

CE Credit - Topic Review

Optic Nerve Sheath Meningioma: A Case Report

Jill Gottehrer, OD, FAAO

CRO (Clinical & Refractive Optometry) Journal

ABSTRACT:

Introduction: The clinical manifestations of optic nerve sheath meningiomas may mimic other orbital or neurological conditions, particularly in asymptomatic patients. This may result in misdiagnosis or delayed treatment, resulting in permanent vision loss. This case report highlights multimodal imaging findings and the clinical outcome of a patient with an optic nerve sheath meningioma.

Case Report: A 71-year-old Caucasian male presented for a routine eye exam. Ocular examination revealed near-normal visual acuity for both eyes. Fundus examination showed indistinct cupping and indistinct disc margins in the right eye. Optical coherence tomography was performed verifying an edematous right optic nerve. Fundus autofluorescence did not reveal any optic disc drusen in either eye. Color vision testing was reduced in the right eye compared to the left. Additionally, formal automated visual field testing showed an inferior nasal step defect in the right eye and a clean visual field without defects in the left eye. Laboratory testing placed giant cell arteritis lower on the differential list. Computerized tomography of the head and orbits without contrast indicated the presence of a mass in the right optic nerve. Magnetic resonance imaging with contrast of the head and orbit was performed confirming the diagnosis of an optic nerve sheath meningioma.

Conclusion: Optic nerve sheath meningiomas typically cause vision loss and/or diplopia; however, not all patients are symptomatic. Multimodal imaging can aid in the diagnosis of optic nerve sheath meningiomas, allowing for faster diagnosis, treatment, and possibly improved prognosis.

INTRODUCTION

Optic nerve sheath meningiomas (ONSM) account for approximately 2% of all orbital tumors and 1% of all meningiomas.¹ These tumors predominantly occur in middle-aged females. ONSM may also present in children and be more aggressive.² Pediatric ONSM are rare, with an overall prevalence estimated between 1:95,000 and 1:525,000.³ Pediatric ONSM have a higher affiliation with neurofibromatosis type 2. Approximately 95% of ONSM cases are unilateral, however bilateral tumors, which account for 5-6% of cases, tend to occur in those with neurofibromatosis type 2.¹ ONSM typically presents with slow, painless, progressive vision loss in the affected eye. The primary goal of treatment of this condition is to avoid secondary complications and sequelae, such as complete vision loss. This case report highlights multimodal imaging findings and the clinical outcome of a patient diagnosed with an optic nerve sheath meningioma.

CASE REPORT

A 71-year-old new patient presented to the eye clinic to update his eyeglass prescription. He reported his last eye exam was three years prior and was informed of cataracts and early age-related macular degeneration. His medical history was positive for hypertension, hyperlipidemia, hyperparathyroidism, and osteopenia. For these conditions he took amlodipine and lisinopril. The patient reported an allergy to penicillin. He denied any history of injury or surgery to his eyes and no significant family ocular history.

His entering visual acuity with correction was 20/30-² in his right eye and 20/30- in his left eye without any improvement by pinhole. With an updated refraction, his best corrected visual acuity improved to 20/25- in his right eye and 20/30+ in his left eye. Pupils were equal, round and reactive to light without an afferent pupillary defect. On testing eye movements, there was no evidence of ophthalmoplegia.

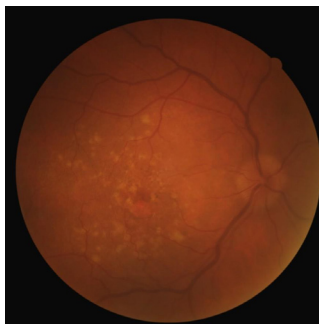


Figure 1. Fundus photo of the right eye showing confluent drusen in the macular area and the optic nerve with negligent cupping and indistinct disc margins



Figure 2. Fundus Photo of the left eye showing confluent drusen in the macular area with the optic nerve margins distinct without any edema or atrophy

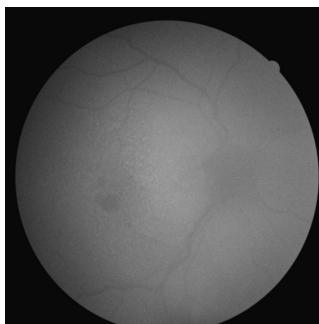


Figure 3. Fundus autofluorescence of the right eye indicating no presence of buried disc drusen and central geographic atrophy

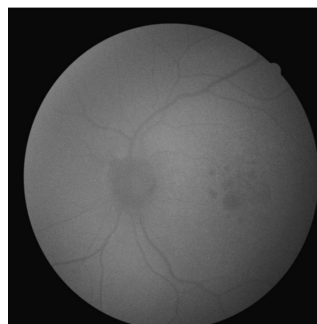


Figure 4. Fundus autofluorescence of the left eye indicating no presence of buried disc drusen and central geographic atrophy

His anterior segment was unremarkable both eyes. His intraocular pressure by Goldmann Applanation Tonometry was 17mmHg in the right and left eye at 2:42 PM. He was dilated with 1% Tropicamide in both eyes at 2:43 PM. On his posterior segment exam, 1+ nuclear sclerotic cataracts and vitreous syneresis were noted in both eyes. The optic nerve cupping was negligent, and the optic disc margins were indistinct in the right eye. However, there was no frank edema, pallor, Paton's lines, or hemorrhages appreciated in the right optic nerve. Comparatively, the left optic nerve had a cup-to-disc ratio of 0.20 round with distinct disc margins, and the rim tissue appeared pink and perfused. The macula in each eye was noted to have confluent drusen with retinal pigment epithelium hyperplasia centrally, without any hemorrhaging or macular neovascularization present. The arterioles and venules in both eyes were noted to be of normal caliber and there were no retinal holes/tears/detachments 360 in the periphery of both eyes.

Same day color photos (Figures 1 and 2) and fundus autofluorescence were obtained (Figures 3 and 4), indicating no buried optic nerve drusen in either eye.

Optical coherence tomography (OCT) macula was obtained (Figure 5), showing the photoreceptor layer intact both eyes with the presence of drusen and no macular neovascularization nor inner retinal or subretinal fluid.

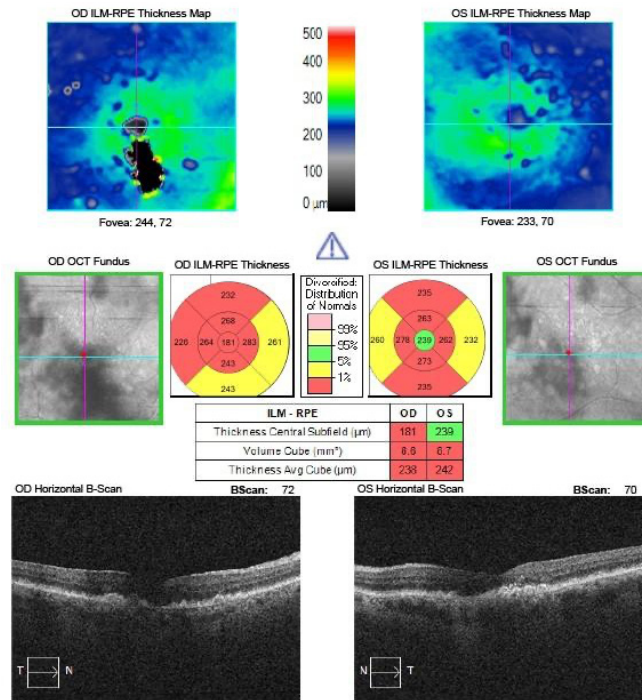


Figure 5. OCT macula showing the photoreceptor layer intact both eyes with the presence of drusen and no macular neovascularization nor inner retinal or subretinal fluid in both eyes

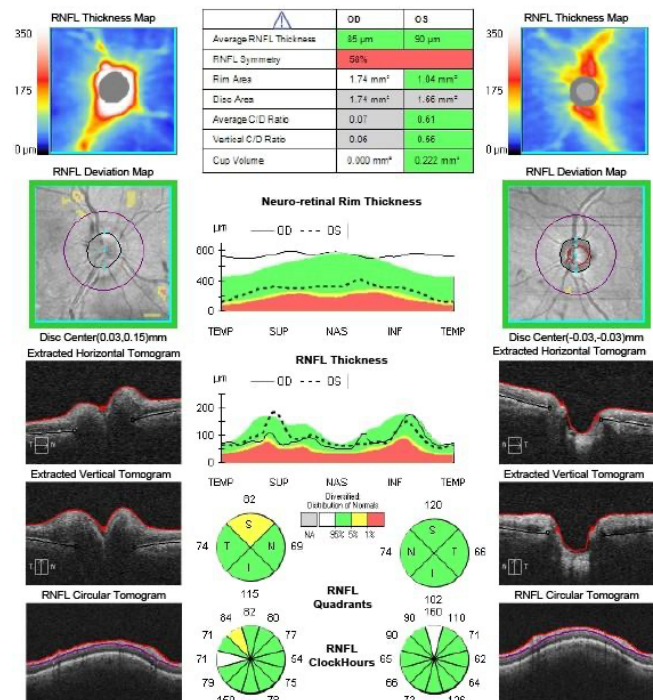


Figure 6. OCT RNFL showing the left eye being within normal limits 360° but the right eye showed an edematous appearance with borderline thinning superiorly

On OCT retinal nerve fiber layer (RNFL) (Figure 6), the left eye was within normal limits 360° but the right eye showed an edematous appearance with borderline thinning superiorly, potentially indicative of progression toward atrophy.

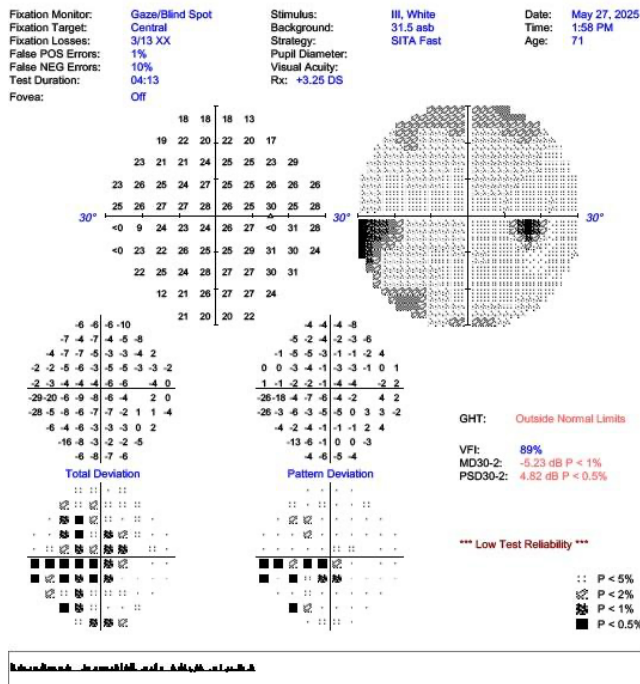


Figure 7. Humphrey Visual Field 30-2 of the right eye showing an inferior nasal defect

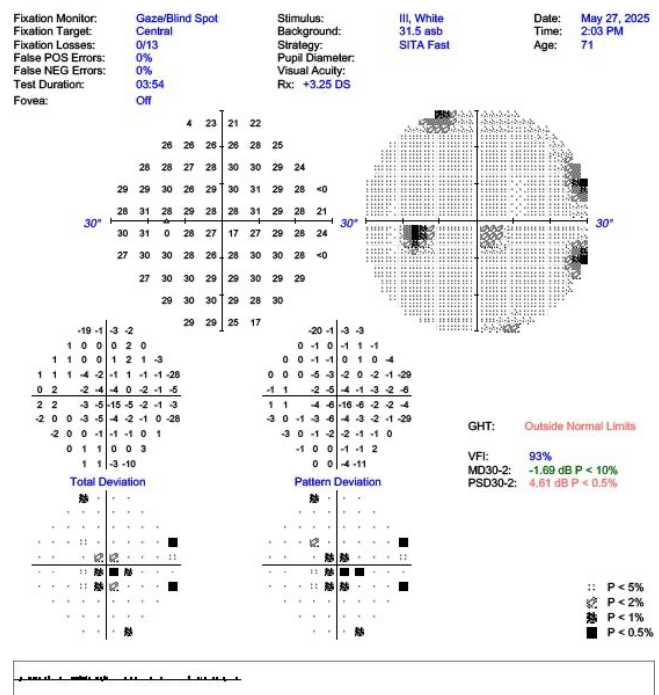


Figure 8. Humphrey Visual Field 30-2 of the left eye showing paracentral defects

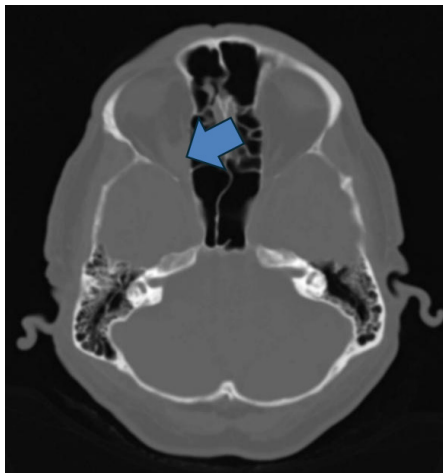


Figure 9. Non-contrast head CT (axial view) showing a right orbital mass (blue arrow)

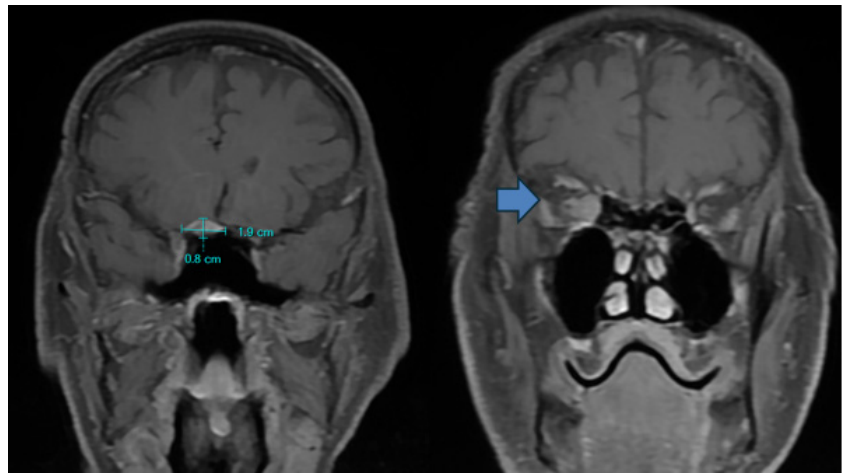


Figure 10. Non-contrast head CT (coronal view) showing a right-sided "doughnut" sign (blue arrow)

Same-day color vision was performed using Ishihara. The patient correctly identified 5/10 plates in the right eye and 9/10 plates in the left eye. Humphrey Visual Field 30-2 SITA Fast was subsequently ordered (Figure 7 and 8). His right eye was reliable with a mild inferior nasal step noted. His left eye was also reliable with paracentral defects noted which can be attributed to his age-related macular degeneration.

A potential differential diagnosis of giant cell arteritis was covered by ordering complete cell count, erythrocyte sedimentation rate, and C-reactive protein, which yielded non-significant results, lowering the concern for

temporal arteritis. A same day non-contrast head and orbit computerized tomography (CT) was ordered. His head CT revealed a soft tissue process in the superior medial aspect of the right orbit, suspicious for a mass with uncertain dimensions. (Figures 9 and 10). His CT orbit revealed a soft tissue mass or process within the posterior aspect of the right orbit.

A brain magnetic resonance imaging (MRI) with contrast was further recommended per radiology. His MRI confirmed a mass involving the right orbital apex, abutting the medial rectus and optic nerve (Figure 11).

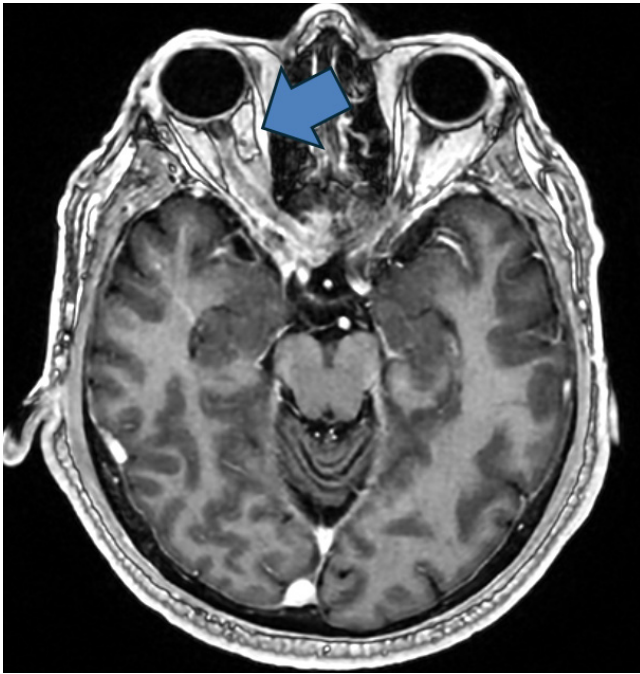


Figure 11. Brain MRI with contrast (axial view) showing a “tram-track” pattern involving the orbital segment of the right optic nerve (blue arrow)



Figure 12. Orbital MRI with contrast (coronal view) exhibiting a mass surrounding the right optic nerve (blue arrow)

There was no definitive intracranial extension. A second mass was noted in the right planum sphenoidale with intense enhancement, highly suggestive of a meningioma. Per radiology, the lesions appeared separate. On his MRI of the orbits, an irregular bilobed intraconal mass isotense to the brain on T1 and T2 extending through the optic canal to the planum sphenoidale was noted. The intracranial component measured 1.6 x 1.4 x 0.5 cm without extension into the optic chiasm and the intraorbital portion measured 1.7 x 1.4 x 1.1 cm. Following contrast enhancement, the mass demonstrated strong enhancement, surrounding the optic nerve (Figure 12).

The radiology report stated the etiology of the mass favored meningioma with other differentials including optic nerve glioma, lymphoma, or schwannoma, however, these differential diagnoses were less likely.

Due to these findings, a consultation for neurosurgery was placed for further treatment and management. Neurosurgery confirmed the diagnosis of an optic nerve sheath meningioma and recommended observation over treatment due to the patient's excellent visual acuity and lack of symptoms such as diplopia. A neuro-ophthalmology consult along with a repeat brain MRI, and orbits was recommended in six months.

DISCUSSION

While optic nerve tumors are rare, ONSM comprise approximately one-third all optic nerve neoplasms.¹ ONSM are most commonly located in the intraorbital or intracanalicular segments of the optic nerve sheath. These

tumors arise from the meningeal cells (arachnoid cap cells) lining the optic nerve sheath. These tumors generally expand circumferentially around the optic nerve, which can result in compression of its pial vascular supply. ONSM typically invades the entire optic nerve, from the canal posteriorly to the globe anteriorly.^{4,5} Optic nerve sheath meningiomas can extend into the chiasm via the optic foramen. Optic atrophy occurs due to compression of the optic nerve. Only a small portion (10%) of orbital meningiomas originate in the orbit itself while the remaining cases originate intracranially and extend into the orbit.¹ Tumors that arise from other intracranial or intraorbital locations and secondarily affect the optic nerve sheath are referred to as “secondary ONSMs”. Optical canal ONSMs represent 8% of ONSMs and can cause compression of the optic nerve due to the tight, anatomical space.

Some patients with ONSM are asymptomatic. Other patients with ONSM present with slow, painless and progressive vision loss that can progress to complete blindness if left untreated. Dutton found 24% of patients had a visual acuity of counting fingers or worse while 45% of patients had a visual acuity of 20/40 or better.⁶ The classic triad in patients with an ONSM is vision loss, optic atrophy, and optociliary shunt vessels, but most patients do not present with all three components.¹ A relative afferent pupillary defect may be observed in the involved eye. Visual field defects may present and are non-specific; they can present as altitudinal defects, generalized constriction, or an enlarged blind spot. Other clinical signs may include proptosis and transient vision loss associated with eye movement, known as gaze-evoked amaurosis. Dyschromatopsia and strabismus may also be present.

In early stages, chronic optic nerve edema may be seen on the clinical exam due to the tumor compressing the intraorbital portion of the optic nerve. On OCT RNFL, increased RNFL thickness can be noted. As the tumor progresses, this will lead to optic atrophy, seen as RNFL thinning on OCT.^{7,8} As optic disc edema subsides, optociliary shunt vessels may develop in about 30% of patients, representing venous collaterals that connect to the retinal or choroidal circulation, forming a bypass for blood to drain. Optociliary shunt vessels may appear in other ocular conditions such as central retinal vein occlusions due to chronic central retinal vein obstruction.

A computed tomographic (CT) of the head and orbit can show the hyperostosis produced in secondary tumors. CT imaging can also show calcification of the tumor and the classic “tram-track” sign, which is parallel thickening and enhancement around the optic nerve. The tumor typically has a smooth lobulated contour.

The diagnosis of ONSM is confirmed with magnetic resonance imaging (MRI), particularly with gadolinium-enhanced fat-suppression sequences. MRI is the gold standard for the diagnosis of ONSM and precludes the need for tissue biopsy. On MRI, ONSM will appear isointense to grey matter on both T1 and T2 weighted sequences, with pronounced contrast relative to the non-enhancing optic nerve. This is due to ONSM being sensitive to gadolinium contrast. On axial imaging, ONSM will present with the characteristic tram-track sign, corresponding to the enhancing outer ONSM encircling the inner non-enhancing optic nerve. On coronal cuts, ONSM may present on imaging as a “doughnut” or “non-enhancing dot” sign. MRI typically demonstrates tubular expansion of the meninges surround the optic nerve (62%), with less common patterns including globular (23%), fusiform (11%), and focal enlargement of the optic nerve (4%).⁹

Differential diagnoses of ONSM include optic nerve glioma, optic neuritis, orbital lymphoma, orbital metastases, idiopathic orbital inflammatory disease (orbital pseudotumor), neurosarcoidosis, and giant cell arteritis.¹⁰ On MRI imaging, an optic nerve glioma will demonstrate uniform enhancement of the optic nerve and will not have the classic tram track sign. Although they may have a similar clinical presentation, ONSM is painless compared to optic neuritis and MRI imaging will further differentiate the two. Both giant cell arteritis and ONSM will show enhancement of the optic nerve on MRI imaging; however, ONSM will display the pathognomonic tram-track sign. Additionally, patients with giant cell arteritis often report profound, painful vision loss in one or both eyes and may have accompanying inflammatory markers in their bloodwork.

Co-management with a neuro-ophthalmologist and/or neurosurgeon is recommended. Observation is recommended for patients whose visual function is intact or remains stable, particularly in patients with a central visual acuity of 20/50 or better. Serial visual fields and retinal nerve fiber layer OCT is recommended in addition to repeat MRI every year.¹¹

Tumor resection is challenging without risking severe vision loss due to the intimate relationship between the ONSM and the optic nerve. However, it is recommended in cases of disfiguring proptosis with severe vision loss or in cases of intracranial extension. Observation is recommended if the eye with the ONSM is blind and the tumor is confined to the orbit. However, there are some arguments / is an argument to be made for surgical resection to limit involvement of surrounding tissues. If the eye has a visual acuity of light perception or no light perception and intracranial extension is present, both the tumor and the optic nerve should be removed.⁵

Radiotherapy may be an alternative to surgery.¹² Stereotactic radiotherapy has demonstrated good long-term control. It demonstrated either stable or improved visual field rates of 83.3%-100%. Risks include radiation-induced retinopathy or optic neuropathy.¹³

There is little risk of mortality from ONSM. If the tumor is left untreated, progressive vision loss results from direct compression of the optic nerve and impairment of its vascular supply. In some cases, ONSM may infiltrate the optic nerve.⁵ Most patients eventually develop blindness in the eye affected by the ONSM. For those cases that undergo surgical resection, vision deteriorates in approximately 33%-50% of cases.⁵

CONCLUSION

Optic nerve sheath meningiomas are a rare, benign intracranial tumor. Multimodal imaging is necessary to distinguish it from other orbital or neurological conditions. Accurate diagnosis of optic nerve sheath meningioma is important as delayed or missed diagnosis can cause irreversible vision loss.

REFERENCES:

1. Parker RT, Ovens CA, Fraser CL, Samarawickrama C. Optic nerve sheath meningiomas: prevalence, impact, and management strategies. *Eye Brain*. 2018;10:85-99. doi:10.2147/EB.S144345
2. Narayan D, et al. Cystic change in primary paediatric optic nerve sheath meningioma. *Orbit*. 2016;35(4):236-238. doi:10.1080/01676830.2016.1176212
3. Levin LA, Jakobiec FA. Optic Nerve Tumors of Childhood: A Decision-Analytical Approach to Their Diagnosis. *International Ophthalmology Clinics*. 1992;32(1):223.
4. Jacobi DM. Optic nerve sheath meningioma. *Clinical and Experimental Optometry*. 2019;102(2):188-190. doi:10.1111/cxo.12819
5. Rassi MS, et al. Prognostic factors in the surgical treatment of intracanalicular primary optic nerve sheath meningiomas. Published online September 21, 2018. doi:10.3171/2018.4.JNS173080
6. Dutton JJ. Optic nerve sheath meningiomas. *Survey of Ophthalmology*. 1992;37(3):167-183. doi:10.1016/0039-6257(92)90135-G
7. Gaur D, Pai HV. Optical coherence tomography in the evaluation of disc edema. *Kerala Journal of Ophthalmology*. 2019;31(1):17. doi:10.4103/kjo.kjo_91_18
8. Tonagel F, Wilhelm H, Kelbsch C, Richter P. Characteristics of peripapillary retinal nerve fiber layer atrophy in glaucoma, optic nerve sheath meningioma, and sphenoid wing meningioma. *Graefes Arch Clin Exp Ophthalmol*. 2022;260(2):577-581. doi:10.1007/s00417-021-05354-2
9. Saeed P, Rootman J, Nugent RA, et al. Optic nerve sheath meningiomas. *Ophthalmology*. 2003;110(10):2019-2030. doi:10.1016/S0161-6420(03)00787-5
10. Ing EB, Garrity JA, Cross SA, Ebersold MJ. Sarcoid Masquerading as Optic Nerve Sheath Meningioma. *Mayo Clinic Proceedings*. 1997;72(1):38-43. doi:10.4065/72.1.38
11. Pandit R, Paris L, et al. Long-term efficacy of fractionated conformal radiotherapy for the management of primary optic nerve sheath meningioma. *British Journal of Ophthalmology*. 2019;103(10):1436-1440. doi:10.1136/bjophthalmol-2018-313135
12. Sasano H, et al. Efficacy of intensity-modulated radiation therapy for optic nerve sheath meningioma. *Graefes Arch Clin Exp Ophthalmol*. 2019;257(10):2297-2306. doi:10.1007/s00417-019-04424-w
13. Ratnayake G, Oh T, Mehta R, et al. Long-term treatment outcomes of patients with primary optic nerve sheath meningioma treated with stereotactic radiotherapy. *Journal of Clinical Neuroscience*. 2019;68:162-167. doi:10.1016/j.jcns.2019.04.018