



Traumatic chiasmopathy following mild trauma in a patient with thyroid orbitopathy

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ABSTRACT

Purpose: Traumatic injury to the optic chiasm is rare and most frequently caused by high-velocity head trauma. It classically results in bitemporal hemianopsia and often presents in conjunction with multiple other traumatic injuries, such as skull fractures and cerebrospinal fluid leaks. We present the case of a 40-year-old woman with pre-existing thyroid orbitopathy who struck her forehead after a fall from standing height.

Observations: This patient suffered immediate profound unilateral vision loss from traumatic optic neuropathy and possible optic nerve avulsion. The fellow eye manifested a temporal hemianopsia with delayed retinal nerve fiber layer and nasal hemimacular ganglion cell layer thinning on optical coherence tomography, consistent with chiasmal pathology. Magnetic resonance imaging showed no definitive lesions at the optic chiasm or more posteriorly along the afferent visual pathway.

Conclusions and importance: This patient's severe vision loss suggests that proptosis from thyroid orbitopathy can sensitize the anterior visual pathway to trauma. In this case, we propose that the lack of laxity in the intra-orbital optic nerves allowed transmission of stretching forces to the optic chiasm in the setting of low-velocity blunt trauma.

1. Introduction

Traumatic chiasmopathy — injury to the optic chiasm — is a rare result of head trauma, although cases have been described in the literature since the late 19th century.¹ It most commonly results from closed head injuries secondary to high-velocity mechanisms such as motor vehicle accidents and falls from substantial heights.^{1–4} Similar to other chiasmal lesions, traumatic chiasmopathy results in bitemporal hemianopsia and may or may not be associated with radiographic abnormalities such as optic chiasm enhancement on gadolinium-enhanced magnetic resonance imaging (MRI).⁴ Here, we report a case of traumatic optic neuropathy and chiasmopathy in a patient with thyroid orbitopathy who sustained mild head trauma as a result of a fall from standing height.

2. Case report

A 40-year-old woman presented to her local emergency department with profound acute vision loss after a mechanical fall in which she struck her right brow and forehead against the edge of a bathroom

counter. She was unsure if she lost consciousness, but experienced immediate vision loss in the right eye and noted mild qualitative visual changes in her left eye, including abnormal color perception and portions of letters missing when trying to read. Her medical history was significant for Graves' disease which had been treated with radioactive iodine ablation 12 years earlier, thyroid orbitopathy with bilateral proptosis (Fig. 1A), herpes zoster stromal keratitis of the right eye, and Sjogren's syndrome. She had experienced 2–3 prior episodes of spontaneous luxation of the right globe, which she would reduce manually. However, she denied any vision loss following the previous luxation episodes and felt that her baseline vision in the right eye had been normal. She denied globe luxation at the time of her present injury.

On initial presentation, her visual acuity was bare light perception in the right eye and 20/20 in the left eye without correction, with full confrontational visual fields and Ishihara color plates in the left eye. Her intraocular pressure (IOP) was 31 mm Hg in the right eye and 13 mm Hg in the left eye. Her pupils were round and symmetric, with a pronounced right relative afferent pupillary defect. She exhibited a 3 cm laceration over the right eyebrow, right upper eyelid ecchymosis, and bilateral proptosis with Hertel exophthalmometry measurements of 31 and 27

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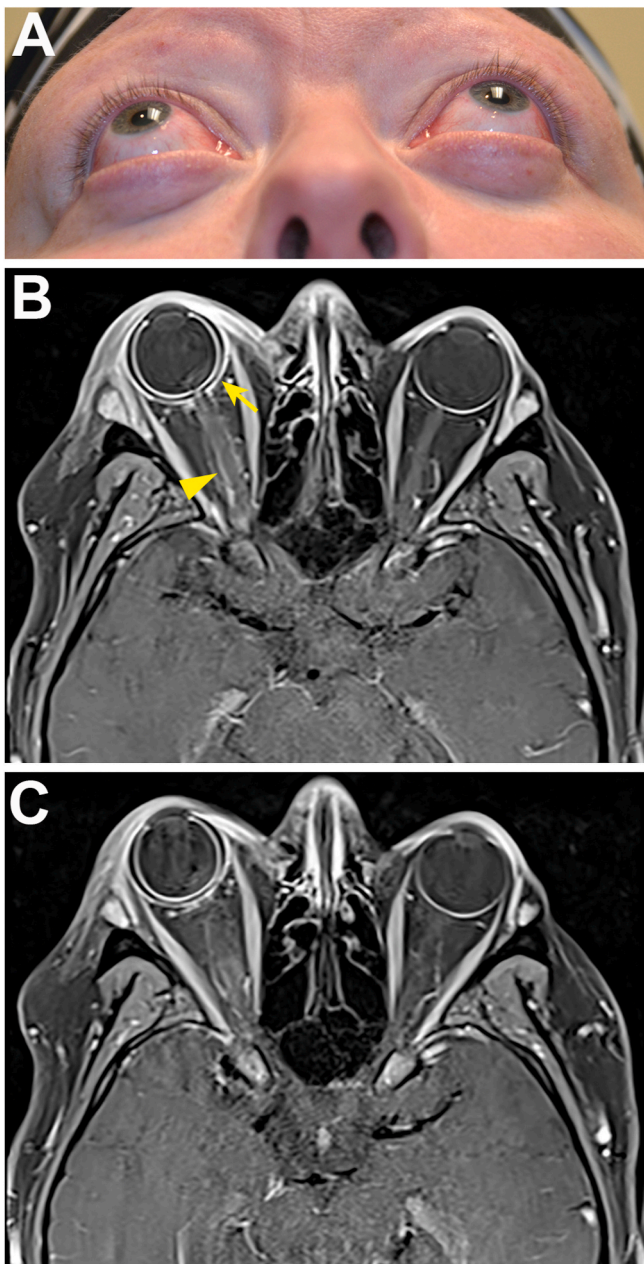


Fig. 1. External photograph and magnetic resonance imaging. **A)** External photograph from an inferior, “worm’s eye view” perspective demonstrating the patient’s bilateral proptosis from thyroid orbitopathy, obtained one year after injury. **B,C)** Gadolinium-enhanced, fat-suppressed T1-weighted magnetic resonance imaging of the orbits at the time of trauma. Right-sided mild vitreous hyperintensity, diffuse scleral enhancement (arrow), and mild enlargement and enhancement of the optic nerve and optic nerve sheath (arrowhead), along with bilateral proptosis are apparent (B). The optic chiasm was poorly captured but shows no evidence of abnormal enhancement (C).

mm in the right and left eyes, respectively. There was trace subconjunctival hemorrhage of the right eye, and the globe was intact. Dilated fundus examination of the right eye revealed a focal plume of dense preretinal and vitreous hemorrhage obscuring the optic disc and macula but an intact retinal periphery. In the left eye, the vitreous was clear, the optic disc had an intact rim with no pallor, hemorrhage, or edema, and the retina was normal. Computed tomography of the brain and orbits demonstrated proptosis and superficial soft tissue swelling around the right brow, with well-formed globes and no bony injury. Her presentation was deemed consistent with severe right traumatic optic

neuropathy with possible optic nerve avulsion, and she was discharged with topical ocular hypotensive agents to decrease the IOP in the right eye.

The following day she presented to the emergency room at our institution at the behest of an outside ophthalmologist, who was concerned about the possibility of optic nerve sheath hematoma. By this time, her vision loss in the right eye had progressed to no light perception (NLP), and she now manifested inferior and nasal hemorrhagic conjunctival chemosis of the right eye, but the examination was otherwise unchanged. MRI of the brain and orbits with gadolinium contrast showed enhancement of the right sclera and enlargement and mild enhancement of the right optic nerve and optic nerve sheath, without evidence of orbital hemorrhage (Fig. 1B and C). There was also bilateral proptosis with increased orbital fat but normal extraocular muscles bilaterally. Conservative management was recommended.

On follow-up one week later, the patient reported no improvement in vision in the right eye, while she was no longer aware of any visual abnormalities in her left eye. She remained NLP in the right eye with an amaurotic pupil and had 20/20 visual acuity with no dyschromatopsia in the left. Her IOP had normalized. There was now no view of the right fundus, and the left fundus appearance was unchanged. Ultrasonography of the right eye showed vitreous hemorrhage and extensive retinal detachment. Unexpectedly, Humphrey visual field testing of the left eye demonstrated a temporal defect respecting the vertical midline (Fig. 2A). Spectral domain optical coherence tomography (SD-OCT) imaging of the left eye demonstrated normal macular architecture and thickness (not shown) and normal peripapillary retinal nerve fiber layer (RNFL) thickness (Fig. 2B). Since the right eye remained NLP, it was unclear if the temporal visual field defect in the left eye represented a homonymous or heteronymous defect. Because evaluation of the optic chiasm had been limited by patient motion, MRI of the orbits was repeated. Although once again subject to significant motion artifact, it showed interval improvement of the scleral and optic nerve enhancement, a normal pituitary gland, and no obvious enhancement or other lesions of the optic chiasm or post-chiasmal optic pathways. Referral to a vitreoretinal surgeon was deferred due to the NLP status of the right eye.

Over the next 6 months there was no visual recovery in the right eye, which had developed a dense cortical cataract. Her proptosis had equalized (26 mm Hertel measurements in each eye). The temporal scotoma in the patient’s left eye persisted but had decreased in severity (Fig. 2A). The patient manifested subtle temporal pallor of the left optic disc, and SD-OCT of this eye revealed interval thinning of the temporal and nasal RNFL (Fig. 2B) and pronounced nasal hemimacular ganglion cell layer (GCL) thinning (Fig. 2C). Visual acuity, perimetry, RNFL thickness, and macular GCL thickness subsequently remained stable in her left eye at last follow-up 15 months from the initial injury.

3. Discussion

The constellation of this patient’s temporal hemianopsia, nasal hemimacular GCL thinning, and nasal and temporal RNFL thinning (“bowtie” optic atrophy) in the left eye are suggestive of co-existing chiasmal/retrochiasmal injury in the context of severe traumatic optic neuropathy with possible optic nerve avulsion in the right eye. Due to the NLP status of the right eye (precluding the identification of a homonymous or bitemporal visual field defect), precise localization of the lesion affecting the left eye cannot be determined: injury to either the optic chiasm or the right optic tract could produce the patient’s pattern of optic atrophy and visual field loss in the left eye. A discrete lesion to the left optic nerve itself is an unlikely alternative, as the pattern of visual field loss and GCL thinning respecting the vertical midline would be highly atypical. Without definitive evidence of intracranial injury on MRI, we strongly favor chiasmal injury as the etiology of the left optic neuropathy. Regarding the right eye, although the patient developed a complete retinal detachment during the week following her injury, the NLP vision with intact peripheral retina observed in her visit to our

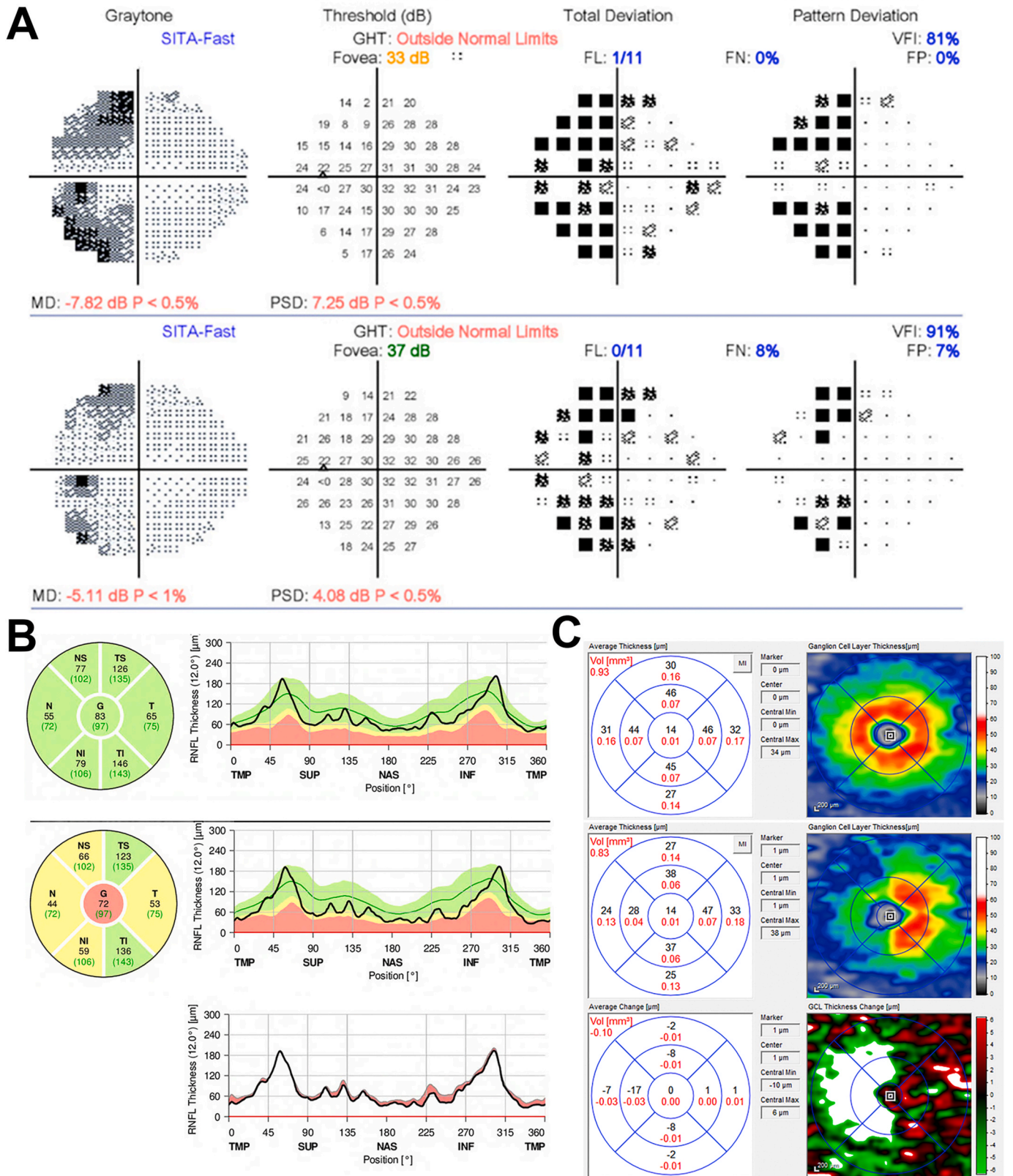


Fig. 2. Ancillary testing at early and late follow-up. A) Humphrey visual field perimetry one week following injury (top panel) and at 6-month follow-up (bottom panel) show temporal defects respecting the vertical midline in the left eye, with modest interval improvement. B) Peripapillary retinal nerve fiber layer (RNFL) thickness measured by optical coherence tomography (OCT) was normal one week following injury (top panel) and showed interval nasal and temporal thinning six months later (middle panel). The bottom panel depicts the interval RNFL thinning in pink. C) Automatically segmented ganglion cell layer (GCL) thickness maps derived from macular OCT scans at one week following injury (top panel) and at 6 months (middle panel). The macular thickness change map (bottom panel) reveals marked interval nasal hemimacular GCL thinning. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

emergency department strongly argues in favor of optic neuropathy as the cause of her acute profound vision loss, rather than retinal injury.

Traumatic chiasmal injury is rare and, as with other chiasmal syndromes, classically results in bitemporal hemianopsia.^{1–5} Asymmetric vision loss between the two eyes is common, and several cases of profound unilateral vision loss with temporal visual field defects in the fellow eye have been reported.^{2,3,5} High-velocity closed head injuries – particularly motor vehicle accidents – are the most common mechanism of injury, and traumatic chiasmopathy is often accompanied by other cranial neuropathies, cerebrospinal fluid leaks from calvarial fractures, and endocrine dysfunction from pituitary injury.^{2–8} As in this case, neuroimaging frequently does not reveal lesions of the optic chiasm at the time of injury.^{2,5} Multiple pathogenic mechanisms for traumatic chiasmopathy have been proposed, including direct mechanical injury to crossing axons, ischemia from shearing of vessels perfusing the chiasm, and compression from perichiasmal hematomas or cerebral edema.^{3,9,10}

The nature of the inciting trauma in this case – a fall from standing height – is remarkable for its relatively low impact velocity. We propose that this patient was vulnerable to anterior optic pathway injury due to her pre-existing proptosis from thyroid orbitopathy, which had been severe enough to produce several prior episodes of spontaneous globe luxation. Proptosis-related tension on the optic nerves could make them more susceptible to injury, with the potential to propagate stretching forces posteriorly to the optic chiasm and to damage decussating axons from the fellow eye. Our patient was uncertain if she suffered direct ocular trauma, which could have resulted in a sequence of rapid retro-pulsion and recoil of the right globe within the orbit to produce acute stretching of the optic nerve. Her subsequent development of a traumatic cataract would seemingly argue in favor of direct ocular trauma. However, the more common mechanism of rapid deceleration from blunt head trauma could also have produced stretching forces leading to this patient's optic neuropathy and chiasmopathy.

RNFL thinning on OCT has been reported in one prior case of traumatic chiasmopathy,¹¹ but to the best of our knowledge ours is the first report to show hemimacular GCL thinning and to present serial OCT images of the RNFL and macula to illustrate the development of partial optic atrophy in this setting. These inner retinal changes are consistent with slow retrograde degeneration of retinal ganglion cells following distal axonal injury. Such OCT abnormalities represent the anatomic correlate to the patient's temporal hemianopsia and confirm injury to chiasmal or retrochiasmal ganglion cell axons despite the absence of overt pathology on MRI.

4. Conclusions

In summary, this case evinces that even relatively mild trauma can cause optic nerve and chiasmal injury when idiosyncratic orbital anatomy – in this case, proptosis from thyroid orbitopathy – eliminates the physiological laxity of the intra-orbital optic nerve. In this setting, chiasmal injury can occur without concomitant injuries, such as skull fracture, CSF leak, endocrine dysfunction, and neurological injury. Use of the GCL-segmented OCT macular thickness map in this case was more informative in localizing the underlying lesion along the visual pathway

than the more commonly used peripapillary RNFL thickness analysis.

Patient consent

The authors thank the patient, who has provided written informed consent for publication of this case report and accompanying images.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Credit author statement

Matthew L. O'Sullivan: writing – original draft; formal analysis; visualization

Sidney M. Gospe III: conceptualization; formal analysis; writing – review & editing;

Declaration of competing interest

The authors declare that they have no competing interests.

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