A Case of Extensive Suprasellar Meningioma Presenting with Insidious Visual Impairment

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Abstract

Background:

- Meningiomas are a common tumor of the brain resulting from abnormal cellular proliferation in arachnoid cap cells associated with the dura mater.
- Most individuals present with neurological abnormalities due to brain mass shift or compression.

Objective:

- To demonstrate how a slow-growing, histologically benign intracranial tumor, may produce severe visual impairment via an insidious course.

Results:

- The patient recovered from no light perception to eccentric light perception at 6 months. While limited, this represents a significant measurable change from baseline

Conclusion:

- Meningiomas may present insidiously in patients without known risk factors, which can delay recognition. Continued diagnostic consideration across evolving clinical findings, despite competing comorbidities, is important for managing the patient and optimizing outcomes.

Introduction

- Meningiomas have long been studied due to their high prevalence in the spectrum of neurological tumors.
- Their development is less random and has a propensity to develop in certain populations with unmodifiable risk factors.
- There is a 2:1 increase in women compared to men. However, the most important predictive factor for development of meningiomas is a defect in the NF2 tumor suppressor gene located at 22q12.2 which produces merlin.[2]
- A loss of heterozygosity of NF2 in both sporadic cases and those diagnosed with neurofibromatosis type II is considered a primary mechanism of development in up to 70% of cases.[1,2]
- Following the WHO classification of central nervous system tumors updated in 2021 there are now 15 variants which can be classified as NF2 and non-NF2-mutated.[5]
- These variants have a preference to their growth location based on mutation.
- The NF2 type is predominantly on the convexity of the meninges with more atypical features and non-NF2 is on the skull base with benign features.[1]
- This case features the latter with a left-sided suprasellar mass most likely originating from arachnoid villi cap cells associated with the dura mater.[6]
- A retrospective analysis between 1994 and 2013 found an incidence of 100 cases of meningioma located in a similar region, specifically the tuberculum sellae. Of them, 89% showed asymmetric visual defects.[3]

Methods

- A case report and literature review were both performed. Written consent was obtained from the patient.

Clinical Course

Presentation:

A 33-year-old female presented to the ophthalmology clinic for a routine diabetic fundus exam. She also complained of worsening of her left eye vision, which has historically been her weaker eye. She has been a type II diabetic for 2.5 years. Most recent HbA1c of 13.6. The patient was found to have poor vision of left eye (OS) as a teenager and was prescribed glasses.

- Her poor left eye vision had been attributed to amblyopia, likely refractive.
- Optical coherence tomography (OCT) and fundus exam confirmed no optic nerve edema, pallor or diabetic retinopathy 3 years prior.
- At the time of her exam, now 3 years later, the patient presented with complete no light perception (NLP) vision OS.
- Per patient, vision had been worsening over the past year.
- Patient denied diplopia, photophobia, flashes or floaters, waviness or distortion of both eyes.
- Intraocular pressure 21 and 19 mmHg right eye (OD) and OS, respectively.
- Adnexal/periorbital exam revealed no evidence of proptosis or enophthalmos.
- Anterior exam was normal, there was no evidence of conjunctival or corneal abnormalities.
- No anterior chamber cell, flare, hyphema or hypopyon.
- No iris defect, nodules, neovascularization, masses, heterochromia, lenticular opacities, or synechaia.
- Fundus exam yielded mild temporal pallor OS.
- No retinopathy or maculopathy.
- OCT revealed a significant retinal nerve fiber layer (RNFL) (Figure. 1A/B) and ganglion cell layer (GCL) (Figure. 2) loss compared to imaging 3 years prior.
- The mass appeared to be separate from the pituitary measuring 2.5 x 1.9 x 2.0 cm with a dural tail.
- MRI of the brain and orbits with and without contrast revealed an eccentric left-sided suprasellar meningioma with encasement of the A1 segment of the anterior cerebral arteries as well as likely encasement of the prechiasmatic optic nerve with optic nerve atrophy (Figure 3 A/B).

Figure 1A:

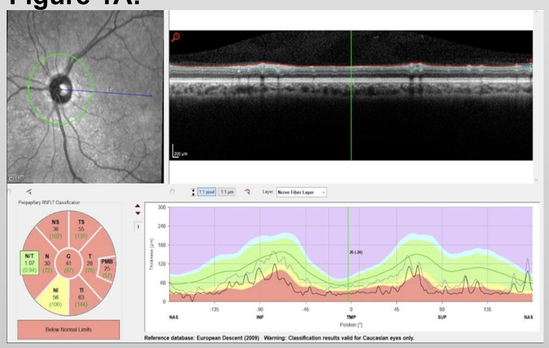

Figure 1B:

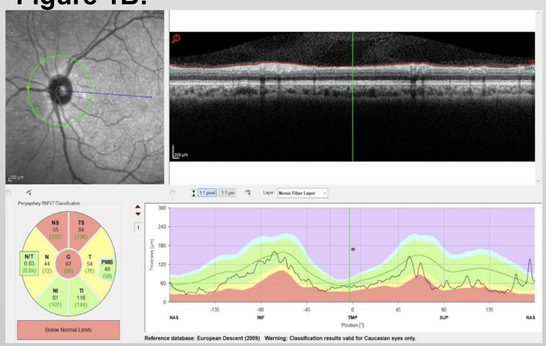

Figure 1:
A: Left eye from 2025. OCT shows decreased thickness of the retinal nerve fiber layer. A pale optic cup is shown in the upper left image denoting damage to the optic nerve.
B: Left eye OCT from 3 years prior showing mild thinning of the nasal and temporal RNFL. Marked thinning of the superior nasal and temporal RNFL.

Figure 2:

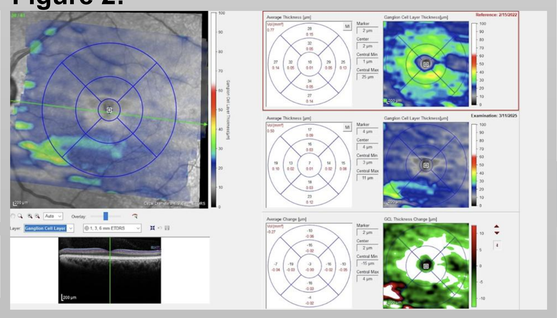

Figure 2: Left eye GCL imaging showing decreased thickness over the course of approximately three years.

Figure 3A:

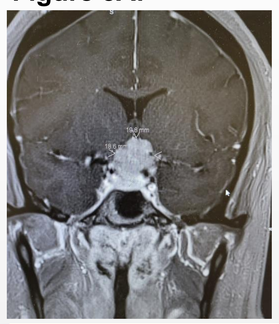

Figure 3B:

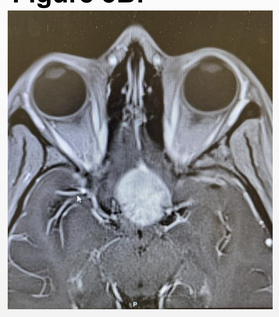

Figure 3
A: Coronal image obtained via MRI showing suprasellar mass.
B: Axial image obtained via MRI showing suprasellar mass slightly off midline to the left with pre-chiasmatic left optic nerve compression.

Results

- Neurosurgery was consulted, given the tumor size and proximity to critical structures, surgical resection/debulking via an orbital zygomatic approach was urgently performed.
- Biopsy of the lesion was confirmed by pathology to be a meningioma WHO grade 1.
- Features included nests of cells with eosinophilic cytoplasm and tumor cells arranged in whorls.
- Immunohistochemical staining was positive for EMA and progesterone receptors.
- Biopsy was negative for mutations in brain tumor genes IDH1, IDH2.
- After resection, the patient's visual acuity improved to seeing shadows.
- The patient was discharged with follow ups.
- 6 months post op, OS vision remained markedly compromised with now eccentric light perception vision, from an initial no light perception.

Discussion

- Given the results of GCL imaging 3 years prior that recorded a vast decrease in GCL thickness, the tumor was most likely present for a long period of time, however, too small to cause substantial symptoms (Figure 2).
- OCT findings showed mild and marked thinning of the RNFL in the nasal/temporal and superior nasal/temporal fields, respectively, 3 years prior (Figure 1B).
- The pathogenesis of the visual impairment was primarily due to compression of the prechiasmatic optic nerve, where the tumor mass directly compressed the fine optic nerve fibers.
- This led to axonal damage, loss of retinal ganglion cells reflected by the extreme RNFL thinning, and ultimately, optic nerve atrophy
- In similar studies of optic nerve involvement with meningiomas, an inverse relationship was found between onset of visual loss and intervention with visual improvements post-operatively as well as thinning of the RNFL as a negative prognostic factor. [4]
- The tumor compressed the A1 segment of the anterior cerebral artery, and even though it does not directly supply the optic nerve, the vessel's compression could potentially affect the microvasculature in the region.
- Specifically, the small pial vessels that contribute to the optic nerve blood supply.
- The primary reason for vision loss was direct mechanical compression of the optic nerve.
- Since meningiomas are often slow growing, individuals who wait until total or severe vision loss over the course of years have an increasingly poor prognosis with regards to visual recovery.

Conclusion

- This case highlights that albeit benign, meningiomas can be locally destructive hence clinicians should be alert to compressive lesions as a potential etiology for unexplained vision loss especially with corresponding changes to the RNFL and GCL.

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