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Indian Journal of Clinical and Experimental Ophthalmology

Journal homepage: www.ijceo.org

Case Report

Idiopathic intracranial hypertension, PCOS & hypothyroidism in adolescent

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ARTICLE INFO

Article history:

Received 19-08-2020

Accepted 29-08-2020

Available online 31-03-2021

Keywords:

Idiopathic Intracranial Hypertension

VI Nerve Palsy

Hypothyroidism

ABSTRACT

An 18-year old Obese female with a history of hypothyroidism & PCOS presented with complains of headache for 5 days associated with nausea and double vision for 2 days. Her neurological examination together with Cranial Nerves was Normal except for bilateral restriction of abduction, suggestive of bilateral abducens nerve involvement. Fundus findings revealed bilateral grade III disc edema. Lumbar puncture indicated increased opening pressure and MRI was normal, which further ruled out the other Causes for Raised Intracranial Pressure. Patient was treated with IV mannitol and then switched over to oral acetazolamide. Patient improved over 1 week. Patient was further followed with serial visual field tests.

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1. Introduction

Idiopathic Intracranial Hypertension (IIH) is identified by raised intra-cranial pressure (ICP) that by definition does not have any identifiable cause. It predominantly involves obese adult Females of childbearing age, but can involve all age groups. Patients may present with blurring of vision, nausea, headache, double vision, etc. without underlying space occupying lesions of central nervous system. Pseudotumor cerebri and benign intracranial hypertension (BIH) are the terms interchangeably used for IIH, but IIH is the preferred term.

2. Case History

An 18 years old female, obese (weight- 100kgs) with BMI- 32.65kg/m² presented with severe throbbing headache associated with nausea for past 5 days and double vision for past 2 days. Patient is a known case of hypothyroidism for last 7 months- On treatment (tab. Thyroxine 50mcg OD). Also a known case of poly cystic ovarian syndrome for last 6 months- on tab. metformin 1000mg qHS. She had no comorbidities like systemic hypertension, Diabetes, etc.

On initial evaluation, she was afebrile and hemodynamically stable. On neurological examination, she was fully alert, conscious and oriented, having fluent speech and normal comprehensive abilities. Signs of meningeal irritation were absent. Evaluation of cranial nerves revealed bilateral VI nerve palsy while remainder of cranial nerves were functionally normal.

On ocular examination, visual acuity in both the eyes was 6/9 improving to 6/6 with pin hole, colour vision was unaffected, extra-ocular motility was partially restricted in abduction in both the eyes & anterior segment was within normal limits. Intraocular pressure (IOP) was found to be 16 and 14 mm Hg respectively in right & left eyes. Fundus examination revealed established disc edema with obscuration of major vessels as they leave the disc in all 4 Quadrants. Modified Frisen's grade III disc edema.

Magnetic resonance imaging (MRI) with magnetic resonance venography (MRV) studies of brain were normal.

Opening pressure on lumbar puncture was 250 mm H₂O, which was above the normal limit. CSF study was within normal Limits (protein- 0.25 gm/Lit, CSF glucose- 0.43 gm/Lit, 3 white cells/mm³ and lymphocytes 100%).

Patient was advised head end elevation to 30°. Physician & gynaecologist opinions were taken and the treatment

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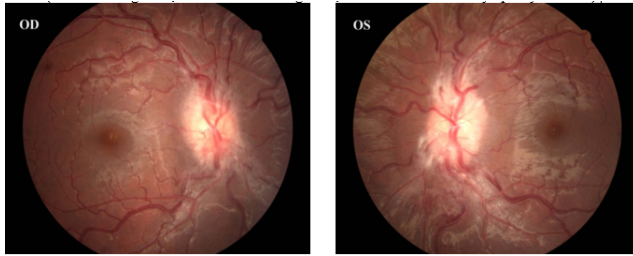


Fig. 1: Both eye fundus showing modified frisen grade III disc edema

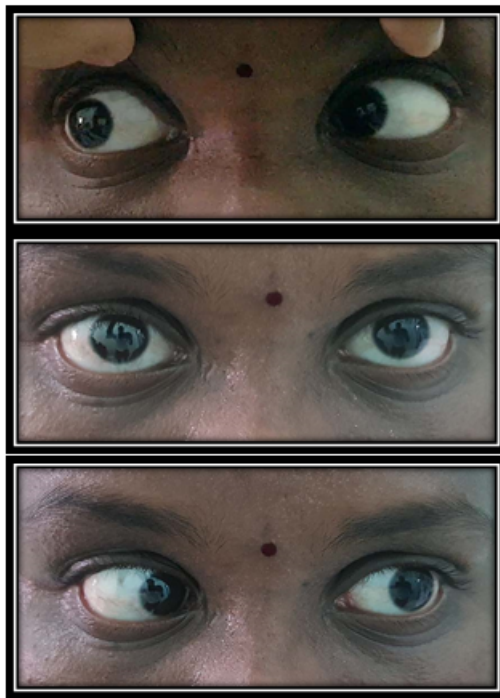


Fig. 2: Bilateral abduction restriction of extraocular movements

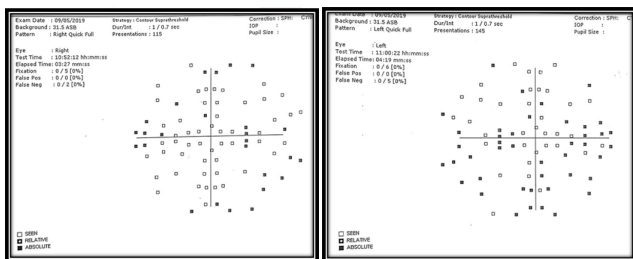


Fig. 3: Both eyes visual fields showing enlarged blind spot and peripheral constriction of fields

for hypothyroidism & PCOD were continued. As per neurologist, Inj. IV Mannitol 200 ml stat & then 100ml IV Q8hourly for 3 days alongside monitoring of input/output. After 3 days, patient was switched from IV medications to Tab. acetazolamide by gradually increasing the dosage.

On discharge, patient was advised weight reduction & the list of drugs which have potential of worsening the condition were to be avoided.

On follow up visit at 1 week, patient was symptomatically better and the complains of headache and diplopia had resolved. Extra ocular movements restriction improved gradually. Fundus examination revealed bilateral resolving disc edema. Repeat visual field testing showed some improvement.

On follow up visit at 1 month, patient was symptom free. Normal extra ocular movements were suggestive of reversal of VI cranial nerve palsy. Fundus examination of both eyes showed complete resolution of the disc edema. Repeat visual field testing revealed Improvement. Patient was further followed up with serial visual field tests.

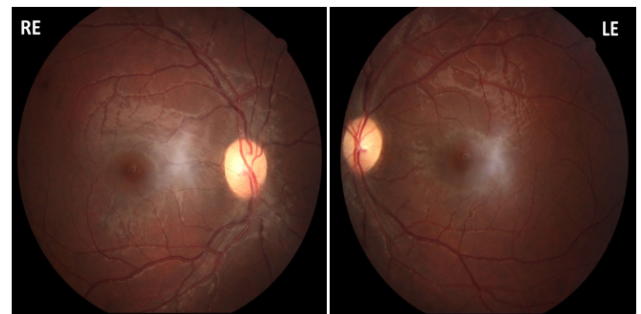


Fig. 4: Resolved disc edema on follow up visit at 1 month

3. Discussion

The diagnosis of IIH with bilateral VI nerve palsy was suspected following detection of bilateral disc edema and bilateral restriction of abduction in presence with headache associated with Nausea & diplopia. She fulfilled all the diagnostic criterias of IIH in a patient with papilledema such as elevated intracranial pressure, bilateral 6th nerve palsy, increased lumbar puncture opening pressure, normal CSF studies & normal neuroimaging study which further ruled out the other etiologies of elevated intracranial pressure. Moreover, our patient was an obese woman fulfilling the expected profile for IIH.

Idiopathic intracranial hypertension (IIH) is defined by the modified Dandy criteria which is as follows:

1. Symptoms and signs of increased intracranial pressure;
2. No localized neurologic signs, except for unilateral or bilateral 6th nerve palsies;

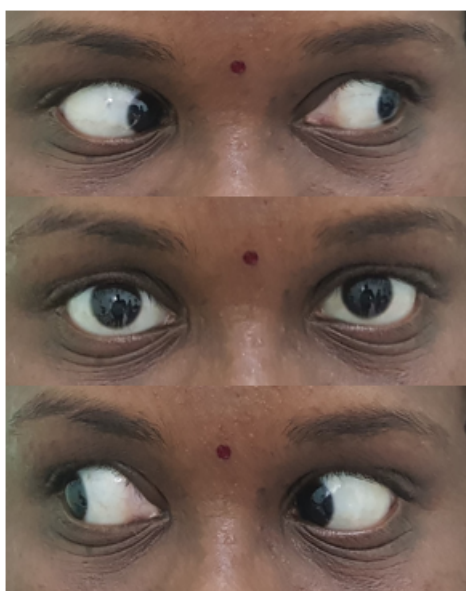


Fig. 5: Complete improvement of abduction restriction on follow up visit at 1 month

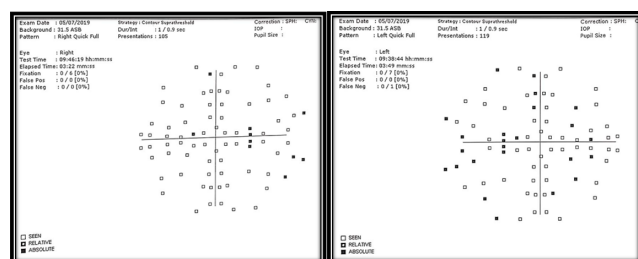


Fig. 6: Visual fields showing improvement in both eyes on follow up visit at 1 month

3. Increased CSF opening pressure >200 mmH₂O, but normal CSF composition;
4. No evidence of hydrocephalus, mass, structural, or vascular lesions on imaging; and
5. No other cause of increased intracranial pressure (ICP) identified¹

The incidence of IIH is 0.9/100,000 individuals annually. It rises to 3.5/100,000 in females aged 15–44, and to 19.3/100,000 in women of age 20–44 years who were having body weight 20 % or more above their ideal.²

The rise in prevalence of PCOS in women with IIH may highlight PCOS-driven morbid obesity, which further leads to development of IIH. Increased intraabdominal and cardiac filling pressures, in association with obesity, may lead to a retrograde increase in CSF pressure.³

The symptoms most commonly reported by IIH patients include headache (94%), transient visual obscuration (68%), pulsatile tinnitus (58%), photopsia (54%), double vision (38%), vision loss (30%) and retrobulbar pain on eye

movements (22%).⁴

Paralysis of Abducens nerve (10–13%) is seen associated with IIH, although oculomotor, trochlear, trigeminal, facial, glossopharyngeal and hypoglossal nerve palsies have been also reported in isolated case presentations.⁵

Papilloedema is an important feature of elevated intracranial pressure and its presence is seen in approximately 97% of cases of IIH. In absence of papilledema, 6th nerve palsy would be essential for the definite diagnosis of IIH.⁶ Interestingly, the patient had bilateral papilloedema and bilateral abducens nerve paralysis both.

In patients with IIH of an adolescent age group, VI Cranial Nerve Palsy was found more often in those with higher BMI & raised CSF Opening Pressure.⁷

The pathophysiology behind cranial nerve palsies coexisting with IIH is poorly understood. The probable mechanism for the Abducens nerve involvement seems to be due to stretching of the nerve across the apex of petrous bone.⁸

Regarding the medical treatment, the first-line drug for management of IIH in children & adolescents is Acetazolamide, a carbonic anhydrase inhibitor. But the treatment duration is not standardised; it is therefore adapted based on the clinical symptoms and the progression of visual acuity, with a mean range of 2 to 6 months.² Unresponsive cases of idiopathic intracranial hypertension can be managed with either optic nerve fenestration, lumboperitoneal shunting or Transverse Dural Sinus stenting. IV mannitol or a lumbar Puncture are usually reserved for Acute Severe exacerbations.

4. Conclusion

IIH must be strongly suspected in young obese women presenting with headache & transient visual complaints. The diagnosis of IIH remains that of exclusion and neuroimaging studies should be always performed to rule out other structural and obstructive lesions.

Our aim was to report the possibility of idiopathic intracraial hypertension presenting with bilateral CN VI palsy in an adolescent patient with poly cystic ovarian syndrome, obesity & hypothyroidism.

Patients with IIH may have associations like obesity, polycystic ovary syndrome, thyroid disorders, addison's disease, anemia, blood-clotting disorders, kidney disease, lupus, sleep apnea, underactive parathyroid glands and should not be ignored while diagnosing and managing the case of idiopathic intracranial hypertension in children.

5. Source of Funding

None.

6. Conflict of Interest

None.

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Cite this article: Vansadia AB, Gnanadurai SC, Waris SAN. Idiopathic intracranial hypertension, PCOS & hypothyroidism in adolescent. *Indian J Clin Exp Ophthalmol* 2021;7(1):171-174.