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Unilateral Papilledema Due to Idiopathic Intracranial Hypertension - A Case Report

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Abstract

Idiopathic intracranial hypertension is a disorder of elevated cerebrospinal fluid pressure of unknown cause. It usually occurs in women in the childbearing years. There is no evidence of deformity or obstruction of the ventricular system and neuro-diagnostic studies are otherwise normal except for increased cerebrospinal fluid pressure. In addition, no secondary causes of intracranial hypertension can be found. We present a case of Idiopathic intracranial hypertension in a young obese female presenting as unilateral papilledema.

Keywords: Idiopathic Intracranial hypertension; Obesity.

Introduction

Idiopathic intracranial hypertension (IIH) is a common cause of unilateral papilledema without any other focal deficits. Although classically bilateral, unilateral and highly asymmetrical papilledema is rarely described in IIH. This atypical presentation can pose a diagnostic challenge if the treating physician is unaware of such association. Unilateral swelling of the optic disc is most frequently caused by a local optic nerve or intraocular pathology. Truly unilateral papilledema caused by intracranial hypertension is rare and poses a diagnostic dilemma.

Case Report

A 25-year-old obese female with no significant past medical history presented to ED complaining of itchiness and dryness of both eyes for 2 days. She denied any changes in vision or photophobia. She also developed redness of both eyes, right-sided headache, and right facial numbness starting one day ago.

She mentioned that she has chronic on and off right sided headache for a couple of years associated with photophobia and phonophobia. Review of systems was negative including

but not limited to tinnitus, ear pain/discharge, vertigo, weakness of limbs, chest pain, shortness of breath, palpitations, Raynaud's phenomenon, rashes, joint pain or any changes in bowel or bladder habits. She has no known allergies. She is a non-smoker and doesn't do any drugs. She drinks alcohol occasionally. She is not on oral contraceptive pills or any other medications. There is no family history of autoimmune or neurological diseases.

Physical examination revealed obese female with BMI of 37.3 kg/m² but was otherwise unremarkable. Laboratory investigations revealed normal cell counts. Serum chemistry showed mildly elevated creatinine but was otherwise normal.

Ophthalmology was consulted in ED and she was found to have right sided papilledema. CT scan of the head and MRI brain done subsequently were unremarkable. There was no evidence of hydrocephalus or intracranial space occupying lesion. She was admitted to the medical floor and neurology was consulted.

Lumbar puncture was done which revealed opening pressure of 515 mm of water. CSF analysis revealed cell count 2, CSF protein 21 mg/dL, CSF glucose 64 mg/dL, and CSF gram stain were negative. She was diagnosed to have Benign Intracranial Hypertension (Pseudotumor Cerebri) and was started on Acetazolamide as per neurology recommendations. Vasculitis workup including p-ANCA, c-ANCA, and dsDNA was negative, but ANA was positive. Complement levels were normal. Anti-Smith, anti-cardiolipin antibodies, and lupus anticoagulant were negative. HIV and RPR were negative.

The patient was discharged on the third day and she followed up in Neurology Clinic 2 weeks later when she stated that her headache has resolved, and she feels much better.

Discussion

Idiopathic Intracranial Hypertension (IIH) is defined as an elevated intracranial pressure but no clinical, laboratory or radiological evidence of hydrocephalus, infection, tumor or vascular abnormality. The Modified Dandy criteria describe clinical, laboratory and radiological findings required for a diagnosis of IIH. The criteria required are 1) symptoms and

signs of increased intracranial pressure (e.g. papilledema and headache), 2) CSF pressure > 250 mm of water in lateral decubitus position, 3) No localization signs except for sixth nerve palsy, 4) normal CSF composition, 5) normal-to-small (slit) ventricles on imaging with no intracranial mass, 6) no unexplained symptoms or signs, 7) exclusion of other causes on specific forms of imaging in, particular, MRI/venography should be included to rule out intracranial venous sinus thrombosis [1].

IIH usually occurs in obese women in childbearing years [2]. The symptoms of increased intracranial pressure are a headache, pulse-synchronous tinnitus (pulsatile tinnitus), transient visual obscurations and visual loss. Signs of IIH are diplopia due to sixth cranial nerve paresis and papilledema with its associated loss of sensory visual function [2]. The term pseudotumor cerebri or IIH is commonly used for a condition characterized by bilateral papilledema and absence of hydrocephalus and space-occupying lesions [3]. Unilateral papilledema is one of the unusual manifestations of IIH [4]. The true incidence of unilateral papilledema is unknown. In a review study by Bruntse, 25 out of 1346 patients had unilateral papilledema [5]. Another study by Frederick showed 6 out of 26 patients had unilateral or highly asymmetric papilledema [6]. There are some case reports on unilateral papilledema due to IIH [7-9].

The actual reason for unilateral papilledema in IIH is unknown but there is some hypotheses. It was postulated that the optic nerve sheath anomaly protects the optic nerve from increased intracranial pressure. Huna-Barn et al. reported 11 cases of unilateral papilledema and found no gross differences between the optic nerve sheaths on CT or MRI [9]. The possibility of differences in lamina cribrosa of two optic discs is another hypothesis [7]. Another study found that experimental elevated intracranial pressure causes an axoplasmic blockage at the level of lamina cribrosa [10].

Intracranial hypertension in the absence of an intracranial mass lesion or hydrocephalus can be caused by an alteration of one or more of the four determinants of cerebrospinal fluid (CSF) pressure: (1) Intrasagittal sinus pressure, (2) Resistance of arachnoid villi to the egress of CSF, (3) Rate of production of CSF, and (4) Compliance of the CSF space. The pseudotumor

cerebri syndrome of obese young women may be caused partially by increased CSF production [9-11].

Conclusion

Unilateral papilledema due to idiopathic intracranial hypertension is uncommon. Early identification and treatment of IIH is crucial to prevent complications. We report this case to make clinicians aware of unilateral papilledema as a potential presentation of idiopathic intracranial hypertension.

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