Posterior Globe Flattening without Papilloedema in Idiopathic Intracranial Hypertension

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ABSTRACT

A 64-year-old woman had a one-year history of transient visual obscurations in the left eye and was found to have left optic disc oedema with preserved visual function. She was diagnosed with unilateral papilloedema related to idiopathic intracranial hypertension. Magnetic resonance imaging of the orbits showed flattening of the globe in the right eye, which did not have disc oedema. Flattening of the posterior globe without papilloedema suggests that the barrier to transmission of cerebrospinal fluid pressure to optic nerve head is at the level of the lamina cribrosa and may be due to connective tissue changes in this area.

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Papilloedema; idiopathic intracranial hypertension; magnetic resonance imaging; raised intracranial pressure

A 64-year-old obese woman presented with a 12-month history of transient visual obscurations in the left eye only. She had mild daily holocephalic headaches, but no pulsatile tinnitus or diplopia. Optic disc oedema in the left eye was noted at the time of symptom onset by multiple ophthalmologists. On examination, visual acuity was 20/20 in both eyes and there was no relative afferent pupillary defect. Intraocular pressure was 10 mmHg in the right eye and 11 mmHg in the left eye. Humphrey visual fields were normal in both eyes. Dilated fundus examination revealed a normal appearing right optic disc and left optic disc oedema with a large peripapillary subretinal haemorrhage (Figure 1). Intravenous fluorescein angiography of the optic nerves was normal in the right eye and demonstrated leakage from the left optic disc. Optical coherence tomography demonstrated a normal average retinal nerve fibre layer thickness of 86 μm in the right eye and was elevated with an average thickness of 149 μm in the left eye. Magnetic resonance (MR) imaging of the orbits revealed flattening of the posterior globe with mild dilatation of the peri-optic cerebrospinal fluid (CSF) space in the right eye and to a lesser extent in the left eye (Figure 2). MR

imaging of the brain and MR venography showed an empty sella and distal transverse sinus stenosis, respectively. MR angiography of the brain and MR imaging of the spine were normal. A lumbar puncture in the left lateral decubitus position revealed an opening pressure of 37 cmCSF and resulted in resolution of her transient visual obscurations. She was diagnosed with unilateral papilloedema related to idiopathic intracranial hypertension (IIH) and treated with weight loss and acetazolamide. Her left optic disc oedema resolved after six months.

Papilloedema in IIH is usually symmetrical, but can be asymmetrical or unilateral. This patient had unilateral papilloedema, but flattening of the globe evident on MR imaging in the eye without papilloedema. Unilateral papilloedema is thought to be related to anatomical variations, which preclude transmission of CSF pressure to the optic nerve head in the eye without papilloedema. Various theories have been proposed including fibrous adhesions in the subarachnoid space surrounding the optic nerve and changes in the lamina cribrosa.¹ More recently, a smaller optic canal size was thought to preclude transmission of CSF pressure to the optic nerve head,² but this result was not reproduced in a subsequent

Figure 1. Colour fundus photographs demonstrating a normal appearing optic nerve in the right eye and optic disc oedema with a peripapillary subretinal haemorrhage in the left eye.

Figure 2. Axial T1 post-gadolinium (left) and T2 (right) MR imaging of the orbits demonstrating flattening of the posterior globes (solid arrows) and distention of the peri-optic CSF space in the right eye (dotted arrows).

publication.³ The observation of posterior globe flattening and dilatation of the peri-optic CSF space behind the globe without papilloedema, localises the barrier to transmission of CSF pressure to the lamina cribrosa. Indeed, a previous publication of patients with unilateral papilloedema found that they tended to be older than those with bilateral papilloedema and this observation was hypothesised to be due to age-related changes in the lamina cribrosa.¹ Previous studies have found that total collagen in the lamina cribrosa increases with age, resulting in a less elastic and flexible structure, presumably sparing the optic nerve head from CSF pressure.⁴ This older patient with unilateral papilloedema and flattening of the globe supports this theory. Further study into the mechanism of unilateral papilloedema is warranted to better understand the

connective tissue changes that occur in these patients.

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