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The visual system and primary care optometry - encounters with the extraordinary

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Abstract
This paper will examine three primary care presentations of disease at different levels of the basic visual pathway encountered over the last year at OPSM Wollongong. This approach will serve to engage the reader with both revision of the nature of vision and an open ended evidence based approach to optometry practice and education. Professional educational development is fundamental to our role as health professionals and this paper’s collaborative effort briefly explores and expresses the group dynamic as a team of optometrists approach this responsibility. The authors hope that this thematic group based approach isrepeatable in other teams of primary care clinicians. We acknowledge and commend the work involved with producing Illuminate over the last two years and hope that this legacy of accessible quality education and research can be built upon in future editions as the profession continues to develop a culture of primary care led investigation.

Keywords
Visual system, pituitary tumour, optic neuritis, Multiple Sclerosis, arterovenous malformations, automated visual field examination.

Disciplines
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The Visual System and Primary Care Optometry
- Encounters with the Extraordinary

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Tom has been working at OPSM Wollongong in some capacity since graduation in 2000 and began as a graduate with the then licensee Anthony Roccon. Tom is currently a part-time postgraduate research student of the University of Wollongong he is also a Clinical Academic of the Graduate School of Medicine and an Honorary Fellow of the CHSD at University of Wollongong. Tom and his wife Sonia are expecting their third child (any day now)

Melissa has been working with the wonderful team in Wollongong since graduating in optometry from UNSW. She worked at OPSM for over year, and has recently transferred to Budget Eyewear, just down the road from OPSM. She very much enjoys the varied working environments of OPSM and Budget Eyewear, the best of both worlds!

Binita is currently completing her first year as a graduate Optometrist at OPSM in Wollongong.

Nuong has been a primary eye care practitioner for 16 years. Nuong had her own practice for 6 years, had a travel break and now fully enjoying Optometry as a part timer. Nuong has been with the OPSM team for three years.

Purpose

This paper will examine three primary care presentations of disease at different levels of the basic visual pathway encountered over the last year at OPSM Wollongong.

This approach will serve to engage the reader with both revision of the nature of vision and an open ended evidence based approach to optometry practice and education. Professional educational development is fundamental to our role as health professionals and this paper’s collaborative effort briefly explores and expresses the group dynamic as a team of optometrists in this dynamic primary care optometry setting. The authors hope that this thematic group based approach is repeatable in other teams of primary care that optometry is about vision and that vision clinicians. We acknowledge and commend the is a perception, a type of thought, that can work involved with producing Illuminate over be affected by almost anything that people the last two years and hope that this legacy of experience. But this in itself is not a fault; even accessible quality education and research can be built upon in future editions as the profession continues to develop a culture of primary care led investigation.

Introduction

One of the benefits of working in a group practice is that there is usually another optometrist nearby to discuss the extraordinary presentations. Of course the complex nature of the visual system virtually ensures that with peculiar regularity people present with rare cases of visual effect due to explainable and sometimes unexplainable phenomena.

Our ability to communicate with the patient, colleagues and other health professionals tested this responsibility. The authors hope that may be upset, confused and rarely understand approach this responsibility. The authors hope that this thematic group based approach is repeatable in other teams of primary care that optometry is about vision and that vision clinicians. We acknowledge and commend the is a perception, a type of thought, that can work involved with producing Illuminate over be affected by almost anything that people the last two years and hope that this legacy of experience. But this in itself is not a fault; even accessible quality education and research can be built upon in future editions as the profession continues to develop a culture of primary care led investigation.

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In primary care optometry we never have time to perform every appropriate clinical test and must prioritise the questions that we ask of ourselves and the patient. We must also be very selective with the information that we share with colleagues when collaborating and discussing differential diagnosis and management options. There are times when a photo from a retinal camera or a print out from a visual field analyser can help capture part of the clinical puzzle in a way that is immediately obvious and recognisable for diagnostic purposes and very easily communicable. These tests and their results do not replace but instead enhance the human aspects of primary health care. They represent and justify the patient’s experience and help optometrists to advocate for the best outcome possible from an increasingly complex and stressed medical system.

The cases that are discussed in this paper are representative of the extraordinary encounters that occur in everyday primary care practice. We have focused on instances that highlight the usefulness of our brand new toys; the non-mydriatic retinal camera and automated visual field analyser. The Cases that have been chosen are: optic neuritis; chiasmic compression; and localised interruption of primary visual cortex.

Thus we move through the different parts of the basic elements of the visual pathway from the eyes to the primary visual cortex. (See Figure 1.0)

**Case One;**
**Optic neuritis episode as Primary presentation of Multiple Sclerosis**

**History**
KM, a 25 year old Caucasian female presented to our practice with a one week history of blurred vision in the right eye. She also noted a dull ache in the right peri-orbital region accompanied by pain on eye movement, more from medial to lateral gaze and vice versa. The patient reported no other visual or neurological symptoms. Her general health is excellent, and she did not report taking any regular medications. Her previous eye test was 5 months ago, where she was fitted for the first time with Activize A monthly contact lenses for correction of mild myopia. Her best corrected acuity at that time was 6/6 (OD), 6/6 (OS). She reports using these contact lenses once a week, and finds no problem with them. As part of a best practice approach to use of retinal camera fundus photos were taken at the time and were normal (see fig 1.1,1.2).

**Case Two**
**Pituitary Tumour**

**Case Three**
**Interruption of Primary visual cortex function.**

Figure 1.0 Interruption of the visual pathway (Adapted from Silverthorn, 2007)
Her visual field results showed a generalized depression of the entire central field for the right eye, the left eye was normal for her age.

**Clinical findings:**
Though KM reported vision blur in the right eye there was no significant change in refraction. her best corrected spectacle acuity was:

**OD:** -1.25/-0.25 x 100 6/7.5  
(No improvement with pinhole)  
**OS:** -1.25/-0.25 x 035 6/6

Her pupil reactions were normal, and did no appear to have an obvious relative afferent pupillary defect. She did note reduced colour perception in the right eye compared to left on ishihara colour vision testing. Slit lamp examination of her anterior segments was unremarkable. Examination of the posterior segment of the eyes showed slight congestion and swelling of the optic nerve head in the right eye (pic 1.3); the left eye appeared normal. Her intraocular pressures were 15mmHg for the right eye, and 16mmHg for the left eye.

Thus the change in appearance of the right optic nerve head was confirmed when compared to the retinal photo record from the previous appointment.
Management:
A diagnosis of optic neuritis was made on the basis of the history and symptoms and confirmed by the clinical findings. While the advanced diagnostic tests were being performed other optometrists in the practice were calling to arrange a prompt referral that afternoon to a local ophthalmologist who was known to have connections to Sydney based specialists in the central hospital system.

KM was then referred on by the ophthalmologist to a consultant neurologist in a major Sydney hospital for further investigations to examine the strong association between multiple sclerosis and optic neuritis. These tests included an MRI and evoked cortical potentials.

Her MRI scan showed a significant number of small lesions with typical morphology and distribution for multiple sclerosis. Lesions were noted in both the corpus callosum and in a pericallosal distribution, and it was thus concluded that she does indeed have multiple sclerosis. The evoked potentials showed evidence of impaired conduction in both optic nerves, worse on the right; and impaired central sensory conduction in the left spinal cord. She was advised that there was at least an 80% chance of developing further clinical attacks over the next ten years.

She was reviewed by the neurologist two months later, and reported suffering from a number of minor intermittent symptoms of MS, including lower limb paraesthesiae after walking for fifteen minutes or more, and generally a sense of feeling ‘washed out’ after a hot shower or bath and a number of headaches.

Discussion:
Optic neuritis is defined as an acute inflammation of the optic nerve (Baker, 2006). While etiologies include infection(syphilis, mumps, measles), infiltrative, inflammatory disease (sarcoidosis, lupus), ischaemic vascular disease (diabetes), the most common etiology is the demyelinating disease multiple sclerosis (MS) (Beck 1997, Landau 1998, Trobe 2003). There is always some permanent damage to the optic nerve with a loss of myelin around the nerve axons at the site of inflammation and is therefore said to be a demyelinating condition.

Acute demyelinating optic neuritis is a clinical diagnosis that can often be made on the basis of the history and clinical features. Typical features include: Pain in or around the eye exacerbated by eye movements (present in >90% of patients), unilateral vision loss, usually progressing over a period of hours to days and central swelling of the optic disc (present in one third of patients).

Patients with optic neuritis who have a normal optic nerve appearance are said to have a retrobulbar optic neuritis. Often, there is an impairment of colour perception, especially red, and a relative afferent pupillary defect (Balcer 2006, Chhabra 2003). Visual field loss is typical of acute demyelinating optic neuritis. Classic defects include central scotomas, however, a more broad spectrum of patterns typically occurs. The most common being a diffuse depression of sensitivity in the entire central 30 degrees of the visual field (Balcer 2006, Fang 1999, Kanski 2007).

The majority of patients are female (F:M 3:1) and are usually aged from 20 – 50. Systemic signs and symptoms may include headache, nausea, Uhtoff’s sign (decreased vision with or without limb weakness following exposure to increased temperatures i.e, a bath or exercise), Romberg’s sign (patient falls when they close their eyes), Pulfrich’s stereo phenomenon and fever.

Association with Multiple sclerosis:
Although some patients with optic neuritis have no clinically demonstrable associated systemic disease, the following close association exists between optic neuritis and MS.

Acute demyelinating optic neuritis is the presenting feature in 15 to 20 % of patients with multiple sclerosis, and it occurs in at some time during the course of the disease in 50% of patients (Arnold 2005, Frohman 2005, Foroozan 2002). During a 10 year follow-up, in patients enrolled for the Optic Neuritis Treatment Trial (ONTT), there was a 30% risk of MS and a 38% risk at 10 years. It was also found that an MRI scan was performed at the time of the original acute optic neuritis was helpful in predicting the risk of MS. Even a single white matter lesion on the MRI scan increased the 10 year risk to 56% (Chhabra 2003).

Treatment
The ONTT supports the administration of intravenous methylprednisolone, which hastens recovery of visual function but does not affect long term visual outcome. Studies have also shown that interferon therapy has been shown to reduce the development of multiple sclerosis
in patients with acute demyelinating optic neuritis and two or more myelinating lesions on MRI on the brain. Our patient did not require steroids for her first episode, as there was spontaneous resolution of her optic neuritis; however, the neurologist had recommended that she would benefit from an early introduction of immunomodulatory therapy. Increasing evidence suggests that this therapy should be started at the first presentation or soon afterwards in patients with typical MR imaging. Our patient is currently undergoing IVF therapy, hence has deferred treatment of MS at this stage.

**Clinical Findings**

Initial visual acuity through her habitual prescription was relatively poor:

- **RE**: -1.50  6/18
- **LE**: -1.75/-0.25x80  6/24

During a brief refraction her visual acuity corrected to:

- **RE**: -2.25  6/7
- **LE**: -2.75  6/9

Where the left eye was not able to be corrected beyond 6/9 it was difficult to judge whether this was due to a change in visual acuity as this was the first appointment at this practice this may have existed previously.

Due to time constraints Ishihara and other test of optic nerve function, besides the VF test, were not performed at this point.

Slit lamp examination and undilated fundus examination with a Volk Super field lens showed no major abnormalities. The Optic nerve C/D ratio was recorded as 0.4 in both eyes and both optic nerves were small but did not appear raised, though the left was slightly different colour. The slight asymmetry of optic nerve head appearance was confirmed with retinal photography(Fig 2.1 and 2.2)

**Case Two;**

**Visual status of Lady with recently discovered Pituitary Tumour**

**History**

WP, a 54-year-old female office worker, presented for assessment of visual field at our optometrical practice as she had been directed to have this test performed before her appointment with a local neurologist the following morning.

It was the first time that she had had her eyes tested for some time and the first time that she had been examined at this practice. She has moderate myopia and besides the increase in blurry vision from what she presumed was due to an increase in her short sightedness she was not aware of any change in her vision.

The lady reported that she had recently been made aware of a pituitary tumour after a routine blood test showed that her cortisone levels were very low and a subsequent scan revealed a 3cm pituitary tumour. She had no headache or other signs of brain tumour besides some ongoing fatigue after a virus that had prompted the blood test with the GP.

She had commenced some cortisone treatment with an endocrinologist and explained she was quite upset since she had thought that she was just having trouble getting over a virus and instead had been given new news that she had a brain tumour.
A slight but detectable visual field defect was noted at this stage. Figure 2.5 and 2.6

Visual fields were tested with a 24-2 fast threshold test in both eyes. Figure 2.3 and 2.4

Management
Understandably this lady was reasonably upset from the moment that she arrived at the practice to make the appointment for the visual field test as directed by her local neurologist.

She was under the impression that she could have the test done almost immediately and that the test gave almost instant results; this made coherent and comprehensive management difficult. After some initial clarification that to conduct a visual field test correctly we needed to know more about her vision she did give a reasonably lucid history but given her extraordinary story this took most of her half hour appointment slot. After a brief refraction, fundoscopy and retinal photography, by the time she finished the visual field test she had well and truly had enough. As soon as the results were
compiled and a reporting letter hastily written she disappeared, promising to return.

Some three weeks later her GP was tracked down (through the local division of General Practice) and a courtesy call was made which confirmed that the lady was seeing a neurosurgeon that specialised in pituitary tumour treatment and that the results of her optometrical examination had been passed on. Under the circumstances, with such fleeting contact with the patient, this is a surprisingly reassuring outcome and with the controversy in the literature that surrounds management of acute cases of pituitary apoplexy and the like it is important that the best available neurosurgical opinion is sought (Gruber A, 2006).

Further to the invitation in the first issue of Illuminate; writing up unusual clinical encounters such as this case for peer review is also an integral part of a practice’s clinical management and a sign of professional care and diligence (Gardner et al., 2006). Although optometrical management is theoretically ongoing, at this stage where little else can be done to affect the outcome of this case, until medical and surgical treatment options are explored, at the very least it is hoped that someone else may learn from this presentation.

**Discussion**

The Pituitary gland is situated slightly below the optic chiasm at the base of the brain. A pituitary tumour has systemic health implications and a well characterised effect on vision: the bitemporal field defect measured is usually deepest in the superior quadrants (Kanski, 2001: p 128). This type of tumour can have a very insidious onset and thus the visual effect can be (initially) quite subtle which is demonstrated well by this case study.

The tendency for pituitary adenomas to be slow growing means that the victims are often unaware of the slight changes to their vision and the imperilment of their general health until it is quite late. The peripheral vision effect in this case was too small for WP to be aware of, and the central visual effect on the left eye, which may be due to asymmetrical impact of the tumour, was masked by her poor refractive status (for Chiasmic disease; Kanski, 2007: pp 807-817).

There is such an overwhelming impact on the endocrine system with pituitary disease that seen often in diabetics with poor blood glucose level (BGL) control (Travis, 2006). The risk of diabetes insipidus is also acknowledged and thus refractive status will be monitored closely for fluctuations in future examination of the patient (Gruber A, 2006).

In cases of pituitary apoplexy the risk to vision is a central concern driving decisions about treatment. Understandably the effect on general health is also of prime concern, for example, as Chanson et al points out: “Corticotropic deficiency (secondary adrenal failure) may be life-threatening if untreated” (2004:p1287).

As patients with pituitary tumours, whether microadenoma (<10mm, less likely to effect vision), macroadenoma or post de-compressive surgery, are usually recommended to have regular visual field testing it is not uncommon for optometrists to find themselves providing this sort of examination facility (Collin, 1993).

This assessment of vision by the optometrist does not preclude or replace an ophthalmologist consultation and the neurologist may seek more specific ophthalmological advice (Alexander Poon, 1995). In the circumstances the primary care role of the optometrist proved more expedient for the purposes of the neurologist, other specialists and the patient; this utilisation of primary care resources is an especially common occurrence in rural and regional areas where access to specialist medical advice (particularly ophthalmologists) is difficult to obtain quickly.

Primary care Optometrists also often recommend visual field testing where unusual headaches, suspicious gross anatomical features, elevated IOP and more commonly, optic nerve appearance give an indication of possible peripheral vision defect (Mok, 2008). The nature of the visual field defect can often be traced back to a possible cause by taking into account the anatomy of the visual system.
Case Three:
An Unusual Case of Visual Disturbance, Transient episodes of Hemianopia due to Arteriovenous malformation of the Brain

History
A 36-year-old male, JW, presented complaining of visual disturbances. The disturbance would begin as a red balloon shape moving in from his right side and blocking his vision. The ‘balloon’ would then explode and become like multi-coloured confetti pieces that formed into lines and then completely disappear. This had been occurring three to six times a day for the past two weeks.

JW had a history of arteriovenous malformation (AVM) of the brain and was under the care of a neurologist. He had had radiosurgery treatment for the AVM approximately one and a half years ago. No other general health problems were noted and he was not taking any medications.

He had a visual fields test one year ago and the results were normal for his age.

Clinical Findings
Routine refraction and undilated slit lamp examination findings were unremarkable:

Uncorrected VA: RE: 6/6 LE: 6/6
IOP: RE: 15 LE: 13
Ishihara: RE: 2 errors LE: 2 errors
Anterior eye: NAD OU
Posterior eye: NAD OU

Retinal photos were taken and were also unremarkable.

Visual fields testing revealed an almost perfect right homonymous hemianopia with macula sparing. The patient noticed the same visual disturbances during the test. Figs 3.1 and 3.2.

Management
The patient was referred to their GP for a same day urgent referral to their neurologist. However compliance was poor. When the patient was called a few hours later they needed to be reminded about the urgency of the situation as they were going to wait until their GP returned from holidays in a week’s time. The optometrist then called JW’s neurologist on his behalf and a prompt appointment with their neurologist was arranged and when he saw them the next day he diagnosed the patient as having multiple seizures. He also found that the patient was
experiencing tingling in his right arm in association with his visual episodes. Medication was prescribed, and further diagnostic investigations are continuing. Interestingly the neurologist advised that the visual field defect was temporary and could change corresponding to activity in the brain.

**Discussion**

Arteriovenous malformations (AVMs) are complex tangles of thin-walled blood vessels that can haemorrhage more easily than normal, healthy vessels (CINN, 2007). They are a problem because they do not give the brain a chance to absorb oxygen from the blood, and can therefore result in strokes. Stereotactic radiosurgery causes the AVM vessels to slowly clot off from one to three years after the treatment until the AVM is obliterated. (Mayo Clinic, 2008)

While AVM is a rare condition unlikely to be encountered in everyday practice, the presenting symptoms have differential diagnoses that range from innocuous to life threatening. One thing worth noting is that the patient did not mention his previous history of AVM until after the visual fields test was performed and a suggestion of neurological testing was made. Based on symptoms and routine testing, one might have considered this an unusual case of acephalic migraine (migraine sine migraine), or alternatively a transient ischemic attack (TIA) and impending stroke (CVA).

The right homonymous field defect indicates that the electrical activity of the seizures originated from behind the optic chiasm, more specifically in the left primary visual cortex.

If visual fields testing had not been performed, AVM might not have even been mentioned. This highlights the fact that optometry is still viewed as a trade by some of the public, as opposed to a health profession. Perhaps we need to speak up more to our patients about what exactly we’re doing when we’re dazzling them with our bright lights...

**CONCLUSIONS:**

*Primary Vision-care Encounters with the Extraordinary*

The fascination of everyday clinical encounters in optometry is something that is difficult to appreciate much of the time. We all remember that first time we saw the hidden Mars like terrain of the retina when learning about fundoscopy in University, but it is hard to maintain the same enthusiasm for every retina that we examine (also, generally people get worried quite quickly if you say “Wow, look at that!” when looking at the back part of their eyes).

This paper has used cases that explore some of the possibilities that lay just beyond, or on the edge of what we can know of the primary visual pathway even at its most basic level. For example it is at times difficult to understand how a subtle effect on vision from optic neuritis can prompt a patient (KM from Case 1) to seek an examination yet a large and recurring effect on peripheral vision (JW, case 3) can go unnoticed and leave a person unworried for so long. It does not matter that these particular cases are so unique and rare that an identical case may never be encountered by a colleague. Conversely what this article attempts to highlight is that every patient is extraordinary; even and especially the immaculately predictable 40-year-old early presbyope having the first eye examination they can remember and realising that they are getting old, mortal, on the verge of a midlife crisis; all while we are conducting our examination.

The collaborative nature of developing this article, while being an excellent exercise in developing an appreciation for the depth of knowledge, care and skill of colleagues, has also drawn attention to the relatively predictable experience of encountering truly rare and unique clinical presentations in primary care. The professional resources of optometry are well recognised by the mainstream health community and the general public. To continue to meet the challenges of the future we need to be open to new technology, evolving clinical standards and ever mindful of old fashioned care and comfort that can only be given by a fellow human being.

The cases presented in this article all underline how well practices can use their resources to make a difference to people’s quality of life; no matter how mundane or bizarre their particular life experience may be.
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