Clinical Reasoning: Wilbrand’s Knee, Scotoma of Traquair, and Normal Tension Glaucoma

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Keywords
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Abstract
An otherwise healthy 63-year-old woman was given a diagnosis of normal tension glaucoma (NTG) in the right eye (OD) 2 months before presentation. Standard computerized perimetry showed a unilateral right hemianoptic temporal field defect. On examination visual acuity was preserved, intraocular pressure was normal, there was a right relative afferent pupillary defect (RAPD) with an asymmetric cupping of the disc, but no pallor. Brain magnetic resonance imaging (MRI) showed a meningioma compressing the right optic nerve at its junction with the chiasm. Compressive disorders on the anterior chiasm, albeit rarely, may cause cupping of the disc and unilateral temporal visual field defect (junctional scotoma of Traquair) with normal visual acuity that should be considered in the differential diagnosis of NTG.

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Introduction

Normal tension glaucoma (NTG) is a relatively common form of open-angle glaucoma in which there is no measured elevation of the intraocular pressure (IOP). When the IOP is elevated and the disc is cupped, the diagnosis of glaucoma is straightforward. NTG, on the contrary, may simulate other ocular or neurological conditions. Most concerning to the clinician is an intracranial tumor masquerading as NTG. In the latter case, there is usually an evident pallor of the disc, a disproportionately reduced visual acuity with a visual field defect different from those usually seen in glaucoma (e.g., a nasal step). However, initially visual acuity can be preserved and the disc may appear normal with no pallor. The present case was misdiagnosed as NTG: clinical features of differential diagnosis between compressive optic neuropathy and NTG are discussed with regard to anatomy (Wilbrand’s knee) and type of visual field defect (junctional scotoma of Traquair [JST]).

Case Description

An otherwise healthy 62-year-old nurse presented to her ophthalmologist complaining of painless mild blurred vision in the right eye (OD). Best corrected visual acuity (BCVA) was 20/20 in both eyes (OU), slit lamp examination was unremarkable, and IOP was 17 mm Hg with a normal central corneal thickness OU. On fundus examination, an asymmetric cupping of the disc (right larger than left) was noted and visual field showed a unilateral temporal field defect OD (Fig. 1). A diagnosis of NTG was made. On our examination, 2 months later, BCVA was unchanged. Color vision tested with Ishihara plates was 12/12 OU, but reading was slower in the right eye. A right relative afferent pupillary defect was noted. Optical coherence tomography (OCT) of the optic nerves showed a reduced retinal nerve fiber layer (RNFL) thickness temporally OD, but was normal in the left eye (OS). Ganglion cell complex (GCC) analysis was reduced OD compared with OS (normal) (Fig. 2). Fundus examination was unremarkable except for an asymmetric cupping of the discs, with right larger than left (Fig. 3). No pallor of the right optic disc was evident. The clinical presentation was suspect for right compressive optic neuropathy, and brain magnetic resonance imaging showed a meningioma compressing the right optic nerve at its junction with the chiasm (Fig. 4). After surgical removal, visual field improved (Fig. 5) and BCVA remained stable.

Discussion

In 1904, the German ophthalmologist Herman Wilbrand in his book “The Neurology of the Eyes” had reported that nasal decussating optic nerve fibers detour into the contralateral optic nerve before entering the optic chiasm [1, 2]. The “Wilbrand’s knee” added interest to visual field interpretation, mostly when the Scottish ophthalmologist Traquair [3] described a unilateral temporal scotoma of the visual field in case of anterior chiasm compression, thus “proving” a clinical “evidence” of Wilbrand’s knee. This was understood as the “JST.” For years, the anatomical explanation for this unilateral temporal scotoma was the selective damage of crossed nasal fibers in the Wilbrand knee at the junction between the nerve and the chiasm. However, in 1997, Horton [4] proved that “Wilbrand’s knee of the primate optic chiasm is an artifact of monocular enucleation.” In a few words, if one eye had been previously enucleated, there was a subsequent shifting of intact fibers at the chiasm once retrograde degeneration in one optic nerve had occurred. Later, in 2014, Shin and coworkers [5] reported evidence for Wilbrand’s knee in the normal human optic chiasm by means of anisotropic reflecting properties
of optic axons. Horton showed that the knee imaged using this technique was a photographic artifact as well [6]. Despite the lack of evidence for a true Wilbrand’s knee, compression of the chiasm does not always cause a bitemporal hemianopia but is able to produce a variety
Fig. 3. Color retinography shows an increased cupping of the disc OD, but no pallor of the neuroretinal rim.

Fig. 4. Brain magnetic resonance imaging. Left: coronal view demonstrates a mass (white arrow) in the sellar region. Middle: axial view of the meningioma (dotted arrow) in the right side of the suprasellar cistern. Right: magnification of coronal view of the sellar region shows that the mass (asterisk) compresses the right junction of optic nerve and chiasm (red arrow).

Fig. 5. Standard computerized 30-2 perimetry 1 month after surgery shows improvement of the visual field defect OD (right image) with a residual inferior fascicular defect.
of visual field defects. In a retrospective case series of 53 patients with chiasmal compression, bitemporal hemianopia was noted in 14 (26%), whereas the junctional scotoma and monocular visual field defect were found in 34% and 7% of cases, respectively [7]. Thus, bitemporal hemianopia is found in only one of 4 patients with chiasmal compression, which, in turn, is able to cause a true monocular visual field defect. Whereas the former is caused by stretching of the nasal crossing fibers before and more than uncrossed temporal fibers, the latter is due to distal (close to chiasm) optic nerve compression.

In short, the anterior chiasmal syndrome can present with 2 types of junctional visual field loss. The JST refers to a monocular hemianopic field defect, which can be nasal or temporal. By contrast, the junctional scotoma refers to ipsilateral central field loss in one eye and contralateral superotemporal field loss. The latter is explained from involvement of ipsilateral optic nerve fibers and crossing inferior nasal fibers from the opposite side.

In addition to visual field, a more objective method to evaluate chiasmal compression is OCT. GCC analysis is given more and more importance in the study of compressive lesions. GCC loss is typical and can be seen with minimal or no detectable visual field loss [7, 8] or be detected before loss of the RNFL [8]. Macular GCC analysis is particularly useful as the loss of the normal nasal-temporal asymmetry may be a very early sign of chiasmal compression [9]. Tomographic thinning of the temporal peripapillary RNFL is not typical of glaucoma and should be looked at with suspect for an alternative cause, as was in our case [10]. Neuro-ophthalmologic evaluation with both visual field testing and OCT is essential in the evaluation of all patients with lesions of the chiasm.

Selective damage of nasal crossing fibers or temporal fibers at the junction of the optic nerve with the chiasm can produce a junctional scotoma or a JST, respectively. In these cases, the clinician may inadvertently miss a compressive intracranial cause and focus attention on an intraocular cause of field defect. Interestingly, an expanding mass in the sellar region may asymmetrically compress the chiasm or compress the distal optic nerve(s) before the chiasm producing a true monocular visual field defect with a fascicular defect [11, 12]. To add confusion in the clinical evaluation of these cases, one has to consider that in compressive optic neuropathy, the optic nerve may appear normal initially.

“Cupping” is a term used to describe enlargement of the cup-to-disc ratio and is widely recognized as a feature of glaucoma. However, it is not pathognomonic of glaucoma and may be seen in genetic (such as dominant optic atrophy) or acquired diseases (e.g., ischemic optic neuropathy) [10].

Moreover, a raised cup/disc ratio has been demonstrated in compressive optic neuropathy as well [13]. When the disc is cupped and the IOP is high, the diagnosis of glaucoma is probable. Diagnosis becomes challenging if IOP remains within the generally considered normal limits (21 mm Hg) and optic disc cupping is present.

Trobe and coworkers [14] have shown that 44% of eyes with nonglaucomatous optic atrophy were misdiagnosed as glaucoma in fundus stereophotographs by at least one experienced observer. This happens because pallor, usually suggestive of compression rather than glaucoma, is a subjective sign. History of the patients and previous records may help in the diagnosis and rapid vision loss not consistent with the degree of cupping is a helpful sign.

In fact, visual function (color and acuity) is usually preserved in glaucoma until later stages because of the relative sparing of the papillomacular bundle [15, 16]. However, even in compressive optic neuropathy, visual acuity may be normal initially [11]. The clinician may thus face the case of a fascicular monocular visual field defect with normal visual acuity and an unremarkable (or cupped) optic disc with not so much pallor, and easily led to diagnose an NTG.

Neuroimaging for all patients with a diagnosis NTG is a controversial issue probably unnecessary due to the low yield for detecting intracranial pathology. Nevertheless, up to 8%
of patients diagnosed with NTG harbor a compressive lesion of the anterior visual pathways [15] as compared to 0.8 in 100,000 in nonglaucomatous (normal) patients [17]. Moreover, normal intracranial structures, such as arteries, may account for a number of NTG because of dolichoectasia and vascular conflict with the intracranial portion of the optic nerve [18, 19].

Both neurologists and ophthalmologists should be familiar with the interpretation of visual field defects. Relatively early specific glaucomatous visual field defects include a nasal step defect obeying the horizontal meridian and the classic arcuate defect, which is a comma-shaped extension of the blind spot [20]. Thus, glaucoma usually causes nasal field defects although Wall and coworkers [21] recently showed that nearly half of glaucoma patients with mild central visual loss have defects in the inferotemporal visual field. The key point is that a temporal visual field defect with respect to the vertical midline with a relative normal optic disc appearance should not be interpreted as glaucoma.

Consequently, the most useful single feature is the disc/field correlation and the potential mismatch between visual function (visual acuity and color vision), visual field, and optic nerve appearance. Our case illustrates the need for clinicians to be vigilant in case of (presumed) NTG. A full neuro-ophthalmologic assessment of the optic nerve, including acuity, pupils, color vision, visual field, and fundoscopy, is surely within the skills of a competent ophthalmologist and could reduce the diagnostic delay.

The aforementioned tests should be mandatory before a diagnosis of NTG is made. In our opinion, features such as field loss respecting the vertical meridian, optic disc pallor, reduced color vision, right relative afferent pupillary defect, and, particularly, disc/field mismatch should alert the clinician to the possibility of a compressive lesion. OCT (RNFL and GCC) may help the clinician and may be abnormal even when visual field is still intact. Our case shows how even a monocular temporal visual field defect (JST) respecting the vertical meridian may be misunderstood as NTG if the optic disc appears cupped but not pallid in a patient with a preserved visual acuity.

Statement of Ethics

This research was conducted ethically in accordance with the Declaration of Helsinki. This retrospective review of patient data did not require ethical approval in accordance with local/national guidelines. Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Francesco Pellegrini: supervision and final editing. Alessandra Cuna: image collection. Daniele Cirone: English editing. Cristina Ciabattoni: case description. Ettore Caruso: MEDLINE search. Antonio Zappacosta and Emanuela Interlandi: discussion.
Data Availability Statement

All data generated or analyzed during this study are included in this article. Further enquiries can be directed to the corresponding author.

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