Idiopathic Intracranial Hypertension with Minimal Ocular Signs

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I. BACKGROUND

Idiopathic intracranial hypertension (IIH) is characterized by increased intracranial pressure due to an unknown cause. The hallmark ocular sign is papilledema which may lead to progressive optic atrophy and subsequent vision loss. IIH is seen most frequently in obese women of childbearing age. Other potential risk factors include certain medications such as tetracyclines, steroids, oral contraceptive pills, vitamin A, gout, nephropathy, and acne medication. Common symptoms include headache, transient blurred vision, double vision, and pulsatile tinnitus. IIH is a diagnosis of exclusion, so an MRI should be run to rule out an intracranial mass or other secondary causes. Once imaging has ruled out other etiologies, a lumbar puncture should be completed to confirm the diagnosis. An opening cerebrospinal fluid (CSF) pressure greater than 25 cm H2O with a normal composition is diagnostic for IIH. First-line therapy for patients that are overweight is weight loss. Medical therapy includes acetazolamide, a carbonic anhydrase inhibitor that reduces the rate of CSF production. Other medical therapies if acetazolamide is insufficient or not tolerated well include topiramate or furosemide. Discontinuation of any potentially contributory medications should also be considered. Surgical treatments are reserved for refractory cases and include optic nerve sheath fenestration and CSF diversion procedures.

II. HISTORY

A 32-year-old Caucasian female presented to the Veterans Hospital with a chief complaint of shooting eye pain OS more than OD, exacerbated when sneezing or coughing. She also had been experiencing headaches for the past year that have increased in intensity over the last two months. She endorsed bilateral tinnitus that has worsened with the onset of her other symptoms. Her last eye exam was two months prior and was told that her symptoms were likely due to an ocular migraine. She reported the pain and headaches were affecting every aspect of her life and taking 400 mg of ibuprofen every morning only mildly relieved her symptoms.

Medical history: dysfunctional voiding, GERD, allergic rhinitis, history of alcohol abuse, PTSD, tobacco user, female stress incontinence

Ocular history: hyperopia OD, regular astigmatism OU, choroidal nevus OD

Medications: loratadine, medroxyprogesterone inject subcutaneously every 3 months, omeprazole

BMI: 23.5

III. EXAM FINDINGS

Corrected distance VA: 20/20 OD, OS, OU

Pupils: PERRL (±APD)

EOMs: Full, smooth, and unrestricted OD, OS

Confrontation visual fields: full to finger movement OD, OS

Anterior segment: unremarkable

Appplanation tonometry: OD: 14 mmHg, OS: 14 mmHg

Posterior segment: Optic nerve is flat with distinct margins, and a small C/D ratio <0.10 OD with mild nasal elevation without vessel obscuration and possible Papill’s folds nasally OU. No visible drusen was seen at the optic nerve head OU. See Figure 1.

IV. DIFFERENTIAL DIAGNOSIS

Mild papilledema

Pseudo-papilledema

Ocular migraine

Intracranial mass

V. NEURODIAGNOSTIC STUDIES

Lab results: normal WBC, HGB, HCT, PLT, BUN, CRE, GLU, TSH, PTH, ALP, AMY, U/L, S/CAB

MRI of brain with and without contrast

- No acute intracranial lesion

- No- enhancing mass identified

- No fibrous or cystic mass

- No intracranial fluid collection

- No midline shift

Magnetic resonance venography (MRV)

- Opening pressure: 26 cm H2O

- Closing pressure: 18 cm H2O

- Composition: normal

VI. DIAGNOSIS AND MANAGEMENT:

A diagnosis of mild papilledema OU secondary to idiopathic intracranial hypertension was made after confirming that there was no intracranial lesion or other pathology on MRI, and an elevated opening pressure with lumbar puncture. The patient was started on acetazolamide with gradually increasing dosages each week. At the 1000 mg dosage, the patient had difficulty tolerating the medication and reported severe hand tingling, diarrhea, nausea, weakness, dizziness, and headaches, and was considered lightheaded. Her dosage was decreased to 750 mg which was the maximum dosage she was able to tolerate. The patient reported complete resolution of symptoms since starting acetazolamide. She elected to self-discontinue her medroxyprogesterone contraceptive.

VII. DISCUSSION

Idiopathic intracranial hypertension typically occurs in young, obese women, but cases outside this demographic do exist. A thorough case history highlights the importance of the optometrist’s role in advocating for these patients. Bilateral optic nerve edema can present in both mild and severe forms. A thorough case history is important as many reports have found oral contraceptives associated with IIH, but less consensus between depot medroxyprogesterone acetate contraception and IIH has been established. Despite this, increased awareness and research of this relationship is necessary as more women enter childbearing age. Other potential risk factors include certain medications such as tetracyclines, steroids, oral contraceptive pills, vitamin A, gout, nephropathy, and acne medication. Increased awareness and research of this relationship is necessary as more women enter childbearing age. Other potential risk factors include certain medications such as tetracyclines, steroids, oral contraceptive pills, vitamin A, gout, nephropathy, and acne medication.

VIII. REFERENCES


