Optic Nerve Edema in a Patient with a Pituitary Tumor

Mallory McLaughlin, O.D.

A patient presents with unilateral optic nerve head edema and an inferior nasal visual field defect. Magnetic resonance imaging yields a pituitary tumor. Proposed mechanisms of pathophysiology of this unusual presentation are discussed.

CASE HISTORY

- Demographics: 66 year old African American male
- Chief complaint: Hazy vision in the morning upon wakening OU, relief with warm compresses
- Past ocular history:
  - Diabetic without retinopathy OD, OS
  - Non-visually significant cataracts OD, OS
  - Vitreoretinal Traction OD
  - History of spark from M16 rifle hitting the left eye during military service, treated and patched. Resolved.
- Past medical history:
  - History of bladder cancer 2007, s/p transurethral resection, radical prostatectomy, pelvic lymph node dissection
  - Type II Diabetic, last HbA1C 7.5%
  - Hypertension
  - Gunshot Wound to the right thorax and right leg in 1969
- Medications:
  - Levothyroxine
  - Sildenafil
  - Glipizide
  - Losartan
  - Atorvastatin
  - Aspirin

PRESENTING EXAM: March 2016

- Clinical:
  - Visual Acuities: 20/25- OD, OS
  - Pupil testing: No afferent pupillary defect OD, OS
  - Confrontation visual fields: Full to finger counting OD, OS
  - Color vision normal OD, OS (Ishihara)
  - Early sectoral optic nerve edema versus crowded optic nerve head OS
  - Normal optic nerve appearance OD
FURTHER TESTING ORDERED:

- Imaging:
  - B-Scan OS: not conclusive for optic nerve head drusen
  - Fluorescein Angiogram OS: Nasal optic nerve head hyperfluorescence
- Goldmann visual field testing:
  - Essentially full OD, OS
- Humphrey visual field testing:
  - Essentially full OD, OS
- Laboratory studies:
  - STAT ESR and CRP—within normal limits
  - ANA, syphilis screen, quantiferon gold, ACE, lysozyme- all within normal limits
- Radiology:
  - Enhancing 2.1 cm mass centered in the sellar region. The mass appears to abut or be in close proximity to the cavernous carotid arteries and the optic chiasm. Differential considerations include a pituitary macroadenoma amongst other etiologies.

FOLLOW-UP EXAM: August 2016

- Clinical:
  - New complaint of blurred vision in the inferior nasal quadrant only OS x 3 weeks
  - Visual Acuities: 20/20- OD, OS
  - New onset afferent pupillary defect OS
  - New onset inferior nasal confrontation visual field defect OS
  - Color vision normal OU (Ishihara)
  - Crowded left optic nerve with indistinct margins and increased edema now 360 degrees, greater inferior
  - Normal optic nerve appearance OD

DIFFERENTIAL DIAGNOSIS

- Unilateral papilledema secondary to increased intracranial pressure
- Compression of the cavernous carotid artery by the pituitary tumor
- Compression of a unilateral optic nerve by a lobulated pituitary adenoma or due to post-fixed chiasm
- Incidental finding of an asymptomatic pituitary tumor with unrelated optic nerve head edema
- Diabetic papillopathy
- Optic nerve sheath meningioma
- Hypertensive optic neuropathy
- Central retinal vein occlusion
- Optic neuritis
- Pseudo-papilledema
  - Optic nerve head drusen
Congenital anomalous appearance of the optic nerve head

- Inflammatory infiltration of optic nerve (tuberculosis, sarcoid, metastasis, leukemia etc)
- Posterior uveitis (syphilis, sarcoid, etc)

**DISCUSSION**

The classic ocular presentation of a pituitary tumor is optic nerve head pallor and bitemporal hemianopic visual field defects\(^1\text{-}\(^3\). The bitemporal visual field defects are typically most dense superiorly. These sequelae are secondary to compression of the nasal retinal ganglion cell axons as they cross in the optic chiasm causing optic atrophy due to retrograde axonal degeneration\(^4\text{-}\(^5\). However in this case, the left optic nerve head was edematous and the patient had a unilateral inferior nasal visual field defect. A closer look at imaging and careful consideration of the pathophysiology was necessary in this seemingly paradoxical presentation.

When evaluating the location of a visual field defect caused by a pituitary tumor, one must carefully consider the anatomy of the optic chiasm and its relationship to the pituitary gland. In 78% of patients, the optic chiasm is superior to the pituitary gland and diaphragm sellae. If the chiasm is located on the tuberculum sellae, it is classified as “pre-fixed”. If the chiasm is located overlying the dorsum sellae, it is classified as “post-fixed”. Pre-fixed chiasm and post-fixed chiasm occur in 5% and 17% of patients respectively\(^6\). If the chiasm is post-fixed, a large pituitary tumor growing superiorly may compress the optic nerve anterior to the optic chiasm. Additionally, a lobulated tumor may compress an individual optic nerve rather than the optic chiasm. If the patient in this case were to have a post-fixed chiasm or a lobulated tumor compressing the optic nerve, it may account for his unilateral visual field defect. However, neither of these theories were supported by MRI imaging. Even if imaging were consistent with those theories, the patient would likely have a superior visual field defect and optic nerve head pallor.

Papilledema secondary to increased intracranial pressure is an important differential diagnosis in this case. Papilledema is most often bilateral, although unilateral cases have been observed. Transfer of increased cerebral spinal fluid pressure to the optic nerve head requires an intact optic nerve sheath. Asymmetric congenital or acquired abnormalities in the optic nerve sheath have been proposed as potential etiologies of unilateral papilledema. If a mass compresses the intracranial optic nerve, it may compromise the transfer of fluid to the optic nerve head. It is possible that the pituitary tumor in this case asymmetrically compressed the right intracranial optic nerve, rather than the optic chiasm, explaining the normal appearance of the right optic nerve head. Asymmetries between the bilateral venous sinuses and lamina cribrosas have also been postulated as etiologies of unilateral papilledema\(^7\text{-}\(^8\). Regardless, it is unlikely for pituitary tumors to cause increased intracranial pressure due to their small size and supratentorial position\(^3\). Pituitary tumors with extra-sellar extensions have been known to cause hydrocephalus if the tumor extends posteriorly impeding the flow through the third ventricle\(^9\). There were no radiological or symptomatic findings in this patient consistent with this explanation.

Cavernous invasion by the pituitary tumor and compression of the left cavernous carotid artery could be a potential cause of ischemia and thus edema of the optic nerve head. Of all pituitary tumors,
6-10% percent invade the cavernous sinus\textsuperscript{10,11}. In all cases, invasion is unilateral\textsuperscript{10,12}. Unfortunately, magnetic resonance imaging only has 55% sensitivity in detecting cavernous sinus invasion pre-operatively\textsuperscript{13}. Often, the diagnosis of invasion is made intra-operatively. In this case, the lumen of the bilateral cavernous carotid arteries and the size of the bilateral ophthalmic arteries were visually symmetrical. However, computed tomography angiography or surgical exploration would be necessary to rule out this potential pathophysiology with certainty.

It must also be considered that the pituitary tumor was an asymptomatic incidental finding and the optic nerve head edema was unrelated to the pituitary tumor. Hypertensive optic neuropathy, central retinal vein occlusion, optic neuritis, pseudo-papilledema, inflammatory infiltration of optic nerve and posterior uveitis were ruled out as differential diagnoses by the patient’s history, diagnostic testing, signs and symptoms. Idiopathic non-arteritic anterior ischemic optic neuropathy (NAION), optic nerve sheath meningioma, and diabetic papillopathy are proposed etiologies of the unilateral optic nerve head edema if we suppose that it is unrelated to the pituitary tumor.

In a retrospective study of 3,235 patients with diabetes, diabetic papillopathy was diagnosed in 24 eyes (0.37%). Of those 24 cases, 50% were unilateral and the average recovery rate of the optic nerve head edema was 7.8 ± 3.7 months\textsuperscript{13}. In this case, the patient had no other signs of diabetic retinopathy. However, diabetic papillopathy can occur in the absence of diabetic retinopathy. Only 63% of the patients with diabetic papillopathy in the aforementioned study had diabetic retinopathy\textsuperscript{13}. Non-arteritic anterior ischemic optic neuropathy is an unlikely etiology because of the duration of the worsening optic nerve head edema. NAION typically resolves within 6-11 weeks\textsuperscript{14,15}. In this case, the edema increased in severity over 5 months without resolution.

Although not visualized by the MRI in this case, optic nerve sheath meningioma is an important differential diagnosis of optic nerve head edema. Optic nerve sheath meningiomas present unilaterally in 95% of cases and with optic nerve head edema in 48% of cases. Common symptoms experiences are loss of visual acuity (96%), visual field defects (83%) and proptosis (59%)\textsuperscript{16}. The location of visual field defects is dependent on the location of the tumor. Although unlikely, it is possible that the MRI sections through the optic nerve were not thin enough to positively image an optic nerve sheath meningioma.

**MANAGEMENT**

After the initial presentation of the patient and MRI images revealed a pituitary tumor, the patient was referred to the neurosurgery department at the University of Illinois at Chicago for surgical consideration. When optometric follow-up examination revealed worsening of the optic nerve head edema, new onset afferent pupillary defect and new onset visual field defect, the neurosurgeon was contacted directly and informed of the new findings. Repeat Goldmann visual field testing will be obtained. Computed tomography angiography and lumbar puncture with opening pressure could be considered as academic endeavors.

If surgical management of the mass should be considered, endoscopic transsphenoidal resection is the most common surgical method for pituitary masses. The surgery has a low morbidity and mortality rate\textsuperscript{17}. If the optic chiasm is compressed, visual recovery of symptoms is achieved in 70-80% of cases\textsuperscript{18,5}.
However, should the pituitary tumor have invaded the cavernous sinus and compressed the internal carotid artery, the surgical difficulty and morbidity and mortality rate will be increased\textsuperscript{11}. In some cases, medical treatment or radiation may also be considered\textsuperscript{2} and hormone replacement therapy may be necessary in cases of hypopituitarism\textsuperscript{17}.

To monitor ocular and visual health, the patient will followed at regular intervals with visual acuity, visual field, color vision, ocular motility, and dilated fundus examinations.

CONCLUSION

The paradoxical presentation of this case reminds the clinician to carefully consider and rule out all differential diagnoses before arriving at a diagnosis of exclusion, such as NAION. Clinical cases do not always present with the most classic, academic, predicable presentation. Correlation of clinical picture with anatomy and pathophysiology of every neuro-ophthalmic case is imperative.

REFERENCES


