Asymmetric Papilledema in a Patient with a Pituitary Microprolactinoma, a Clinical Twist

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Abstract

A 33 year old woman with a known pituitary microprolactinoma was referred to our clinic for visual field evaluation. On examination and subsequent testing she was found to have asymmetric bilateral disc elevation. Visual field testing revealed generalized constriction in the right eye and a full visual field in the left eye. On follow-up magnetic resonance imaging (MRI), the pituitary prolactinoma was stable in size (8 mm) without impingement on the visual pathways or extension into the cavernous sinus. Magnetic resonance venography (MRV) did not reveal any intracranial venous occlusion or sinus stenosis. Subsequent lumbar puncture revealed an opening cerebrospinal fluid (CSF) pressure of 250 mmH2O with normal CSF composition, which clinched the diagnosis of idiopathic intracranial hypertension (IIH).

Keywords: Disc Edema; Papilledema; Pituitary; Adenoma; Microadenoma; Prolactinoma; Microprolactinoma; Idiopathic Intracranial Hypertension

Introduction

Optic nerve elevation is an unusual presentation in patients with pituitary microadenomas. We report an asymmetric case of papilledema secondary to IIH in a patient with a pituitary microprolactinoma. This case is clinically important because the history of pituitary prolactinoma as a primary pathology can hinder and delay further investigations in order to find a secondary pathology to explain the patient’s vision loss, particularly considering the fact that unilateral papilledema is uncommon in IIH. The patient was properly treated for IIH that saved the remaining of her vision. However, she lost some peripheral vision which did not recover with treatment. It is conceivable that if she did not have pituitary prolactinoma her further investigation with lumbar puncture was performed sooner that could save her vision.

Case Report

A 33 year-old woman was referred to our clinic with complaints of a visual disturbance in her right eye. She had been previously diagnosed with a pituitary microprolactinoma when she initially presented a year prior with increasing headaches and nausea. On MRI brain she was found to have an 8 mm suprasellar lesion consistent with a pituitary microadenoma without impingement on the optic nerves or chiasm or extension into the cavernous sinus. The patient was referred to an endocrinologist who found an elevated prolactin level of 111 µg/L (normal: < 20 µg/L) and diagnosed her with a pituitary microprolactinoma. She was started on treatment with bromocriptine.

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Later bromocriptine was substituted for cabergoline due to intolerance, and the patient was also treated with dexamethasone. During the course of this treatment, the patient's prolactin level decreased to 49 µg/L, but her morning cortisol level increased to 1237 nmol/L (normal: 140 - 690 nmol/L).

On history the patient continued to complain of headaches, which had increased in frequency to once a week, and also reported a new shadow in the peripheral vision of her right eye. She admitted to gaining approximately one hundred pounds over the last couple of years. She denied transient visual obscurations, diplopia, pulsatile tinnitus or other neurological symptoms. Her past medical history was significant for social anxiety, panic attacks, and depression, which had progressively worsened and were being treated with sertraline and clonazepam. Her past surgical, ophthalmic, and social histories were unremarkable.

On examination the patient weighed 152 kg (BMI: 48.5) and her blood pressure was 120/76 mmHg. Her uncorrected vision was 20/20 bilaterally, pupils were equal and reactive to light and accommodation with no relative afferent pupillary defect (RAPD) noted, and the intraocular pressures were 12 mmHg in both eyes. She had full extraocular movements and was orthophoric in primary position and in all cardinal positions of gaze. Trigeminal nerve function was preserved bilaterally. No dyschromatopsia was reported on red saturation test and she correctly identified all plates on Ishihara pseudosochromatic testing in both eyes. Slit lamp examination revealed normal anterior segments. On dilated fundus examination the patient had slight optic disc atrophy with gliosis and blurred margins in the right eye, and acute, florid disc edema in the left eye (Figure 1). The optic disc cups were effaced in both eyes and a protruding disc was noted in the left eye without Paton’s lines or choroidal folds. The vessels were slightly attenuated in the right eye and were of normal caliber in the left eye with absent spontaneous venous pulsations bilaterally. The vitreous, macula, and periphery were unremarkable in both eyes.

**Figure 1:** Asymmetric bilateral disc edema with atrophic changes and gliosis in the right eye and florid disc edema in the left eye.

On visual field testing (Figure 2), the patient had a circumferential visual field defect sparing fixation in the right eye with blind spot enlargement in a pattern reminiscent of advanced glaucoma. The visual field in the left eye was full without evidence of blind spot enlargement (Figure 2). Ocular coherence tomography (OCT) examination of the optic discs revealed decreased average retinal nerve fiber layer (RNFL) thickness in the right eye (50 µm) consistent with chronic atrophy, and supranormal average RNFL thickness in the left eye (179 µm) consistent with the acute, florid disc edema previously noted on examination (Figure 3). Her neurological examination was unremarkable.

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Given the above findings a diagnosis of asymmetric papilledema was entertained and the patient was referred to a neurosurgeon. An urgent brain MRI/MRV with sella protocol was performed to rule out interval growth of the pituitary prolactinoma. MRI revealed a stable

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pituitary microadenoma (Figure 4). MRV revealed no evidence of intracranial venous occlusion or transverse sinus stenosis. It was decided to proceed with lumbar puncture, as the relatively small size of the lesion posed minimal risk of uncal herniation. The patient was referred to a neurologist for the lumbar puncture who also ruled out focal neurological signs. The opening CSF pressure was 250 mmH\textsubscript{2}O and the CSF composition was normal, clinching the diagnosis of papilledema secondary to IIH as per the modified Dandy criteria [1,2]. At this juncture the patient was treated with acetazolamide and weight loss to which she responded well. Her visual fields slightly improved in the right eye and stayed stable in the left eye; they are currently being monitored on a quarterly basis.

Discussion and Conclusion

Prolactinomas are the most frequently occurring pituitary tumors [3]. Those greater than 10 mm, or macroprolactinomas, may cause different visual field defects depending on the location of the lesion and the anatomy of the perichiasmal region. Mass effect on the overlying visual pathways may result in a central visual field defect, junctional scotoma, visual field defects respecting the vertical midline in at least one eye (i.e. monocular temporal or bitemporal visual field defect), complete vision loss in one or both eyes, or a combination of these. In our case, visual field analysis was key to the diagnosis of papilledema as the cause of the patient's visual disturbance. The patient's constricted visual field in the right eye with preservation of her central vision steered the diagnosis away from mass effect on the optic nerves or chiasm. It also ruled in intracranial hypertension secondary to obesity and/or hormonal dysregulation.

While the optic disc appearance in this case was reminiscent of Foster-Kennedy syndrome, this was quickly ruled out on visual field analysis and subsequent neuroimaging. Rather, the appearance of bilateral, albeit very asymmetric, disc edema in this case may represent changes related to anatomical variation in the optic nerve subarachnoid trabeculations representing pseudo Foster-Kennedy syndrome. Anatomical variation between the two optic nerves at the level of the subarachnoid trabeculations may result in differential ICP transfer to the perilaminar area as initially described by Hayreh [4]. This area is particularly susceptible to changes in ICP for several reasons. It constitutes the area where ganglion cell axons make an acute right turn from the retina into the optic nerve. Within the optic canal, ganglion

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cell fibers are crammed into their narrowest configuration within the optic nerve. This is also a vascular watershed area where the retinal and choroidal circulations meet. Hence, an increased number of subarachnoid trabeculations may be protective against papilledema due to decreased transfer of ICP to the particularly vulnerable optic nerve head [4,5]. Interestingly, the patient did not have an RAPD in the right eye despite her markedly asymmetric visual fields and right optic disc atrophy. This was presumably due to impaired optic nerve function in the left eye secondary to axoplasmic flow stasis in the setting of acute, florid disc edema.

Given the relatively small size of the pituitary microprolactinoma, it is unlikely that its mass significantly contributed to the patient’s increased ICP. This is why a decision to proceed with lumbar puncture was made after the lesion was found to be stable in size on MRI. Rather, one can ascribe the patient’s increased ICP to her body habitus and/or to hormonal dysregulation of the hypothalamic pituitary axis caused by the microprolactinoma and/or its ensuing treatment. As such, a mechanism of increased ICP due to impaired absorption of CSF at the level of the arachnoid granulations is most likely in this case. It was interesting to note that shortly around the time of the patient’s referral to the Ophthalmology service she was being treated with steroids and had gained a significant amount of weight. Her morning cortisol and prolactin levels three months prior to her ophthalmology assessment were more than twice the upper limit of normal value. Her blood pressure during this time remained within the normal range, which ruled out malignant hypertension as a cause of her disc edema. She also denied being on medications associated with IIH. An association between hypercortisolemia and intracranial hypertension has been well documented [6-9]. Obese patients have been shown to have increased 11β-hydroxysteroid dehydrogenase type 1 (11β-HSD1) CSF activity, which may affect CSF absorption through the arachnoid granulations and result in intracranial hypertension [6-8]. A definite link between hyperprolactinemia and intracranial hypertension has been more elusive [10] as other reported cases of increased ICP associated with prolactinomas have also been confounded by abnormalities in cortisol levels [11].

Although rather uncommon, this case underscores the importance of considering a diagnosis of IIH in overweight patients with pituitary microadenomas. While mass effect on the optic pathways [12], Foster Kennedy syndrome [13] and interval growth of the pituitary adenoma resulting in a space occupying lesion and/or obstructive hydrocephalus [14-16] are important considerations when evaluating a patient with a similar presentation to this case, increased ICP secondary to obesity and/or hormonal disturbances must also be considered. In this context, visual field analysis remains an invaluable diagnostic and monitoring tool. Prompt recognition and treatment of this condition may be vision saving. In our patient, visual field became stable after we initiated proper IIH treatments but her visual field loss in the right eye did not completely recover.

It is conceivable that if the patient was sent for proper eye examination earlier, her vision in the right eye could have been saved. Therefore, we recommend that clinicians must have a low threshold to investigate for a second pathology in patient with suprasellar lesions if there is evidence for vision loss and symptoms beyond what can be merely explained by the established pathology that was pituitary microprolactinoma in our patient.

Disclosure

The authors have no proprietary or financial interest concerning products or instruments described.

Bibliography


