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## Journal

UCLA Radiological Sciences Proceedings, 4(3)

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## Publication Date

2024

## DOI

10.5070/RS44353362

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# Tilted Disc Syndrome with Bitemporal Hemianopia in a 67-Year-Old Woman with High Myopia and Mixed/Combined-Mechanism Glaucoma: A Report of a Rare Case

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*UCLA Radiol Sci Proc.* 2024;4(3):38-44

**Abstract:** Bitemporal hemianopia typically results from compression of the optic chiasm by sellar, suprasellar, or chiasmal lesions. Most of the cases of bitemporal hemianopia are secondary to pituitary masses. Defects in the temporal half of the visual field that mimic those that are caused by such pituitary or chiasmal lesions are known as bitemporal "pseudohemianopia" and involve orbital pathology. Tilted disc syndrome is an eye anomaly that may result in bitemporal visual field deficits similar to those that are caused by extrinsic or intrinsic mass effect on the optic chiasm. We report an incidentally found tilted disc syndrome in a patient with a history of surgically treated high myopia and the symptoms of bilateral, gradual vision loss.

**Keywords:** *bitemporal hemianopia, tilted disc syndrome, optic chiasm, mixed/combined-mechanism glaucoma, high myopia, primary open-angle glaucoma, anatomically narrow iridocorneal angle*

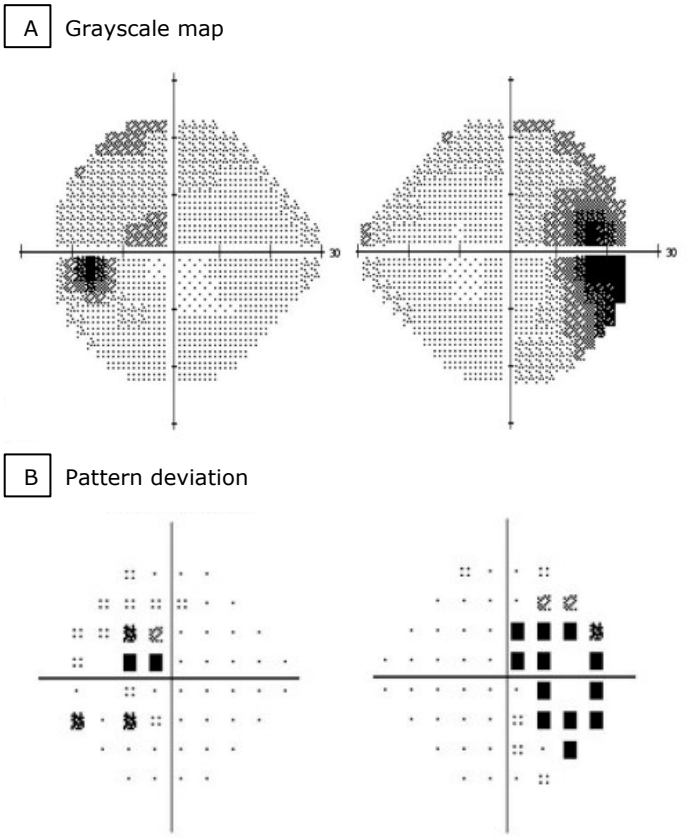
**Key Points**

- Tilted disc syndrome may result in bitemporal hemianopia mimicking that of intrinsic or extrinsic compression of the optic chiasm.
- A high index of suspicion for tilted disc syndrome should guide the radiologist in the evaluation of the visual pathway when sellar, suprasellar, or chiasmal lesions are absent.
- Tilted disc syndrome may be associated with both high myopia and mixed/combined-mechanism glaucoma.

## Case Presentation

A 67-year-old Egyptian woman with a medical history of hyperlipidemia, hypertension, not visually significant nuclear sclerotic cataract, high bilateral myopia that was treated with laser-assisted in-situ keratomileusis (LASIK) and refractive surgery, and suspected glaucoma of both eyes presented for an ophthalmologic examination after experiencing gradual vision loss. The patient also had elevated intraocular pressure (IOP) with numbers reaching 22 mmHg and 24 mmHg in the right eye and the left eye, respectively. Although initially the patient's glaucoma was categorized as primary open-angle glaucoma (POAG), a slit-lamp examination revealed shallow anterior chambers, and the subsequent gonioscopy showed D30s and D25s iridocorneal angles (Spaeth gonioscopic grading system) in the right eye and the left eye, respectively, as well as peripheral anterior synechia present superiorly in the left eye, suggestive of anatomically narrow, potentially occludable, angles. The patient's uncorrected visual acuity was 20/60 on the right eye and 20/30 on the left eye. The result of a Humphrey visual field 24-2 test showed bilateral temporal visual field defects (Figure 1), supratemporal and

**Figure 1.** Humphrey Visual Field 24-2 Test of a 67-Year-Old Woman with Mixed-Mechanism Glaucoma and Gradual Vision Loss.




(A, B) The test reveals bitemporal visual field defects – supratemporal and infratemporal defect in the right eye and temporal paracentral defect in the left eye – that do not cross the vertical midline.
(A) The grayscale plots illustrate raw threshold sensitivity values for light stimuli. Clusters of high threshold points lie in the peripheral visual fields.
(B) The pattern deviation probability plots highlight points of visual field depression in the peripheral visual fields, statistically significant against normalized age-matched groups.

infratemporal defects in the right eye and temporal paracentral defect in the left eye. Optical coherence tomography (OCT) performed on ophthalmic imaging platform Spectralis SD-OCT (Heidelberg Engineering GmbH), revealed abnormal thinning in the right nasal quadrant as well as in the left temporal, the superior, and the inferior quadrants of the retinal nerve fiber layer (RNFL) compared with normative data for RNFL thickness in the Egyptian population[1] (Figure 2). Subsequent funduscopic examination (Figure 3) showed the right eye with inferonasal tilt of the optic disc and a significant, predominantly γ-zone, peripapillary atrophy (PPA) as well as the left eye with a less tilt of the optic disc inferiorly and a moderate β-zone PPA.

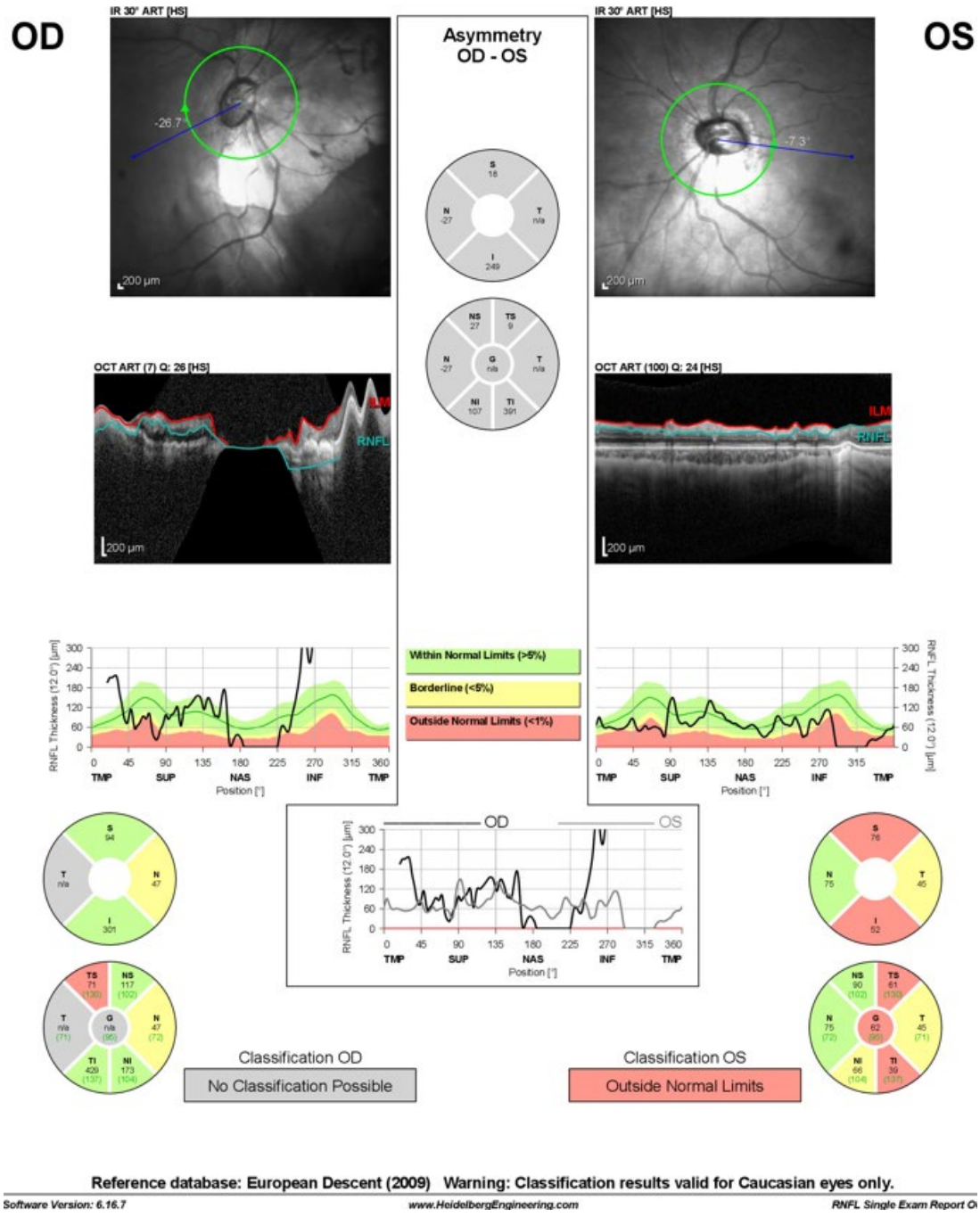
After the initial diagnosis of tilted disc syndrome (TDS) was established based on the results of ophthalmologic examination, the patient was referred for magnetic resonance imaging (MRI) of the pituitary to exclude underlying chiasmal or pituitary lesions. No intracranial abnormalities were identified on MRI (Figure 4A) but flattened temporal aspects of the posterior sclera and slight oblique insertion of the optic nerves were noted (Figure 4B). The presence of tilted discs explained the temporal visual field defects, and the diagnosis of tilted disc syndrome was confirmed based on the results of ophthalmologic and radiologic examinations.

To lower IOP, the patient was treated with prostaglandin analog, latanoprost 0.005%. In addition, to prevent an acute occlusion of the iridocorneal angle, the patient was treated with laser peripheral iridotomy. Subsequently, the patient underwent a cataract extraction surgery of the left eye and implantation of an intraocular lens. Following these treatments, the patient's condition improved, and there was no need to continue the IOP-lowering therapy.

## Discussion

The temporal visual field input is relayed by the nasal retinal fibers, which then decussate at the optic chiasm positioned superior to the sella turcica, before projecting into the contralateral visual cortex in the occipital lobe. Disruption of these fibers at any segment along the visual pathway may produce peripheral vision loss in half of the visual field of one or both eyes, termed hemianopia. Chiasmal, and specifically pituitary, lesions are the most common cause of bitemporal hemianopia, and therefore, MRI is used to evaluate for intracranial pathology when patients present with such deficits of the visual field.[2] When these neuraxial abnormalities are not present, subsequent consideration should be given to less common orbital anomalies, including tilted disc syndrome, which affects between 0.4% and 3.5% of the general population.[3]

**Figure 2.** Optic Coherence Tomography of a 67-Year-Old Woman with Mixed-Mechanism Glaucoma and Gradual Vision Loss



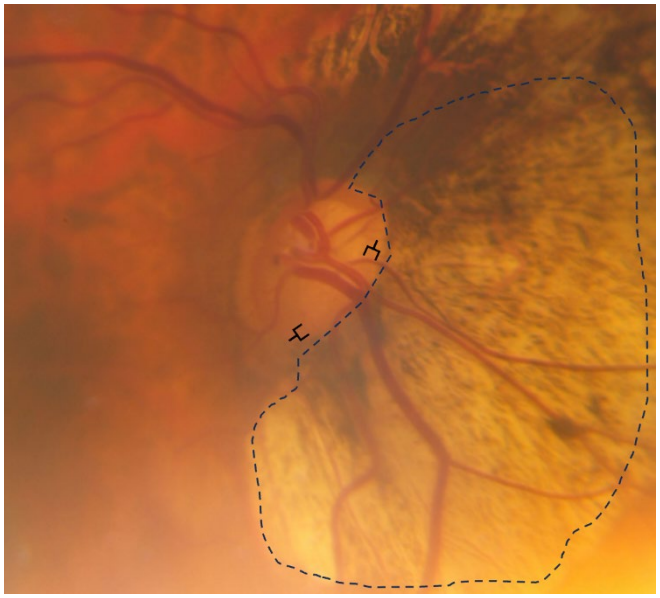

The right eye (OD) shows thinning of the retinal nerve fiber layer (RNFL) in the nasal quadrant. Because of the tilt of the optic disc, RNFL displacement, and the thinnest distribution of RNFL in the temporal quadrant (especially in patients with high myopia as it occurred in our patient), the temporal measurement was not possible to calculate. The inferior quadrant is artifactually thick because of the segmentation errors (OD tomogram, blue/teal line). The left eye (OS) shows abnormal thinning in the inferior, the superior, and the temporal quadrants, with a thickness of 52 µm, 76 µm, and 45 µm, respectively.

[Normative data[1] acquired with Spectralis SD-OCT for RNFL thickness in the Egyptian population: The mean (G)—101.74±10.05 µm (range: 79.0–123.0 µm). The mean (N)—80.0±14.1 µm (range: 53.0–119.0 µm). The mean (T)—73.72±11.8 µm (range: 51.0–137.0 µm). The mean (NS)—105.79±21.4 µm (range: 67.0–166.0 µm). The mean (TS)—137.8±17.04 µm (107.0–181.0 µm). The mean (NI)—114.8±26.15 µm (range: 57.0–172.0 µm). The mean (TI)—147.9±19.13 µm (range: 107.0–179.0 µm)]

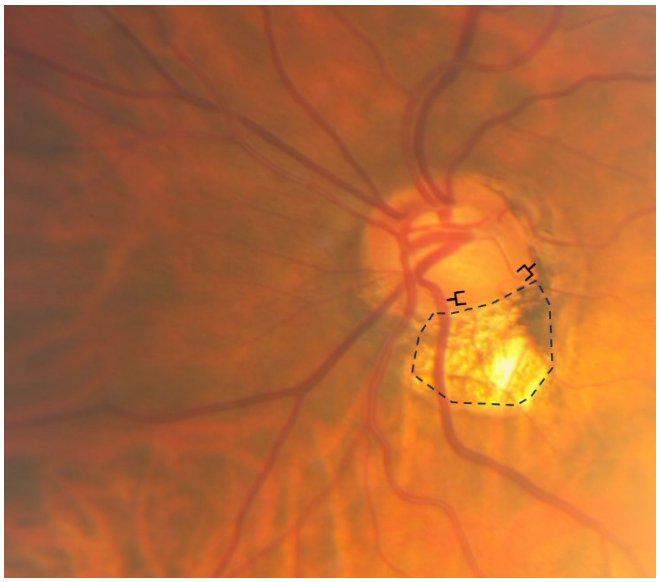
Abbreviations: G, global sector; N, nasal sector; T, temporal sector; NS, nasal superior sector; TS, temporal superior sector; NI, nasal inferior sector; TI, temporal inferior sector.

**Figure 3.** Fundoscopic Examination of a 67-Year-Old Woman with Glaucoma and Gradual Vision Loss

A Right eye



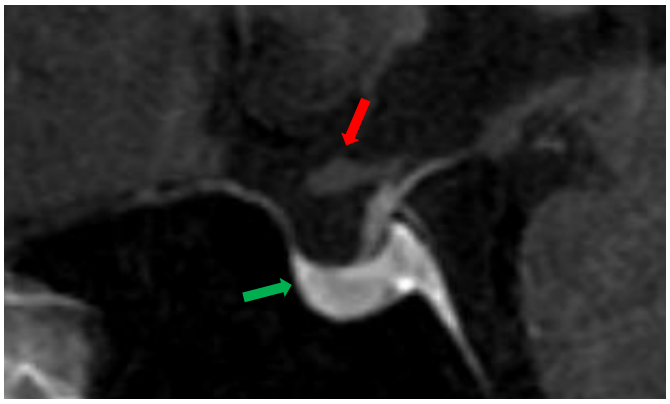
B Left eye



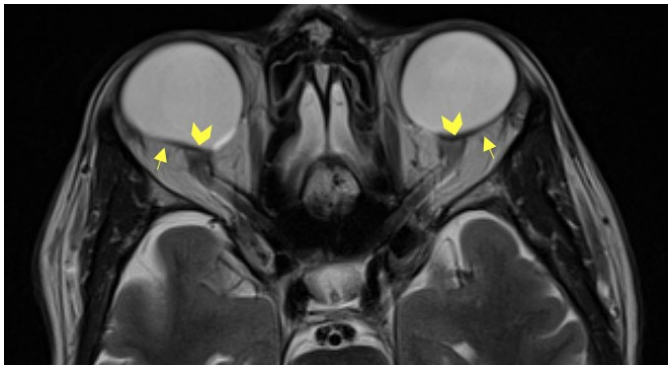
(A) The right eye shows inferonasal tilt of the optic disc and neuroretinal rim thinning (A, area between brackets) with marked peripapillary atrophy and staphylomatous appearance (A, area outlined by dashes) especially in the inferonasal quadrant.
(B) The left eye shows less tilt of the optic disc inferiorly, more moderate thinning of the neuroretinal rim (B, area between brackets) and less peripapillary atrophy (B, area outlined by dashes), most significant in the inferior aspect.

**Figure 4.** Magnetic Resonance Imaging (MRI) of the Pituitary Gland and the Orbits in a 67-Year-Old Woman with Bitemporal Hemianopia, Glaucoma, and Gradual Vision Loss

A T1-weighted MRI, sagittal view



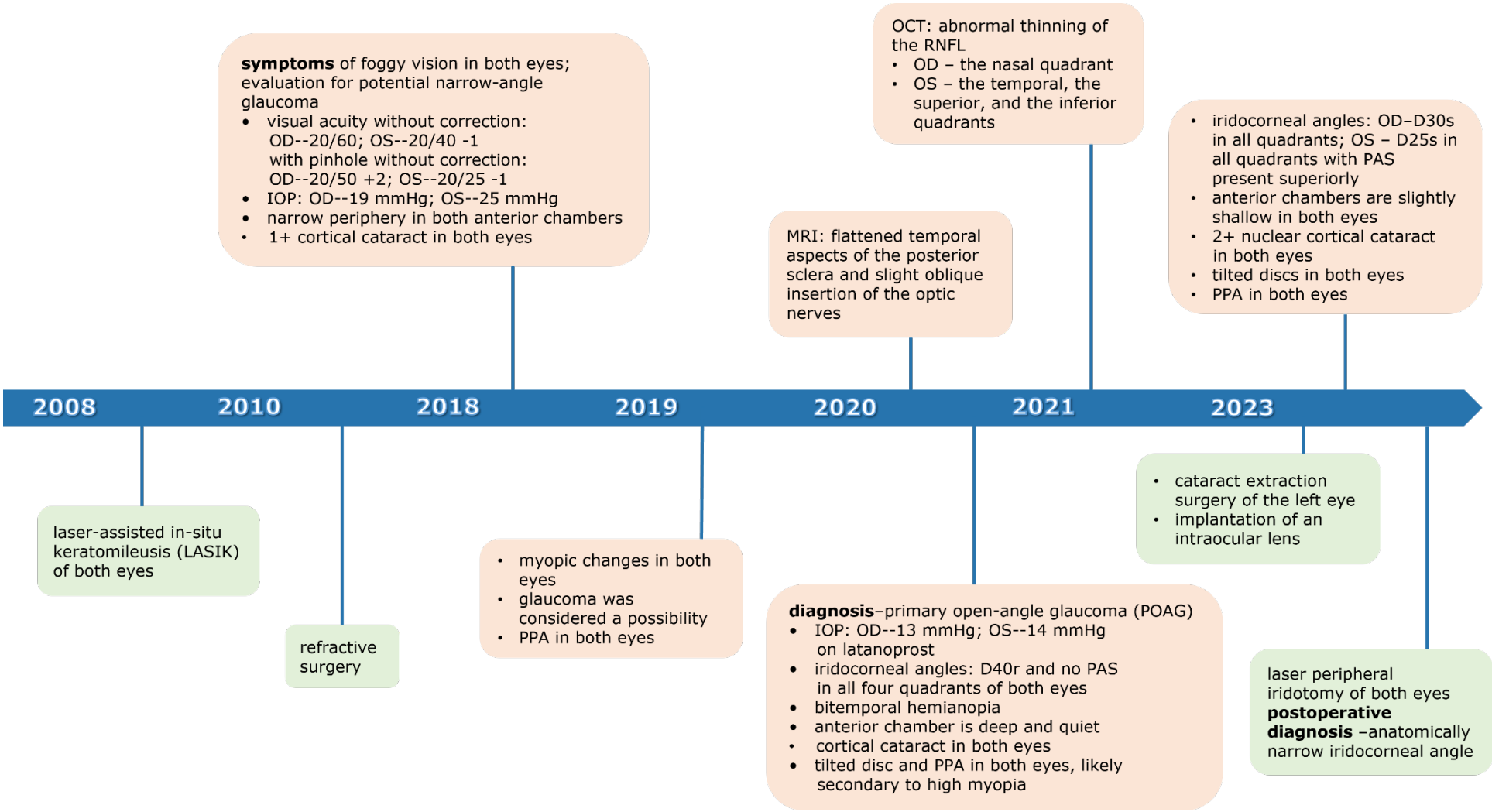
B T2-weighted MRI, axial view



(A) T1-weighted MRI of the pituitary gland, sagittal view, shows normal pituitary gland (A, green arrow) and the optic chiasm (A, red arrow). (B) T2-weighted MRI, axial view, at the level of the orbits shows flattened appearance of the temporal sclera (B, arrows) of the posterior globes with oblique insertion angle of the optic nerve (B, arrowheads).

Orbital development begins in the third week of gestation once the neural plate has formed.[3,4] Lateral bulges grow outward from the neuroectoderm to become optic vesicles and later invaginate to form a double-layered optic cup and the optic fissure.[3,4] The edges of the optic fissure fuse around the sixth week of gestation and envelop the optic stalk.[3,4] In individuals with a congenital form of TDS, an incomplete closure of the optic fissure leads to focal hypoplasia and thinning of the retinal and the choroidal epithelium, predominantly in the inferonasal aspect.[3,5] This presents an impediment to axonal development in the defective retinal segments, subsequently manifesting as visual field defects.[5]

**Case report timeline**.




Abbreviations: IOP, Intraocular pressure; LASIK, laser-assisted in-situ keratomileusis; OCT, optic coherence tomography; OD, the right eye; OS, the left eye; MRI, magnetic resonance imaging; PAS, peripheral anterior synechia; POAG, primary open-angle glaucoma; PPA, peripapillary atrophy; RNFL, retinal nerve fiber layer

While TDS can be unilateral, it was found bilaterally in the wide range, between 22% and 89%, of the cases, presumably because of wide variation in classification of the tilted disc.[6,7] Bilateral TDS may cause visual defects characterized by bitemporal hemianopia mimicking chiasmal compression.[3,8] The spectrum of severity of tilted disc syndrome depends on the degree of the fundus involvement, from mildly tilted disc to severe fundal deformity.[9]

It is believed that tilted discs may occur not only as a congenital condition, but also as an acquired condition caused by the progression of myopia.[3,10] As the eye elongates in an anterior-posterior orientation during childhood, especially in those with progressing axial myopia, the optic nerve insertion (and therefore optic disc) displays increasing tilt with levels of myopia rising.[3,10] Although association between myopia and tilted disc is still debated,[6] it is possible that the optic disc tilt described in this case report was the result of high myopia noted in our patient's medical history. The prevalence of myopia and myopic open-angle glaucoma in populations with tilted discs[6] is between 57% and 81%. However, myopia is rarely found in subjects with narrow iridocorneal angle[11] that was observed in our patient, and mixed/combined-mechanism glaucoma is an uncommon (2.3%) condition.[12] In addition, in our search of literature, we did not find reported cases of mixed/combined-mechanism glaucoma that co-occurred with TDS, which makes us believe that our case is a first reported co-occurrence of associated tilted disc syndrome, high myopia, and mixed/combined-mechanism glaucoma.

On conventional MRI of patients with tilted discs, the orbits appear with increased anterior-posterior dimension and decreased nasal insertion angle of the optic nerves.[13] On 3D MRI and swept-source OCT, the structural changes include asymmetry of the shape of the eyes, outward protrusion of the lower part of the posterior eye segment, oblique attachment of the optic nerve at the upper nasal edge of the protruded segment, protrusion of the upper edge of the Bruch's membrane,[3] and a flattened appearance of the temporal sclera,[10] with the latter being observed on MRI of our patient.

While visual field defects caused by TDS typically cross the vertical midline (which did not happen in our patient), visual field defects caused by chiasmal lesions generally respect the vertical midline.[2,4,7,8] Most commonly, TDS-associated visual field defects are located in the superior temporal quadrant.[3,8] Although the cases of bitemporal hemianopia, especially those in which visual field defects do not cross the vertical midline, necessitate an immediate use of MRI and/or CT to determine the presence of intracranial pathology,[4,8] as it occurred in our patient, the diagnosis of TDS can be made based on the results of funduscopy, fundus photography, and visual field testing.[7,10]

Appropriate ophthalmologic evaluation not only aids in diagnosis of patients with myopia-related tilted disc syndrome and glaucoma, but also provides essential information for prevention and timely treatment of structural and functional changes in the eye caused by these conditions.[10]

## Conclusion

We reported a case of tilted disc syndrome with bitemporal hemianopia which, to our knowledge, has not been reported in association with mixed/combined-mechanism glaucoma. Although in most of the cases, bitemporal hemianopia is caused by sellar, suprasellar, or chiasmal lesions,[2,3,8] it can also be caused by other conditions, including retinopathy[14] as well as abnormalities of the optic disc.[3,8] In cases of bitemporal hemianopia, patients are generally referred for neuroimaging[3,4,8] that is essential for evaluation of the visual pathway for the presence of sellar/suprasellar tumors and other less common causes of chiasmal syndrome. For this reason, radiologists should know about orbital causes of bitemporal hemianopic field defects, including those caused by TDS, and a high index of suspicion for these causes should guide the evaluation of the visual pathway when sellar, suprasellar, or chiasmal lesions are absent.

### Author Contributions

Conceptualization, N.P.; Acquisition, analysis, and interpretation of data, C.J. and J.W.; Writing – original draft preparation, C.J.; Review and revisions, C.J. and J.W.; Supervision, N.P. All authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately

investigated and resolved. All authors had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis.

## Disclosures

None to report.

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