Funny Optic Nerves

Dr. Connolly is a 2014-2015 Pediatric/Binocular Vision Resident at the IU School of Optometry. She received her Doctorate of Optometry from Ferris State University. Dr. Connolly will be joining IUSO as a Pediatric/Binocular Vision Clinical Assistant Professor upon completion of her residency.

Dr. Miller is a 2014-2015 Contact Lens Resident at the IU School of Optometry. He received his Doctorate of Optometry from Ferris State University and completed his Bachelor’s degree in Chemistry at Alma College. Dr. Miller will be joining a private practice in Petoskey, Mich., upon completion of his residency.
Idiopathic Intracranial Hypertension (IIH) in a Child

Katie Connolly, O.D.
Pediatric/Binocular Vision Resident
Atwater Eye Care Center
IU School of Optometry

Case

An 11-year-old Caucasian male presented for a routine eye exam without visual complaints. He denied the presence of headaches, double vision or blur. Previous ocular health was unremarkable. His BMI was 41.9. He was taking albuterol and fluticasone for asthma and had no known drug allergies.

Unaided entrance visual acuities were 20/20 OD, OS at both distance and near. Pupils were equal, round and reactive to light without afferent pupillary defect. Extraocular muscle movements and confrontation visual fields were unremarkable. Subjective refraction was +0.25 OD, +1.00-1.00x165 OS. Intraocular pressure with Goldmann tonometry was 13 mmHg right eye and 15 mmHg left eye. Blood pressure was 136/80 mmHg on the right arm with the patient seated. Anterior ocular health of both eyes was unremarkable with slit lamp examination. Upon dilated fundus examination, optic nerve edema was evident in both eyes without disc or retinal hemorrhages. Macula and retina were within normal limits in both eyes. Spectralis optical coherence tomography (OCT) was performed (Figure 1) and revealed the extent of disc swelling. Humphrey visual field was performed in both eyes. Due to poor reliability with repeat Humphrey visual field testing, Goldmann perimetry was performed which revealed an enlarged blind spot in both eyes.

The patient was referred to a neurologist and an MRI was performed to rule out a space occupying lesion. MRI was normal; while lumbar puncture was elevated above 400 mmH2O with normal composition. The patient was diagnosed with idiopathic intracranial hypertension (IIH). The diagnosis was based on elevated intracranial pressure (ICP) with normal composition without MRI abnormality or evidence of abnormal cerebral perfusion. The patient was started on 75 mg of topiramate daily after adverse side effects were experienced with an initial trial of oral acetazolamide. The patient is monitored every three months by our clinic with dilated fundus exami-
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nation, optical coherence tomography and visual field analysis. After each optometry visit, the patient is seen by neurology. The neurologist uses OCT data (residual papilledema) to assist their dosage adjustment of topiramate. Over the first five months of presentation, topiramate was increased slowly to 600 mg daily and papilledema resolved slowly (see Figure 2). The patient continues to take 600 mg daily 18 months after initial diagnosis due to persistent papilledema which very closely represents the amount of edema present in Figure 2. The patient has been successfully taking topiramate and has not experienced any adverse effects from the medication. Since visual function has remained normal, additional treatment has not been required ie. additional lumbar puncture to relieve pressure. Initially, the patient was also referred to a nutritional expert to begin a strict diet and exercise plan. Unfortunately, 18 months later no improvements have been made in the patient’s weight due to non-compliance with his diet and exercise plan. Treatment compliance has been difficult as the patient remains asymptomatic.

![Figure 2: 5 months](image)

Discussion

The etiology of elevated intracranial pressure in patients with intracranial hypertension is not fully understood. Current theories are increased resistance of cerebrospinal fluid (CSF) outflow, decreased absorption or increased secretion of CSF or altered cerebral hemodynamics.\(^1\) IIH is most common in adult females who are overweight and in their child bearing years of life with a rate of 19.3 in 100,000 versus 0.9 in 100,000 in other populations.\(^1,2\)

In the prepubescent population, there is no gender predilection and frequency is equal between patients with and without obesity.\(^2\) Risk factors of IIH in post pubescent children is similar to that of an adult, therefore the rate of childhood obesity may be the reason for increased prevalence in adolescents.\(^3\) Most children will present with the symptom of headaches and many cases present with strabismus typically from sixth nerve palsy which can resolve following treatment.\(^4\) Younger patients and those without obesity tend to respond less to medical treatment. Based on these characteristics, it is likely that the cause of IIH in prepubescent patients is different than that of adults or obese post pubescent children. Most cases are likely secondary to associated diseases such as autoimmune disorders, endocrine disorders, otitis media, neck injury, nephro-
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tic disease or arteriovenous malformation.\(^1,3\) Separate diagnostic criterion may be required for pre and post pubescent children given the different clinical picture.\(^3\) Important conditions to rule out in pediatric patients with bilateral optic disc edema are malignant hypertension, intracranial mass lesions, hydrocephalus and cerebral venous thrombosis.\(^1,5,6\) Therefore, blood pressure and MRI should always be performed. Magnetic resonance angiography (MRA) should be performed if abnormalities in cerebral perfusion are suspected.\(^1\)

Treatment is aimed at preserving the function of the optic nerve and controlling ICP. Acetazolamide is typically used first line as a diuretic to lower ICP.\(^1-6\) It also reduces CSF production and may change the taste of food which can actually assist with needed weight loss in many cases. Common side effects are fatigue, nausea, vomiting and extremity parasthesias.\(^6\) If acetazolamide is unsuccessful or not tolerated, furosemide is typically used second line which also acts as a diuretic.\(^1-6\) In addition, it alters sodium transport into the brain further reducing ICP. If visual function is normal or if additional headache relief is desired, topiramate may be used. It reduces ICP as it has carbonic anhydrase inhibitor activity.\(^6\) A common side effect of this medication is weight loss which can be advantageous in many cases of IIH.\(^1-6\) Finally, corticosteroids can also be used if the patient has visual symptoms that are not responsive to other medications.\(^2\)

Typically patients are monitored by both optometry and neurology every 1-3 months while papilledema persists. As an optometrist it is important to routinely perform dilated fundus examinations, OCT and visual field analysis to illustrate the level of papilledema and secondary optic atrophy present which can help guide the neurologist’s decision to adjust medication.

References

Asymptomatic Peripapillary CNVM

Shawn Miller, O.D.
Cornea and Contact Lens Resident
Atwater Eye Care Center
IU School of Optometry

Case

A 76 year old Caucasian male presented with a chief complaint of mild blur at distance and near in both eyes. The blur had been present for several months and had a gradual onset. The patient’s systemic history was positive for narcolepsy, an inguinal hernia, benign prostatic hypertrophy, coronary artery disease, diabetes mellitus type 2, hyperlipidemia, hypercholesterolemia, gastroesophageal reflux disorder, polyarthralgia, partial hearing loss, and obesity. The patient was actively taking Aliskiren, Amlodipine, Atorvastatin, Carvedilol, Metformin, Modafinil, and Omeprazole with no allergies to medication. The patient had multiple surgical procedures performed previously, including an appendectomy, transurethral resection of the prostate, aortocoronary bypass, percutaneous transluminal coronary angioplasty, and stomach surgery for a perforated ulcer. The patient’s ocular history was positive for cataract extraction in both eyes with PCIOLs.

Best corrected visual acuities (BCVA) were 20/25+1 OD and 20/20 OS. Extraocular muscle testing was normal and confrontation fields were full to finger counting in both eyes. Pupils were reactive without afferent defect. Anterior segment evaluation was unremarkable and the anterior chamber was deep and quiet in both eyes. PCIOLs were clear and centered in both eyes. Dilated fundus exam of the posterior segment OS revealed a 1 DD choroidal nevus that was flat with no drusen or overlying pigment. OD posterior segment evaluation revealed a suspect peripapillary choroidal neovascular membrane (CNVM) superotemporal to the optic nerve head with subretinal fluid present. Additionally, fibrotic scarring and possible subretinal fluid were present superonasal to the optic nerve head. Blot-like subretinal hemorrhages were present intermittently around the optic nerve head (Figure 1). The peripheral retina and macular findings were unremarkable OU. Fundus photos were taken and ocular coherence tomography (OCT) was

Figure 1: Fundus photo OD

Figure 2: ONH OCT OD and OS
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performed to document and confirm subretinal fluid OD (Figure 2).

The patient was diagnosed with an asymptomatic peripapillary CNVM OD. There were no signs of diabetic retinopathy. The patient was referred to a retinal specialist the day of presentation, but was not seen until one month later. The retinal specialist performed fluorescein angiography and a B-scan ultrasound and confirmed our tentative diagnosis. An intravitreal Avastin injection was performed that day, as well as at the subsequent follow-up. The retinal specialist could find no other apparent cause of the peripapillary CNVM and thus called it idiopathic. The patient seemed to be responding well to his treatment, but unfortunately did not return for his third Avastin injection.

Discussion

A CNVM occurs when there is pathological growth of new blood vessels between Bruch’s membrane and the sensory retina. By definition, a CNVM is considered peripapillary when it occurs within one disc diameter of the optic nerve head. It is estimated that approximately 10% of posterior pole CNVMs are peripapillary.

Peripapillary CNVMs can result in patients being asymptomatic or having a range of symptoms. Visual acuity can range from normal to severely reduce. Patients may experience floaters or distortion, and scotomas may appear on visual field testing. The degree of visual changes largely depends on the location and severity of the peripapillary CNVM. Temporal location tends to have a less favorable outcome due to proximity to the macula. This increases the risk of the CNVM itself progressing to the macula or a subretinal or pigment epithelium detachment affecting the macula and impacting central vision.

The clinical course of peripapillary CNVMs can vary greatly. The neovascular net can remain stable or involute. Additionally, peripapillary CNVMs have been known to recur. Peripapillary CNVMs can be associated with numerous ocular conditions. As would be expected, conditions leading to stretched or thinned retinal tissue can lead to breaks in Bruch’s membrane and therefore a peripapillary CNVM. Conditions which lead to hypoxic or sick retinal tissue, chronic inflammation, or compromised optic nerve head health can also lead to formation of a peripapillary CNVM. Age-related macular degeneration, pathological myopia, angiod streaks, chronic uveitis, sarcoidosis, presumed ocular histoplasmosis, optic disc drusen, and an optic disc coloboma are just a few of many conditions associated with peripapillary CNVM development. Ignoring age, approximately 42-45% of peripapillary CNVMs are associated with age-related macular degeneration and 28-39% are idiopathic.

Literature indicates it is helpful to think of peripapillary CNVMs in terms of the age of the patient. We can loosely divide the patient population into two general groups consisting of older patients, age 40-50 and above, and younger patients, age 40-50 and below. Peripapillary CNVMs in older patients tend to be associated with age-related macular degeneration or are idiopathic. Peripapillary CNVMs in younger patients, however, can be associated with numerous ocular conditions with no consistent pattern of association. The clinical course of peripapillary CNVMs in the older group of patients tends to include progression of the CNVM, whereas younger patients tend to have less CNVM progression but more serious or hemorrhagic sensory retinal detachment.
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A CNVM can appear with retinal serous elevation, intraretinal and subretinal hemorrhaging, exudates, pigmentation, and fibrotic scarring. Differential diagnostic testing to confirm a peripapillary CNVM and further investigate suspected associated pathology includes ocular coherence tomography (OCT), fluorescein angiography, and B-scan ultrasound. The most prominent differential diagnosis for a peripapillary CNVM specifically is Polypoidal Choroidal Vasculopathy (PCV). PCV affects choroidal vasculature leading to distinctive polyps in the vasculature that is best seen (and therefore differentiated from a peripapillary CNVM) with indocyanine green angiography.

Treatment for peripapillary CNVMs include monitoring, laser photocoagulation, photodynamic therapy, anti-VEGF injections, steroid injections, and surgery with vitrectomy and resection. Laser photocoagulation has been shown to result in a modest increase in visual acuity relative to non-treated eyes. However, one concern with laser photocoagulation is the risk of thermal damage to the nerve fiber layer bundles, primarily the papillomacular bundle. Additionally, laser photocoagulation becomes higher risk when the peripapillary CNVM is located temporal to the optic nerve head due to proximity to the macula. Photodynamic therapy (PDT) is not FDA approved to be used within 200 microns of the optic nerve head. However, used off label, PDT has been shown to have effective improvement of visual acuity relative to non-treated eyes. Additionally, PDT does not have the risk of thermal damage as with laser photocoagulation. Anti-VEGF intravitreal injections have a high success rate improving visual acuity, but it is often necessary for the medication to be injected as a series of injections which can be expensive and inconvenient. Steroid injections can be associated with the development of cataracts and increased intraocular pressure. Surgical options including vitrectomy with resection is considered the most complicated of the treatment methods.

References