CASE REPORT

Early Loss of Optic Cup with Increased Intracranial Pressure

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ABSTRACT

A 54-year-old woman developed increased intracranial pressure due to chronic dural venous sinus thrombosis in the absence of significant comorbidity. In the years preceding diagnosis, the patient underwent routine diabetic retinopathy screening; however, gradual loss of optic cup and disc swelling was only retrospectively noted after marked papilloedema was evident at an optometry visit. The patient made a full recovery once the diagnosis was established. This case is novel in the literature as routine photographs, taken for diabetic retinopathy screening, demonstrate the natural history of papilloedema before medical intervention. Interestingly, these images show that a traditionally “late” sign of papilloedema – loss of cup – can be the sentinel abnormality. This finding may have significant clinical implications and reinforces the need for clinicians to compare investigation results with historical patient data.

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KEYWORDS

Optic cup; papilloedema;
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Introduction

This is a case of a 54-year-old female who presented asymptotically to her optometrist for routine diabetic (DM) screening. Upon questioning she admitted to a one-month history of headaches that worsened when bending forwards, with occasional transient visual loss and vomiting. On examination, bilateral papilloedema was noted and she was urgently referred for ophthalmology review. Disc swelling was confirmed in the absence of any other new ocular pathology. Neurological examination revealed symmetrically brisk upper limb reflexes without any other appreciable abnormality. No relative afferent pupillary defect (RAPD), colour vision deficit or new ocular motility problem was identified. Malignant hypertension was excluded and no suggestion of an infective or toxic cause was elicited in the history. The patient was not obese.

Lumbar puncture opening pressure was elevated (39.5 cm water) and magnetic resonance venography (MRV) revealed dural sinus thrombosis at the junction of the transverse and sigmoid sinuses with collateral vessel formation (Figure 1). Given the presence of collateral vessels and minimal acute symptoms, the process was considered chronic.

It was upon retrospective review of clinical images, taken to screen for a separate condition, that the evolution of relatively indolent disc swelling was identified.

Over the course of approximately 5 years preceding the diagnosed papilloedema, the patient was reviewed at regular intervals with serial fundus photography for diabetic retinopathy screening. In each instance, the patient was clinically stable with a superficially healthy appearance of the optic disc.

At no point was hypotony,¹ decreased colour vision or RAPD found, despite serial testing as part of routine practice. Her refraction (+1.75 diopters) and visual acuities (6/5 best corrected, both eyes) remained stable throughout.

The disc images in this series are cropped sections of screening fundal photographs. Figures 1 and 2 in the series are largely unremarkable in isolation although demonstrate a decreasing cup-to-disc ratio (CDR) in both eyes. With the benefit of hindsight, Figure 2 may show subtle swelling [Modified Frisén Scale (MFS) 1²], more noticeable in the left eye. Figure 3 reveals early swelling (MFS 2) with further loss of cup that was initially missed while screening for diabetic retinopathy. Figure 4 shows

Figure 1. Bilateral disc images (cropped) – December 2007 (Baseline).

Figure 2. Bilateral disc images (cropped) – March 2009 (Decreased CDR. Possible MFS grade 1 disc swelling left eye).

Figure 3. Bilateral disc images (cropped) – February 2010 (Decreased CDR. MFS grade 2 disc swelling left eye).

marked papilloedema (MFS 4/5) which prompted the referral to ophthalmology.

In the absence of detectable thrombophilia or significant comorbidity, the patient was not anticoagulated for this chronic process and the condition spontaneously resolved. Upon review 4 years later, mild disc swelling was persistent

([Figure 5](#)) however the patient had no symptoms or functional impairment.

Discussion

We believe this image series represents the first published report of papilloedema evolving in the

Figure 4. Bilateral disc images (cropped) – July 2012 (Bilateral MFS grade 4/5 disc swelling).

Figure 5. Bilateral disc images (cropped) – June 2016 (Bilateral MFS grade 1/2 disc swelling).

context of chronic venous sinus thrombosis. These images, capturing a disease in development, provide a rare insight into the progression of disc swelling before medical intervention altered the natural history.

Comparison of this real-world series with the theoretical Frisén system demonstrates some interesting differences. Notably, the widely used grading system (0–5 scale) lists loss of cup as a late sign seen in grades 4 and 5.^{2,3} This system has been challenged in recent literature,⁴ and a decreased cup-to-disc ratio (CDR) has been demonstrated in the idiopathic intracranial hypertension population across papilloedema severity grades.^{5,6} We believe this is the first case report documenting loss of optic cup as the initial change on fundus photography in a patient with venous sinus thrombosis.

In this case no harm befell the patient as she went on to make a full symptomatic and functional recovery. The report serves to illustrate the importance of

referring to historical data in clinical practice. We believe that this is the first reported case with documented evidence of progressive optic cup loss over several years predating a diagnosis of elevated intracranial pressure and venous sinus thrombosis – and as such is an important addition to the world literature. With this in mind, clinicians must refer to previous results and remain vigilant for dangerous progression in superficially healthy discs.

Abbreviations

CDR – cup-to-disc ratio
DM – diabetes mellitus
MRV – magnetic resonance venography
RAPD – relative afferent pupillary defect

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