Case Reports in Neurology

Case Rep Neurol 2017;9:49–53

DOI: 10.1159/000466685
Published online: March 21, 2017

Case Report

Uncommon Presentation of Idiopathic Intracranial Hypertension in a Patient with Polycystic Ovary Syndrome: A Case Report

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Keywords
Case report · Idiopathic intracranial hypertension · Neurology · Ophthalmology

Abstract
Introduction: Idiopathic intracranial hypertension is a rare condition characterized by increased intracranial pressure without clinical, laboratory, or radiological evidence of intracranial pathology. Early management could prevent irreversible outcomes. ***Case Presentation:*** A 17-year-old single Arabian female of Arab origin presented with a 2-day complaint of horizontal diplopia and transient visual obscurations. She denied any history of headache or decreased vision. The patient was diagnosed with polycystic ovary syndrome a year prior to presentation. Examination revealed bilateral moderate papilledema and limited left eye abduction. However, visual acuity and fields were normal. Increased intracranial pressure was confirmed by lumbar puncture opening pressure (550 mm H_2O). The cerebrospinal fluid composition and imaging of brain and cerebral venous system were normal. The diagnosis of idiopathic intracranial hypertension was confirmed and the patient was treated with acetazolamide 500 mg twice per day. The symptoms totally resolved within 3 days and the

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papilledema disappeared after 2 months. ***Conclusion:*** Awareness of such uncommon presentation of idiopathic intracranial hypertension emphasizes the critical importance of detailed ophthalmic examination and shows the good prognosis of early management.

Introduction

Idiopathic intracranial hypertension (IIH), referred to as "pseudotumor cerebri," is a condition characterized by increased intracranial pressure without clinical, laboratory, or radiological evidence of intracranial pathology such as ventriculomegaly, venous thrombosis, malignancy, or underlying infection [1, 2]. IIH is a rare condition with an annual incidence of 1 per 100,000 populations [3, 4]. The incidence is much higher among women in childbearing age and those who are overweight or obese, which constitutes more than 90% of diagnosed cases [5, 6]. Typical symptoms include headache and transient visual loss, which occurs in 85–90% of the cases, followed by tinnitus, diplopia, and rarely no symptoms [6, 7]. Family history of IIH has been reported in only a few cases [8]. Additionally, IIH has been linked to certain medical conditions such as polycystic ovary syndrome [9]. We are reporting here the diagnosis and the management of IIH in a patient with polycystic ovary syndrome.

Case Report

A 17-year-old single female of Arab origin presented to the emergency department with a 2-day history of double vision in primary gaze and when looking to the left side. The double vision would disappear when looked to the right side or upon closing one of her eyes. She was referred to neurology for further evaluation. Further questioning revealed she had multiple episodes of bilateral transient visual obscuration during the last week. Those episodes were short lasting (few seconds) and precipitated by changing position from lying to sitting or standing. She also reported mild left shoulder pain for 1 week prior to her presentation. The patient denied any history of headache, decreased visual acuity, photophobia, vomiting, tinnitus, dizziness, neck pain, loss of sensation, or loss of motor functions. She had menstrual irregularities and was diagnosed with polycystic ovary syndrome a year earlier and was started on metformin for that reason. She denied taking any other medications including tetracycline, vitamins, hormones, cosmetic supplements, or herbs. She was nonsmoker, denied recent trauma, and denied family history of similar complaints.

On examination, she was conscious and oriented with normal vital signs. She was lying comfortably on the bed, with no pain or distress. Body mass index was 31. Chest and heart were normal and abdomen was soft, not tender with no palpable organomegaly, or masses. Higher mental function, visual acuity, and visual fields were normal. Fundus examination revealed bilateral moderate papilledema with obscuration of major retinal vessels (Frisén grade 3). She had esotropia with limited left eye abduction, but the right eye movement was normal. Vertical and intorsion movements were normal in both eyes. There were no signs of affection of other cranial nerves. Motor, sensory, and cerebellar examinations were unremarkable as well as deep tendon reflex and gait. Computed tomography and computed tomographic venography were normal with no masses or structural abnormalities. Lumbar puncture was done while the patient was relaxed in the lateral decubitus position with hips

and legs extended. The opening pressure was 550 mm H_2O. Examination of cerebrospinal fluid (CSF) composition showed normal cell count and differential, glucose, and protein. Other laboratory investigations including complete blood counts, electrolytes, blood glucose, liver functions, renal functions, and thyroid stimulating hormone were normal. After ruling out secondary causes, the diagnosis of IIH was confirmed and the patient was started on acetazolamide 500 mg twice a day. She reported full recovery of her double vision and visual obscuration within 3 days after lumbar puncture and treatment initiation. The patient was maintained on acetazolamide 1,000 mg twice a day after discharge and decreased to 500 mg twice a day after normal fundus examination on the second month of follow-up. During 1 year of follow-up, the patient had no admissions and never had recurring symptoms. Fundus examination showed mild papilledema 1 month after discharge, which became normal with no papilledema at 2 months and 8 months after discharge. However, mild bilateral papilledema without symptoms were seen at 12 months after discharge as the patient stopped the medication by her own and was instructed to resume acetazolamide 500 mg twice a day and to continue follow-up.

Discussion

The diagnosis of this unique case of IIH was suspected after detecting bilateral papilledema in the absence of headache. She fulfilled all other diagnostic criteria of IIH in a patient with papilledema such as increased intracranial pressure, normal CSF analysis neuroimaging study showing no etiology for intracranial hypertension, and normal examination [2]. Additionally, possible other causes of intracranial hypertension such as cerebral venous abnormalities, relevant medication intake, thyroid disease, anemia, and renal failure have been excluded [2, 10]. Moreover, our patient was an obese woman fulfilling the expected profile for IIH [6, 8].

We believe that our case had an uncommon presentation for several reasons. The lack of headache, which is considered the most common initial symptom of IIH being seen in approximately 90% of patients [6], and second, diplopia is an uncommon presenting complaint in IIH patients [11]. The diplopia in our patient might be due to unilateral left abducens nerve palsy, which led to horizontal displacement of images [12]. Finally, only 6% of patients with IIH were having evidence of polycystic ovary syndrome [9].

Papilledema is an important sign of increased intracranial pressure and is present in approximately 97% of IIH cases [13]. Should papilledema be absent, abducens nerve palsy would be required for a definite diagnosis of IIH [12]. Interestingly, our patient had both bilateral papilledema and unilateral abducens nerve palsy. As papilledema can lead to profound visual loss, it is great to perform detailed ophthalmic examination including perimetry for early assessment and management of papilledema [12].

The patient's symptoms (including diplopia, visual obscuration, shoulder pain, and abduction nerve palsy) had totally resolved few days after lumbar puncture and acetazolamide initiation. Lumbar puncture has been shown to be effective, but the relief is often transient [11]. As papilledema regressed on follow-up while the patient was maintained on acetazolamide, we reconfirm that acetazolamide is a safe and effective drug which is considered the first-line treatment for IIH [11].

In conclusion, we are reporting an uncommon presentation of IIH in a patient with polycystic ovary syndrome. This case may increase the awareness of uncommon presentations of

IIH that can pass unrecognized, emphasize the critical importance of detailed ophthalmic examination for and show good prognosis with early acetazolamide therapy.

Acknowledgements

We would like to thank nurses and medical staff in neurology ward at King Abdulaziz Medical City for their assistance in the care of this patient.

Statement of Ethics

Appropriate ethical approval has been obtained from the Neurology Department at King Abdulaziz Medical City. Written informed consent was obtained from the patient's legal guardian(s) for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Disclosure Statement

The authors declare that they have no competing interests.

Authors' Contributions

Both authors made the diagnosis and managed the patient. Anas Albarrak did the lumbar puncture. Anas Albarrak wrote the manuscript. Suleiman Kojan revised and edited the manuscript. Both authors read and approved the final manuscript.

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