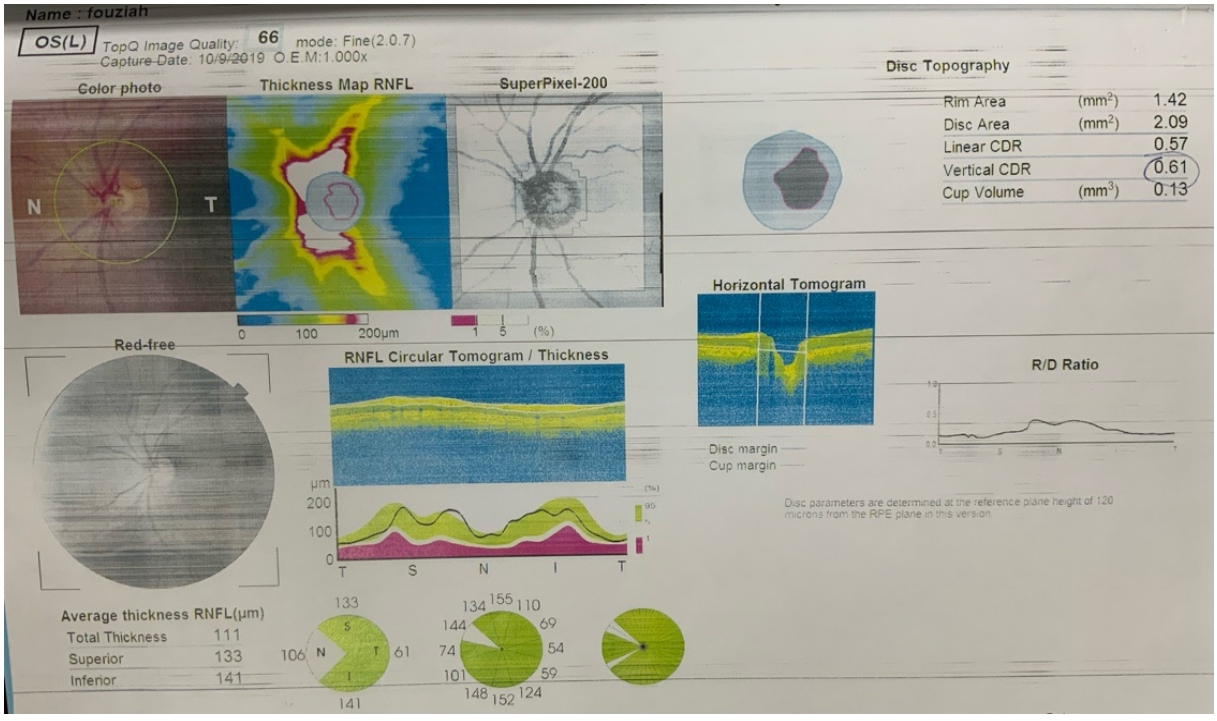

Figure 2. OCT of the left optic nerve shows normal RNFL thickness. This indicated the absence of papilloedema.

constriction in the right eye, while the left eye showed superior arcuate and a small nasal field defect, both of which do not correspond to a specific neurological cause (Figure 5 and Figure 6).

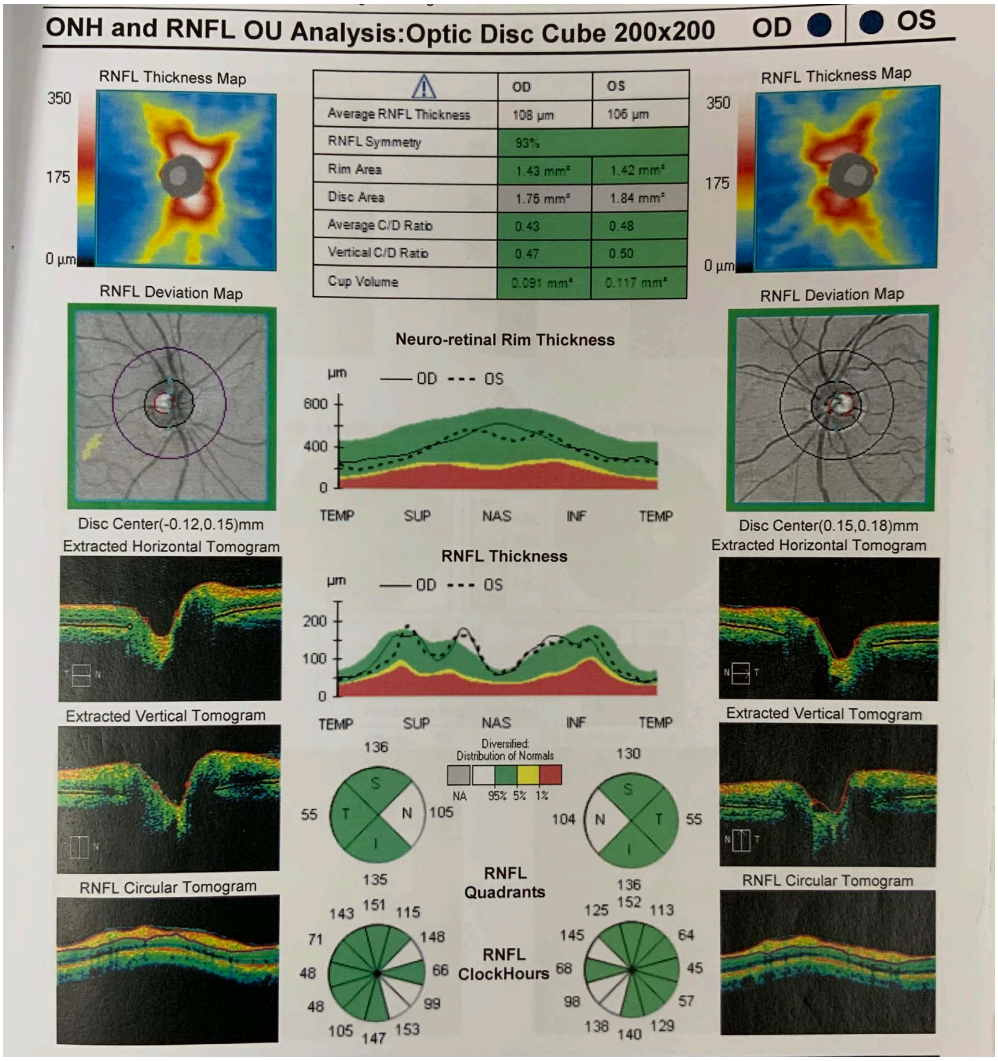

Figure 3. This figure shows the normal ganglion cell layer analysis in both eyes.

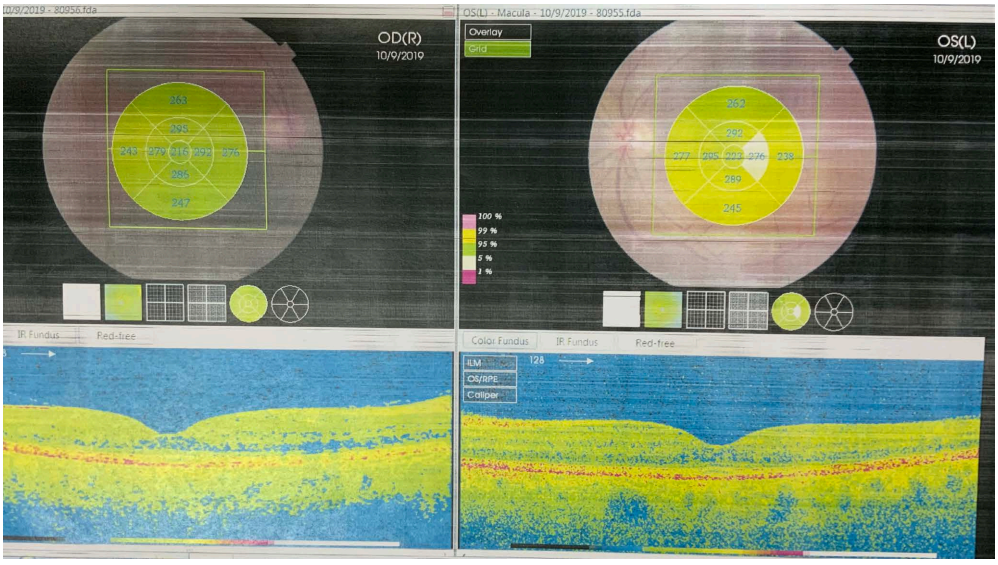

Figure 4. This figure shows the normal macular thickness and contour in both eyes.

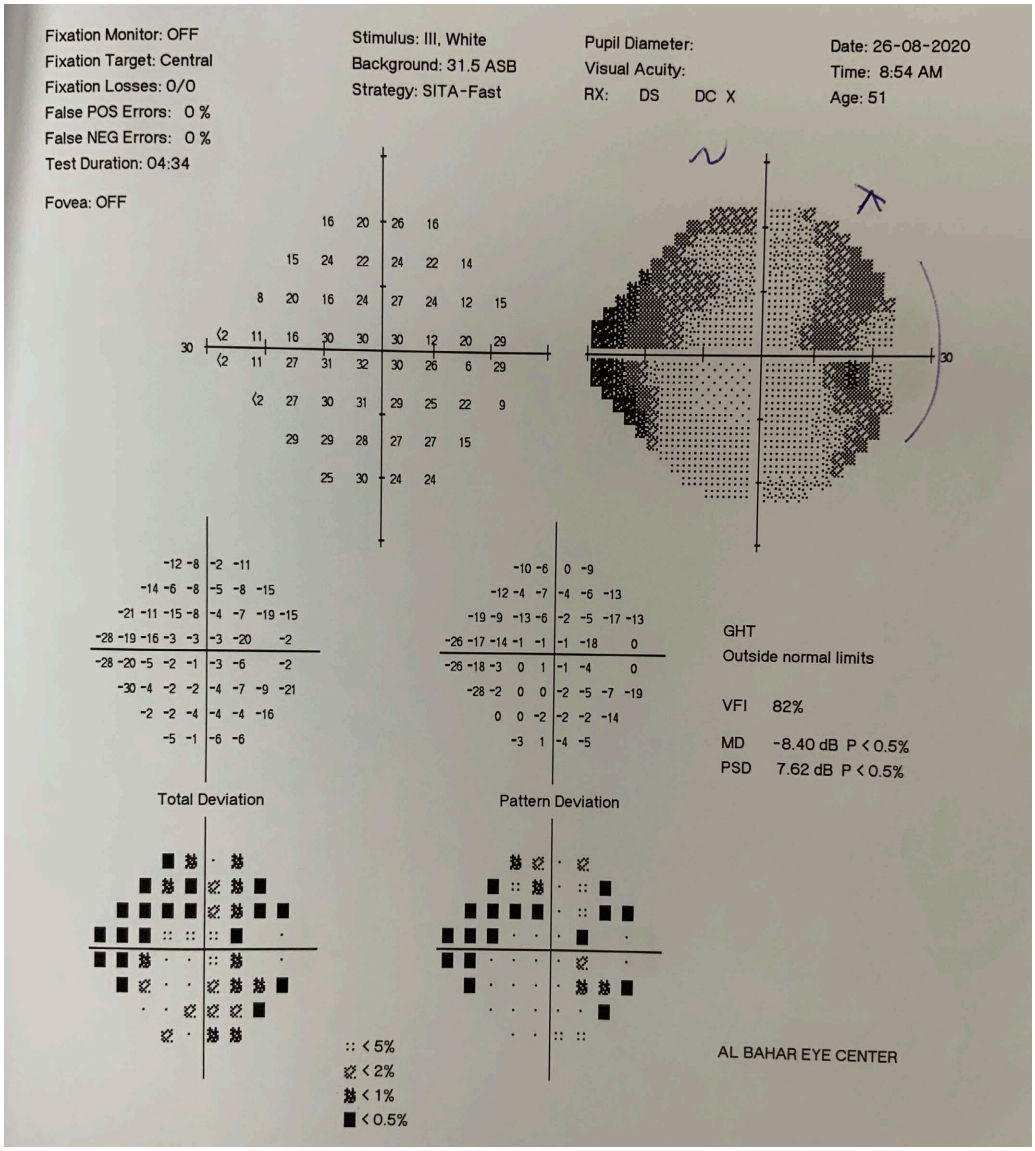

Figure 5. The central 24-2 Humphrey visual field test of the right eye, showed enlarged blind spot and nasal field constriction.

The Patient had done several neuroimagings, all of which showed an empty sella, otherwise were remarked as normal studies (**Figure 7**). The last MRI, MRV, and MRA of the brain were done in November 2019. Both MRV and MRA were normal and cavernous sinus thrombosis was ruled out (**Figure 8**). There was increased cerebrospinal fluid (CSF) around the optic nerves associated with empty sella turcica, which suggested an increase in intracranial pressure (**Figure 9**).

5. Therapeutic Interventions

The patient was started on antiglaucoma medications for the possibility of normal tension glaucoma. Moreover, there was a possibility that her headache was related to her cervical disc prolapse, yet, the patient refused again any orthopedic intervention.

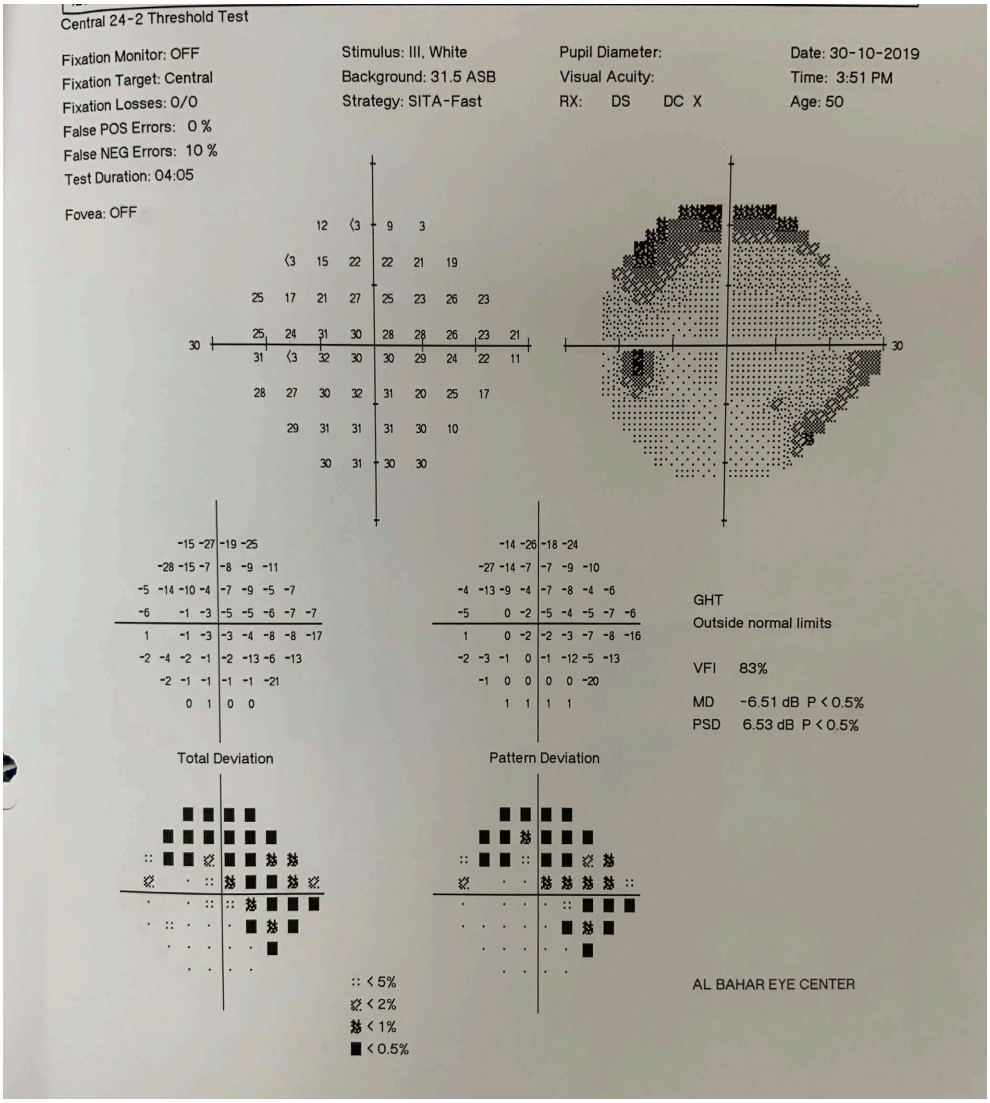

Figure 6. The central 24-2 Humphrey field test of the left eye showing superior arcuate field defect along with a small peripheral nasal field defect.

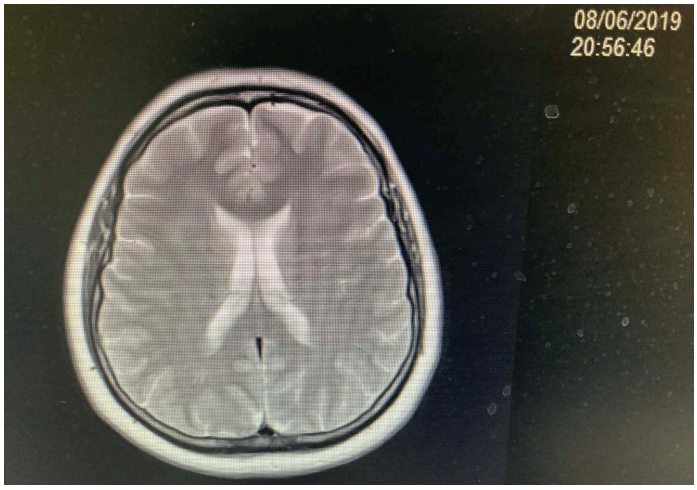

Figure 7. This figure shows a normal MRI study.

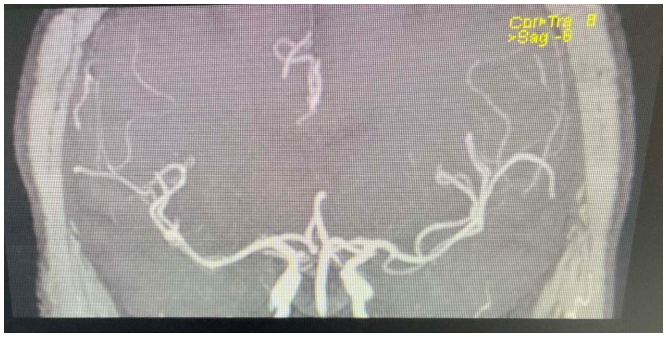

Figure 8. This figure shows a normal MRV study.

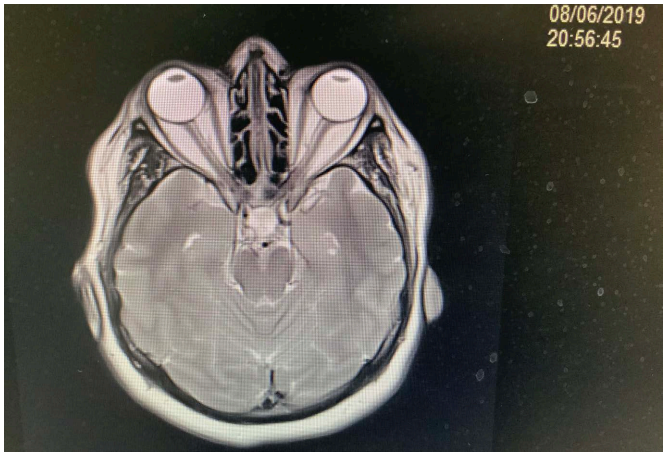

Figure 9. This figure shows increased CSF spaces around the optic nerves.

Several tests were done and included PCRs for Enterovirus, HSV1, HSV2, CMV qualitative PCR, EBV, and VZV. There were all negative. Biochemistry profile showed high levels of chloride, calcium and adjusted calcium, Uric acid and cholesterol levels. Otherwise, CBC, ESR, and thyroid function tests were all normal.

On the 18th of August 2020, a lumbar puncture was done on the patient by her treating neurologist, as a further measure in managing her persistent headache and given the MRI findings. The opening pressure was 33 cm H_2O, and was closed on a pressure of 20 cm H_2O. Her headache had only partially improved after the LP. CSF analysis showed colourless CFS, clear, with no red blood cells (NIL RBC) or white blood cells (NIL WBC). CSF glucose level was 3.6 mol/L, which was less than 80% of her plasma glucose. CSF protein level was normal. The patient was started on Diamox 500 mg twice per day, and was advised to reduce her body weight.

Upon her follow ups in the neurology department, she was still complaining of headache, and was taking daily Solpadeine. She was suspected to have medication overuse headache. Detoxification therapy was offered, but she refused. She was advised to stop Solpadeine and to continue Topamax treatment. Botox therapy for headache was given to her as well.

During her neuro-ophthalmology follow ups, she consistently showed no op-

tic nerve head swelling. She neither tolerated Diamox nor Topamax. Moreover, her continuous headache had persuaded her neurologist to perform another lumbar puncture, which showed an opening pressure higher than 33 cm H_2O. Accordingly, shunting procedure was offered, however, the patient refused any surgical intervention.

Recently, an MRI of the neck showed an enlarged left lobe of the thyroid gland, with large cystic nodule. Final cytologic diagnosis of the nobule of the left lobe of the thyroid was suspicious for papillary carcinoma.

6. Follow ups and Outcomes

The patient's headache was partially, but not totally relieved. She is currently following up in the neurology clinic for her headache, and with the endocrinologist for the thyroid mass. Given her past history of cancers, this patient needs to be followed up regularly for intracranial metastasis.

7. Discussion

The strength of this case report is in the rarity of the condition, the multiple other medical conditions that are managed through multidisciplinary team. The limitations include the failure to fully resolve the patient's headache, and the poor image quality of the OCT of the optic nerve.

Idiopathic Intracranial hypertension (IIH or pseudotumor cerebri syndrome) is a common condition that most commonly occurs in overweight women of childbearing age, however, it may also occur in both genders, and in any age. Characteristically, there is raised intracranial pressure with papilloedema, a normal brain parenchyma on neuroimaging, and no cause is found for the raised intracranial pressure.

MRI signs of high intracranial pressure appears much before the papilloedema develops. Moreover, papilloedema may disappear much later compared to MRI findings after the intracranial pressure is controlled.

Reference [2] showed the modified Dandy Criteria for IIH, which included the following: 1) Signs and symptoms of IIH. 2) Normal neurological exam. 3) Normal neuroimaging apart from evidence of increased CSF pressure (*i.e.* empty sella turcica, optic nerve sheath with filled CSF spaces and smooth walled non flow-related venous sinus stenosis or collapse), and >200 mm water of CSF pressure with otherwise normal neurodiagnostic studies. An abnormal neuroimaging prompts another diagnosis. 4) Awake and alert patient. 5) No other cause of increased intracranial pressure present.

In the presence of equivocal CSF opening pressures of 200 - 250 mm water, at least one of the following is required: 1) Pulse synchronous tinnitus; 2) 6th cranial nerve palsy; 3) Frisée Grade II papilloedema; 4) Negative echo graphs for drusen, and no other disc anomalies mimicking disc edema present; 5) MRV showing lateral sinus stenosis, preferably using ATECO technique (auto-triggered elliptic centric-ordered three dimensional MR venography); 6) Partially empty sella on

coronal or sagittal views and optic nerve sheaths with filled out spaces on T2 weighted axial scans [2].

Secondary causes of IIH were suggested to be called as they are: e.g. Vitamin A-induced intracranial hypertension, or steroid withdrawal-related intracranial hypertension, and so on. It was also proposed that the term pseudotumor cerebri is not needed [2].

Interestingly, it has been found that bilateral transverse sinus stenosis evident on MR venography predicts IIHWOP in migraine patients. Furthermore, it is suggested that bilateral transverse sinus stenosis on MR venography is associated with increased intracranial pressure in the absence of papilloedema in patients with headache mimicking chronic tension type headache [3].

In the absence of papilloedema or 6th cranial nerve palsy, with persistent headache refractory to prophylactic therapies, the differential diagnosis should involve chronic headaches and migraines, as well as IIHWOP (Idiopathic intracranial hypertension without papilloedema). Although the diagnosis may be challenging, an LP is needed to diagnose IIHWOP, along with at least 3 of the neuroimaging signs of high CFS pressure, including empty sella, distention of the perioptic subarachnoid space with or without a tortuous optic nerve, flattening of the posterior sclera and transverse venous sinus stenosis) [4].

In conclusion, chronic headache is a challenging symptom to manage, especially with the possible etiologies that cause it, like migraine, anemia, brain tumors…, etc. IIH without papilloedema is a rare condition that should be suspected in patients with chronic headache whom are unresponsive to treatments. To diagnose IIH without papilloedema, a high opening pressure on lumbar puncture of ≥250 mm H_2O and at least three signs if IIH on neuroimaging are needed.

Patient's Perspective

The patient was aware of the rarity of her condition, as was told by the other physicians she was seen by, and is fully aware of our interest to share her case.

Informed Consent

The paper does not include any recognizable photos of the patient.

Acknowledgements

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Conflicts of Interest

There are no financial conflicts of interest to disclose.

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