ORIGINAL ARTICLE

Idiopathic Acquired Temporal Wedge Visual Field Defects

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ABSTRACT

Our aim is to report 13 unusual cases of acquired, temporal sectoral scotomas. Such stationary “wedge” field defects have been reported previously in cases of presumed *congenital* nasal hypoplasia of the optic disc and as a complication of vitreoretinal surgery. To our knowledge, the literature provides no reports of similar defects occurring spontaneously. This is a descriptive clinical case series of 13 patients presenting with sub-acute monocular temporal visual field loss. All were clinically assessed and investigated with Goldmann perimetry, automated Humphrey visual fields, retinal optical coherence tomography, orbital ultrasound, and standard and multi-focal electroretinography. Cases were followed with serial perimetry for a mean of 3.9 years (range: 6 months to 10 years). Goldmann and Humphrey perimetry both demonstrated “wedge”-shaped defects extending temporally from an apex contiguous with, or lateral to, the blind spot. There was no evidence of optic disc drusen, glaucoma, disc hypoplasia, or focal retinitis. Sectoral optic disc swelling was not present in any patient at presentation. In all cases, the visual field defect remained stable. One patient developed a similar defect in the fellow eye after an interval of 5 years. Here we describe 13 cases of acquired, stationary temporal wedge scotomas, novel in the literature. Although the aetiology is uncertain, we propose damage to the nasal rim of the optic disc as a likely mechanism.

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Introduction

The trajectory of retinal nerve fibres at the posterior pole has been described: axonal bundles traverse toward the disc by three distinct routes.¹ The papillo-macular, or caeco-central projection communicates directly from the nasal macula to the temporal aspect of the disc; insults here result in a caeco-central scotoma. Secondly, fibres of temporal and macular ganglion cells project away from the horizontal raphe in an arc towards ever more superior or inferior aspects of the disc; damage to these crescents manifest as arcuate scotomas that respect the horizontal midline. Finally, temporal to the disc, as there is no horizontal raphe, axons transverse directly to the point of the disc closest to them in a radial fashion; lesions here result in wedge-shaped scotomas extending from the blind spot.²

With this in mind, the pattern of visual field loss can point towards fibres damaged by disease processes preferentially deleterious to one bundle or another; for example, arcuate defects in glaucoma² or caeco-

central scotomas in toxic optic neuropathies.³ Temporal defects are somewhat rarer and less firmly associated with particular disease processes. Here we present 13 patients seen at our units with such signs. Detailed descriptions of three of the cases precede a discussion of possible causes and associations of acquired, stationary temporal wedge defects.

Materials and methods

This is a retrospective, observational case series carried out in accordance with the declaration of Helsinki. The local ethics committee (National Health Service Research Ethics Committee [England]; NHS REC) ruled that approval was not required for this study. Clinical notes and digital imaging were reviewed and summarised in tabular form with detailed descriptions of three illustrative cases. A literature search was initiated with an interrogation of the MEDLINE database via PubMed with the term “Temporal visual field

defect,” then with abstracts refined, papers of interest reviewed, and further literature explored from references therein.

Results

In the interests of brevity, all 13 cases are summarised in Table 1, with 3 cases described here in more detail for illustrative purposes.

Case 1

A 57-year-old woman presented to eye casualty having become aware of a “film” over her right eye. There were no antecedent events or associated symptoms. Ocular examination including confrontation visual fields, 1 week after symptom onset, was recorded as normal. She had a past history of myocardial infarction, hyperlipidaemia, and Raynaud’s phenomenon.

Table 1. Summary of clinical cases.

Patient	Age at diagnosis and sex	Presentation	Visual acuities	Refraction (spherical equivalent)	Intraocular pressure (mm Hg)	Humphrey visual field	Goldman perimetry	OCT—Nasal quadrant RNFL thickness (µm; normal range: >60)	Follow-up
1	57 F	“Film” over right eye	6/9 N5 OD	∞ OU	13 OU	Temporal wedge OD	Wedge to blind spot OD	Not available at first presentation—see patient 1b	10 years
1b	62 F	Visual change in left eye	6/9 N8 OS 6/9 N6 OU	∞ OU	14 OU	Temporal wedge OS	Not available	46 OD 38 OS 60 OD	3 years
2*	56 F	Could not see husband on the left	6/5 N6 OU	−3.50 D OD −7.00 D OS	13 OD 23 OS	Temporal wedge OS	Wedge to blind spot OS	35 OS 63 OD	10 years
3	36 M	Missed object moving on the left	6/5 N5 OU	∞ OU	12 OU	Temporal wedge OS	Wedge to blind spot OS	44 OS 72 OD	2 years
4	67 M	Left visual disturbance	6/6 N6 OU	∞ OU	10 OU	Temporal wedge OS	Wedge to blind spot OS	54 OS 74 OD	3 years
5	43 F	Sudden visual loss in the right eye	6/5 N4 OU	−1.00 D OU	14 OU	Not available	Wedge not including blind spot OD	76 OS 59 OD	5 years
6	38 F	Right facial paresis and became aware of visual field loss	6/6 N6 OU	+1.00 D OU	15 OU	Temporal wedge OD	Wedge to blind spot OD	61 OS 32 OD	2 years
7**	41 F	Aware of bilateral “blinkers”	6/4 N5 OU	−6.00 D OU	12 OU	Not available	Wedge to blind spot OU	56 OS	1 year
8	52 F	Noticed peripheral defect in right eye	6/6 N5 OU	−10.00 D OU	14 OU	Temporal wedge OD	Not available	Not available	1 year
9	42 F	Aware of left peripheral changes after headache	6/6 N6 OU	−2.00 D OU	17 OU	Not available	Wedge not including blind spot OS	Not available	1.5 years
10***	24 F	Peripheral defect after an acute diarrhoeal illness	6/4 OD	∞ OU	Not available	Not available	Inferior Temporal	Not available	8 months
11	59 M	Asymptomatic, found on interval optometrist screening	6/5 OS 6/6 OU	−4.00 D OU	17 OU	Temporal wedge OS	Wedge OS Wedge to blind spot OS	53 OD	1.5 years
12	51 M	Asymptomatic, found on interval optometrist screening	6/5 OU	−5.00 D OU	12 OD	Temporal wedge OS	Wedge to blind spot OS	38 OS Not available	1 year
13	43 F	Asymptomatic, found on optometrist screening	6/6 OU	−2.00 D OD −2.75 OS	10 OS 13 OD 12 OS	Temporal wedge OU	Wedge to blind spot OU	Not available	6 months

*Figure 1; **Figure 2; ***Figure 3.

On examination in our service, 4 months later, the best-corrected visual acuity (BCVA) was 6/9 N5 OD and 6/9 N8 OS with full colour vision using the Ishihara plates. There was no relative afferent pupillary defect (RAPD). The intraocular pressure (IOP) was 13 mm Hg OU. Automated Humphrey visual field (HVF) testing revealed a right temporal wedge defect. Goldmann perimetry showed that the scotoma extended from 5 degrees temporal to the blind spot. The optic discs were noted to be small and crowded bilaterally. There was no disc oedema, pallor, or focal neuro-retinal rim thinning. There was no posterior vitreous detachment.

Magnetic resonance imaging (MRI) of the anterior visual pathways was normal. B-scan ultrasonography did not reveal any drusen. Results of electrodiagnostic testing (EDT), including visual evoked potentials (VEPs), full-field electroretinogram (ERG), and flash and pattern ERG, were normal.

Yearly visual field tests remained unchanged for 5 years, after which she noticed a change in vision in her left eye. Formal testing showed a new left-sided temporal wedge defect. The ocular examination was normal. Repeat MRI of brain and orbits with gadolinium did not show any abnormalities. Optical coherence tomography (OCT) showed nasal thinning of the retinal nerve fibre layer (RNFL) in both eyes. There was no evidence of glaucoma on formal assessment by a sub-specialist.

Case 2

A 56-year-old woman presented when her husband noticed that she did not see him waving on her left hand side. She had had a minor head injury to her occiput without loss of consciousness 1 month previously. She had a diagnosis of Ehlers-Danlos syndrome and idiopathic peripheral axonal neuropathy. In recent years, she had had acephalgic migraines with visual aura but these were not in the area of vision loss.

On examination, her BCVA was 6/5 N6 OU with full Ishihara colour vision and no RAPD. A temporal wedge defect extending from the blind spot was demonstrated on HVF and Goldmann perimetry (Figure 1). IOP recordings were asymmetrical: 23 mm Hg OS and 13 mm Hg OD,

partially explained by asymmetric central corneal thickness: 537 OS, 551 OD. There was also mild asymmetry of the optic discs, 0.6 cup-to-disc ratio OS and 0.5 OD, but no notching.

Neuro-imaging and EDT were normal. An OCT showed general thinning of the RNFL in the nasal quadrant of the left eye (35 μ m) relative to the right eye (60 μ m). In view of the borderline intraocular pressure, latanoprost was initiated as a precaution, although there were no clinical glaucomatous optic disc changes. Her clinical examination and visual field defect have remained static for over 10 years.

Case 3

A 36-year-old man presented with a sudden awareness of loss of vision on his left hand side. There was a history of paranasal sinus disease and turbinate surgery as well as asthma and occasional migraines with visual aura. These auras were typical and binocular.

On examination, the BCVA was 6/5 N5 OU, with full Ishihara colour vision, no RAPD, and IOPs of 11 mm Hg OU. His ocular examination was normal with the exception of a small remnant of glial tissue on the left optic disc. A temporal wedge defect contiguous with the blind spot was documented on visual field testing in the left eye. EDT and neuro-imaging were normal. OCT showed thinning of the nasal quadrant in the left eye relative to the right (44 vs. 63 μ m). His visual defect remained stable on review for 2 years.

Discussion

The differential diagnosis of a temporal wedge visual field defect is listed in Table 2. However, here we have described 13 cases in which extensive investigation was unable to demonstrate a unifying underlying aetiology for static, presumably acquired, wedge scotomas.

Buchanan and Hoyt described three cases of presumed congenital hypoplasia of the optic nerve head (ONH)⁴ where similar absolute, steep-edged wedge defects emanating from the blind spot were seen—bilateral in two cases. All, however, were associated with distinctive atrophic nasal disc margins, small caliber retinal arterioles,

Figure 1. Colour fundus photographs (A, B) and Goldmann (C, D) and automated Humphrey (HVF) (E, F) perimetry plots of patient number 2 (case 2).

and absent RNFL in the nasal sector alone. Similarly, Al-Obailan et al.⁵ describe a subset of supranasal ONH patients who have similar, congenital, temporal wedge defects associated with characteristic disc changes. With our patients, the anatomy of the disc tissue itself appeared unremarkable. In cases 2 and 3 above (unique in our series), the defect was not noticed by the patient, but rather a partner, raising the prospect that the onset was not acute and may even have been congenital. This is further tempered by the general phenotype in ONH reported of high myopia (greater than -5 D, SE), small discs, and a “double ring” sign (blurred nasal disc margins with a

scleral halo) expanded on in the larger case series of Ohguro.¹⁰ Although six of our patients were myopic, none demonstrated any of the other features of ONH previously described. Adult presentation of ONH is also usefully considered in a recent case report from Marsiglia et al.⁶

If congenital disc changes, such as ONH, are less likely, could a more common, acquired, cause be responsible? Although we could not find previous reports in the literature, it would appear logical that a focal anterior ischaemic optic neuropathy affecting the nasal sector of the optic disc could cause a similar scotoma. A hallmark of this process would be the appearance of disc swelling in the acute phase.⁷ Over

Figure 2. Colour fundus photographs (A, B) and kinetic perimetry (C, D) of patient number 7 (case 7).

half of the patients (1, 4, 6, 7, and 9) were seen within 10 days of symptoms onset with disc swelling absent, making this proposition less attractive.

As disc swelling was absent, another common optic neuropathy to be considered would be glaucoma. Although this tends to preferentially cause damage to the arcuate bundles, the entire cross-section of the nerve may be involved (including the nasal border).⁸ It has been noted that scotomas temporal to the blind spot may be easily overlooked on standard automated perimetry as few of these points are tested.⁹ However, these patients were being specifically monitored for such defects and none were found to have progressive defects over extensive follow-up periods. In addition, all but one were normotensive and no progressive RNFL thinning was demonstrated with OCT scanning, as would be expected in a glaucomatous scotoma.¹⁰

A direct retinal insult, rather than an RNFL or disc injury, could cause a discrete wedge scotoma. For example, a focal retinitis such as acute

idiopathic blind spot enlargement syndrome (AIBSE) or acute zonal occult outer retinopathy (AZOOR) could produce similar field defects. Patients are typically myopic young women who present with a peripheral scotoma and positive visual symptoms. On EDT, there is a consistent pattern of dysfunction both at the photoreceptor/retinal pigment epithelial complex but also at inner retinal levels, comprising a delayed 30 Hz flicker ERG and a reduction in the electrooculogram (EOG) light rise.¹¹ Few of our patients lie within the AZOOR demographic, together with the inter-patient consistency in the field defect appearance and the absence of the characteristic EDT findings,¹¹ would also count against this possibility. In addition, further investigation with MR imaging, though negative in all of our cases, would be indicated where there was suggestion of bilaterality or progression: features not seen in our cases.

Although direct retinal insult appears less probable, reports from patients undergoing

Figure 3. Colour fundus photographs (A, B) and kinetic perimetry (C, D) of patient number 10 (case 10).

Table 2. Differential diagnoses of temporal wedge defect.

Nasal optic nerve hypoplasia (NOH)
Glaucomatous
Anterior ischaemic optic neuropathy (AION)
Focal retinitis, e.g.,
Acute zonal occult outer retinopathy (AZOOR) or
Acute idiopathic blind spot enlargement syndrome (AIBSE)
Posterior vitreous detachment (PVD)
Early asymmetric chiasmal lesion

surgery involving the vitreoretinal interface may prove useful: non-progressive, wedge defects have reported after retinal surgery,^{12,13} Interestingly, wedge defects have been reported consistently for some decades as occurring in between 1% and 17% of patients undergoing macular hole surgery.^{12,14} Gass et al. discussed possible aetiologies, including direct mechanical trauma to the peripapillary retina or disc; direct toxic effects of the intraocular gas; retinal phototoxicity; intra-procedure ocular hypertension causing direct glaucomatous damage; ischaemia;

and dehydration injury to the RNFL.¹² Each of these theories is reinforced by the observation that techniques to limit intraocular pressure spikes, surgical time, and retinal dehydration¹⁵ have led to lower rates of such complications.¹²

Although no surgery has taken place in our patients, an interesting recent theory is relevant to our group: that shearing damage may occur during the peeling of the posterior vitreous from the point where it is tightly adherent to the disc and RNFL (usually at the nasal margin)¹⁶ as is often required in such surgery.¹⁴ Post-mortem¹⁷ and OCT¹⁸ studies suggest that congenital remnant vitreous glial condensations may be present in around 30% of eyes at the vitreous interface at the nasal margin of the disc, increasing the likelihood of tractional damage to the nasal disc (and temporal field loss) during induced vitreous detachment.¹⁹

It is conceivable that a similar insult could occur during a spontaneous posterior vitreous detachment (PVD), and this would certainly give an

acute onset of symptoms with a stationary scotoma. This concept is particularly topical given the recent proposal by Parsa and Hoyt, of a novel mechanism for non-arteritic ischaemic optic neuropathy non-arteritic anterior ischaemic optic neuropathy (NAION) where vitreous traction during PVD is said to induce axonal damage leading to symptoms, rather than ischaemia per se.²⁰ Interestingly, Katz and Hoyt have also previously reported four cases of vitreo-papillary traction in the context of incomplete PVD. Three of these cases had a scotoma. In two it was arcuate and in one an enlarged blind spot.²¹ However, sectoral disc swelling was evident acutely in all of their cases, presumably due to direct elevation by the vitreous traction. Again, this was not evident in any of our cases (although case 3 was noted to have a glial remnant at the nasal disc), nor were there concurrent symptoms of an acute PVD (photopsia, new floaters). This, together with the presence of several young and hyperopic patients (who are very unlikely to have suffered acute PVD)¹⁹ in our cohort, makes this a less attractive, if conceptually interesting, theory in our patient group.

It would appear that both the demographic and clinical characteristics of this cohort of patients, albeit with a remarkably similar presentation and clinical findings, make them particularly difficult to describe in terms of a recognised syndrome or disease. Having considered diseases of the vitreous, retina, RNFL, and optic nerve, we surmise that the likely insult is acquired and arises at the nasal aspect of the optic disc. The exact nature of this injury, however, remains to be fully elucidated, necessitating the—hopefully transitory—use of that most unedifying of terms: idiopathic!

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Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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