

Article ▶ An Amblyope That Wasn't: A Case Report of Prolactinoma Discovered During a Binocular Vision Evaluation

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ABSTRACT

Background: For one to be diagnosed with amblyopia, there must be a valid reason for the diagnosis. A decrease in best-corrected visual acuity alone is not sufficient, and if there are no ocular abnormalities found, further testing should be pursued to rule out other causes, possibly systemic.

Prolactinoma, a benign tumor of the pituitary gland, causes prolactinemia, an elevation in the hormone prolactin. Patients with this condition may present with symptoms of headaches, nausea, vision loss, impotence, or loss of sexual desire, although patients (men in particular) may have none of these indicators. If the tumor begins to invade the optic nerve, visual field loss and acuity loss are likely. These findings may be misdiagnosed or not diagnosed at all if complete testing is not performed. The majority of patients can be treated and managed with Bromocriptine or Cabergoline oral medication, both of which are dopamine agonists. They decrease the amount of prolactin, in turn decreasing the size of the tumor and symptoms.

Case Report: A 26-year-old male, referred from his primary care optometrist, presented for a visual efficiency evaluation. The patient was originally diagnosed with functional amblyopia secondary to a questionable microstrabismus. After a complete binocular vision and ocular health work-up, he was referred for an MRI, which revealed a macroprolactinoma impinging on the optic nerves.

Conclusion: This is a case report which validates the importance of finding a cause for decreased vision, in particular when exam elements do not all point to a definitive cause for diagnosed functional amblyopia.

Keywords: amblyopia, binocular vision, prolactinoma

Case Report

A 26-year-old male first reported to the clinic for a routine eye exam with a complaint that his habitual glasses “aren't quite right.” His glasses were less than a year old, but the patient stated that distance and near acuities were blurry (squinting helped). No other ocular complaints were noted. He had a full physical a few months prior, and his cholesterol and blood pressure were slightly elevated, but no medication was prescribed at that time. The patient denied any ocular history of surgeries, reduced vision, or pathologies.

The patient's presenting acuities were 20/25+ OD, 20/40 OS with no improvement on pinhole. Habitual Rx was -1.75-0.50x076 OD, -1.00-1.00x103 OS. He was best corrected to 20/20- OD and 20/40 OS with -1.75-0.25x066 OD and -1.75-0.75x087 OS. Cover test revealed ortho alignment distance and near; stereo acuity was intact with 20” Randot positive random dot stereopsis. Color vision was normal with Ishihara plates OD and OS. Pupils were normal, equal, round and reactive to light, and no APD was present. Extraocular muscles were full with no pain or diplopia. Intraocular pressures were 20mmHg OD and 12mmHg OS. At this time, the four base out test was performed, which confirmed a central zone of suppression

in the left eye. In addition, an FDT was performed as standard exam testing, which yielded multiple defects in the left eye but high fixation losses. The patient was deemed a poor test taker. Anterior and posterior segment were unremarkable for any pathology, with C/D ratios of 0.25/0.25 OD and 0.2/0.2 OS. The patient was given a new glasses prescription and was released to be seen on a yearly basis, with diagnoses of compound myopic astigmatism OU and microstrabismus and amblyopia, OS.

The patient returned four months later with a complaint of “oily film over eyes” which began a few months prior but had since gotten worse, in particular in the mornings. He denied any infections, fevers, or colds. He also reported blurry vision, but he had not filled the most recent glasses prescription. No other ocular complaints were noted, and there were no changes in medications or illnesses. His presenting acuities at this visit were 20/20- OD, 20/70+ OS, PH 20/60 OS. He refracted to 20/20 OD and 20/60 OS with minimal changes in prescription. Anterior segment evaluation showed Meibomian gland dysfunction. No other testing was performed at this time. He was diagnosed with amblyopia and microstrabismus OS and myopia and astigmatism OU.

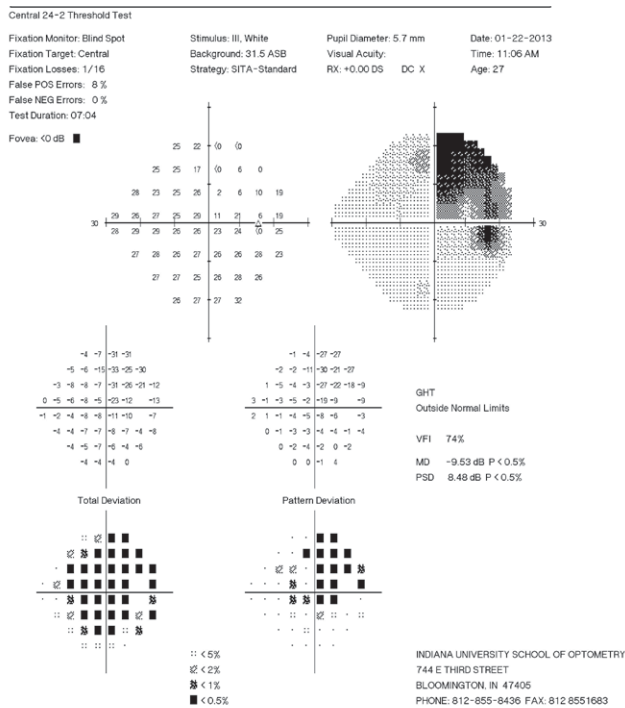


Figure 1. Right eye visual field

In January 2013, he presented for an annual exam with a chief complaint of distance blur through his habitual pair of glasses, left eye. He noted blurred vision for 3-4 years and no significant improvement with new prescriptions. He also noted that the nasal field of vision in the left eye was decreased and almost “lost.” He had had recent surgery on his knee, but no other changes in medication or illnesses were noted, aside from now taking Zoloft.

At this exam, acuities were 20/20-2 OD and 20/60-OS (PH 20/40) at distance, 20/20 OD and 20/50- OS at near wearing -1.75-0.50x070 OD, -1.50-0.75x106 OS. Subjective refraction was -2.00-0.50x075 OD (20/15) and -1.50-0.75x095 OS (20/60); no improvement in acuity OS. Extraocular muscles were full range with no pain or diplopia, and pupils were equal and reactive to light with no APD. Intraocular pressures were 16mmHg OU. The four base out was performed and was positive, confirming a microstrabismus OS. Red cap test was equal in comparison of saturation. The FDT showed multiple defects with high fixation losses, OS more than OD. Anterior and posterior segment were unremarkable for pathology, with C/D ratios of 0.25/0.25 OU. A macular OCT was run to rule out potential causes for decreased vision. Both maculae were flat with no defects. In addition, a 24-2 Humphrey visual field was attempted without success. A plan ensued to refer the patient to the binocular vision department for a visual efficiency evaluation due to the microstrabismus and amblyopia findings in his left eye. It was also assessed that he had an unspecified visual field defect, left eye worse than right eye. The patient was instructed to return to repeat the full 24-2 field in a few weeks.

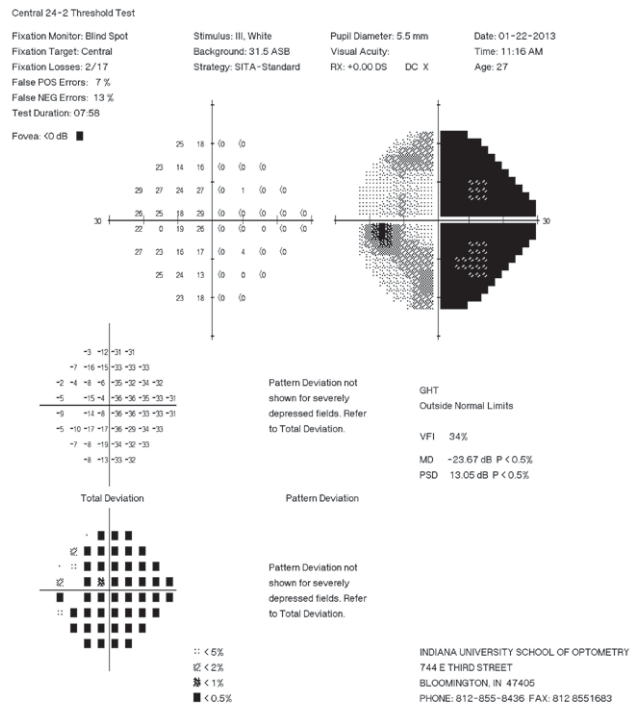


Figure 2. Left eye visual field

One week later, the patient presented for a visual efficiency evaluation with the same complaint of blur OS with glasses (all previous spectacle prescriptions) and subjectively, the vision seemed to be getting worse. He reported that he was diagnosed with amblyopia and microstrabismus in the left eye. No changes in medications or illnesses were noted since his previous exam one week prior.

Acuities at this visit were reduced: 20/30 OD and 20/70 OS, PH 20/60, with correction. He was refracted to yield a very similar prescription to his habitual with minimal improvement in acuity to 20/20- OD and 20/60-OS. Cover test was orthophoria at distance, 6 exophoria at near. Near point of convergence was left eye out at 7cm, and with stamina OS out at 10cm. Prism bar vergences measured: at distance, BI x/6/4, BO x/35/25; at near, BI x/10/8, suppression OS at 5BO. Although exophoria was seen on cover test, the four base out test was done to confirm a microstrabismus as diagnosed in previous exams. It was performed three times; two of three attempts yielded a positive result, but poor fixation was noted. Worth 4-dot testing showed inconsistent results; no definite diplopia or suppression responses were given. Stereo acuity was normal with 20” Randot and positive random dot stereopsis. Eccentric fixation testing was unreliable with questionable nasal fixation. In the amblyoscope, using first-degree, large targets, the objective angle was 2 PD BO, and the subjective angle was 3 BO. There was no movement on unilateral cover test, and it was noted that he had poor inconsistent fixation during this test as well. Color vision was normal aside from missing one plate OS. During red cap test, the patient reported that the cap appeared “ashen and

washed out.” Extraocular motilities were full with no pain or diplopia. Pupil testing revealed a trace to 1+ APD OS. Confrontation fields showed a superior temporal loss OD, complete nasal field loss OS. A formal Humphrey 24-2 field confirmed reliable, consistent field loss, a superior temporal defect in the right eye, and a complete nasal defect in the left eye, (Figures 1 and 2). Healthy optic nerves were viewed with undilated 90D; no pallor was noted, but an OCT was done. Measurements were within normal limits: no atrophy, swelling, or pathologies were found.

It was at this time that extensive patient education was warranted and provided, as the cause of his amblyopia diagnosis did not correlate with the previous microstrabismus diagnosis. The possible causes of visual field loss, afferent pupillary defects, and loss of vision, as well as the need for an MRI, were all explained to the patient. It was deemed that microstrabismus and amblyopia were unlikely causes for his consistent declining vision in the left eye.

Diagnosis

The patient was scheduled for an MRI at the end of the week to scan the brain and orbits with and without contrast. It was discovered that the patient had a pituitary macroadenoma with severe cisternal optic nerve and chiasmal compression and left cavernous sinus involvement (Figure 3). After receiving the report, the patient was scheduled to discuss the results and was promptly referred to a neurosurgeon for consultation that day.

Treatment and Follow-up

The neurologist confirmed the diagnosis and noted the compressive nature of the lesion and also that hemorrhaging was likely in the past. He believed that this might have been associated with a time when the patient had severe migraine headaches for a week as a teenager. The patient’s options for treatment were presented to him: surgery, proton beam therapy, gamma knife therapy, or simply observation. He also had blood work done to ensure that no problems with hypothyroidism occurred. He was then scheduled for a two-month follow-up and scan.

The neurosurgeon made an endocrinologist referral. He diagnosed prolactinoma, the benign pituitary gland tumor, causing highly elevated levels of prolactin. In fact, the blood work showed prolactin levels of 2600 ng/mL; normal levels are less than 20 ng/mL.³ The patient was prescribed Cabergoline, a dopamine agonist, to decrease the levels of prolactin in the blood stream.

The patient was seen at the eye clinic for a visual field after a few months of medical therapy. Although his visual field remained stable, his visual acuity slightly improved, with pinhole in the left eye to 20/40-, the right eye remaining 20/20. Subjectively, he felt as if his vision was improving. Systemically, he reported feeling no symptoms and was still continuing with his diet and exercise regime. At his most recent exam, the patient’s visual acuities were 20/20 OD and 20/30 OS, best

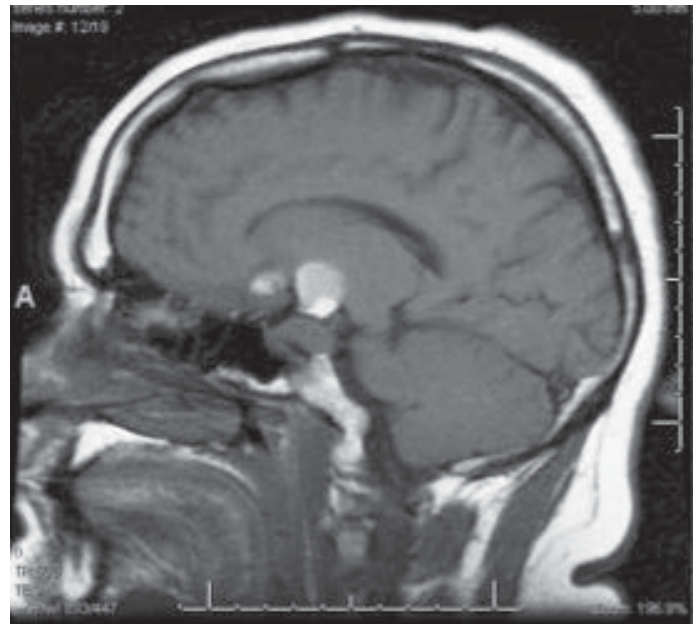


Figure 3. Patient's Magnetic Resonance Image

corrected. He expressed at this exam that his vision “has never been better.” With medication, the tumor shrank to the point that the endocrinologist thought that the patient had had it surgically removed. He is currently still on medication and is being followed for the prolactinoma.

Discussion

Prolactinoma, a benign tumor on the pituitary gland that secretes the hormone prolactin, accounts for approximately 40% of all pituitary tumors¹ and are the most common type of benign pituitary tumor.^{2,3} They are most often found in patients under the age of 40.² Microprolactinomas (<10mm in size) are five times more common in women² but are found in 10 per 100,000 (0.01%) of men.¹ In the male population, the tumors are typically macroprolactinomas, which are greater than 10mm in size.⁴ Prolactinomas often grow greater than 5mm in men before any symptoms are noticed.² As the tumor grows, compression may result, and symptoms of headache, nausea, vomiting, nasal drainage, vision changes, diplopia, and visual field loss may occur. In a retrospective study of 46 male patients with prolactinomas, 74% of those with macroadenomas presented with symptoms of headaches or history of headaches, and many also had vision loss.⁵ If one presents with any of these symptoms, an MRI and blood work are completed in order to determine the presence, location, and type of lesion. It is notable, however, particularly in men, that it is not uncommon to be asymptomatic.²

In most cases, treatment of the prolactinoma is medication, but surgery may be warranted if vision is impacted. Bromocriptine or Cabergoline, both dopamine agonists, are used to reduce the amount of prolactin in the blood to normal levels. Few side effects are seen with these medications, but these may include orthostatic hypotension and nausea. When used in standard dosing, long-term adverse effects have

not been reported.^{6,7} The goals of treatment are to decrease symptoms and normalize pituitary function, providing permanent control of hormones.⁵ Length of medical treatment is dependent on prolactin levels, usually 1-3 years for most patients.^{6,8} It is possible that if therapy is discontinued, prolactinemia will recur, and the tumor size will likely increase; therefore, medication may be used life-long, as the medications are not tumorcidal.^{4,7,8} If the tumor is large, it is harder to treat, and therefore may require radiation therapy. Approximately 70-80% of patients with microprolactinoma are successfully treated with medications alone and may achieve normoprolactinemic remission following medication withdrawal.^{4,9}

In regards to this particular case, the patient was diagnosed with functional amblyopia, defined simply as a decrease in visual acuity. However, there must be a causative agent in order to deem someone amblyopic. Also, functional amblyopia does not result in a progressive decrease in visual acuity in adult patients. Risk factors for amblyopia include strabismus, refractive error, or an anatomical obstruction such as a cataract or ptosis.¹⁰ If none of these factors are present, further testing for pathological causes is imperative.

Amblyopia develops in infancy or early childhood, and acuities can range from 20/30 to 20/200 or worse. If any of the risk factors are present during the critical period of visual development, the visual cortex is not properly stimulated, and therefore vision cannot mature properly.¹¹ The patient in our case was suspect to have microstrabismus, the amblyogenic risk factor. The four base out test, a common test used to confirm microstrabismus, was not actually positive. The image was shifted into the patient's left nasal field, the area of defect, leading to a false positive test and diagnosis.

Due to field and vision loss associated with pituitary tumors, a patient may be misdiagnosed with another ocular entity or deemed amblyopic, when in fact they harbor no amblyogenic risk factors. It is vital to examine all aspects of the ocular exam and determine the cause of amblyopia before diagnosis.

In this case, the patient had reduced visual acuity in one eye with the assumption of a microtropia as the contributing factor and was diagnosed amblyopic. After multiple exams and a progressing decrease in best-corrected acuities over time, it was ultimately determined that the acuity loss was due to pathology. Loss of field found on confrontations and confirmed with a formal visual field led to an MRI, which revealed a tumor impinging on the optic tract.

Conclusion

This is a case that demonstrates how vital it is to define the contributing factor when diagnosing a patient with amblyopia.

Vision loss does not always mean amblyopia, as there must be a cause for any decrease or loss of vision. Also, visual field loss is not consistent with amblyopia. One must perform formal visual fields and, pending results, refer as warranted. The patient in this case exhibited none of the amblyogenic risks, nor did he have any history of ocular abnormalities. This case also portrays the importance of how primary care optometry and binocular vision specialty clinics can assist in treatment for a multitude of diagnoses, providing complete and cohesive optometric care.

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