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CASE REPORT

Fulminant Idiopathic Intracranial Hypertension With Malignant Systemic Hypertension—A Case Report

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ABSTRACT

We describe a young woman who presented with malignant systemic hypertension and fulminant idiopathic intracranial hypertension. This is a rare combination, but both diagnoses should be considered in patients with optic disc swelling in whom cerebral imaging does not suggest an alternative cause. In this case, malignant hypertension was identified and treated before the idiopathic intracranial hypertension was recognised. Visual failure was evident at presentation and prior to blood pressure manipulation. It is likely that a combination of both conditions increased the vulnerability of the optic nerve head to ischaemic damage. It is also possible that reducing blood pressure in such patients, without treating coexisting raised intracranial pressure, may compound an already compromised ciliary arterial perfusion pressure. We therefore recommend careful blood pressure measurement in all patients presenting with idiopathic intracranial hypertension and advise that lumbar puncture is performed in patients with malignant hypertension with optic disc oedema, particularly in overweight young females.

Keywords: Cerebrospinal fluid, computerized tomography, idiopathic intracranial hypertension

INTRODUCTION

The aetiology of idiopathic intracranial hypertension (IIH) remains an issue of debate.[1] It usually affects obese females of child-bearing age. Diagnostic criteria include symptoms and signs that are attributable to elevated intracranial pressure with normal cerebrospinal fluid composition along with the absence of hydrocephalus, and structural or vascular lesions on brain imaging. In fulminant IIH—defined as (i) the acute onset of symptoms and signs of intracranial hypertension (less than 4 weeks between onset of initial symptoms and severe visual loss) and (ii) rapid worsening of visual loss over few days—there are reports of up to one quarter with coexistent systemic hypertension at presentation.[2] Fulminant IIH is a medical emergency and should be recognised early and managed as such. The coexistence of systemic hypertension with IIH has implications in terms of treatment and may indicate a group of patients at more risk of visual complications.

CASE REPORT

A 23-year-old female with no significant past medical history apart from a raised body mass index (BMI) of 39.5 and occasional migraine presented to her optician with a 3-day history of reduced vision in the left eye. There was also left-sided headache and horizontal binocular diplopia. Visual acuity was 6/36 in the right eye (RE) and 6/60 in the left eye (LE). Bilateral optic disc swelling and retinal haemorrhages (Figure 1) were observed and immediate referral to hospital was arranged. There she was found to be hypertensive, with blood pressures up to 220/140 mm Hg. There was proteinuria and evidence of left ventricular hypertrophy on echocardiography. A diagnosis of

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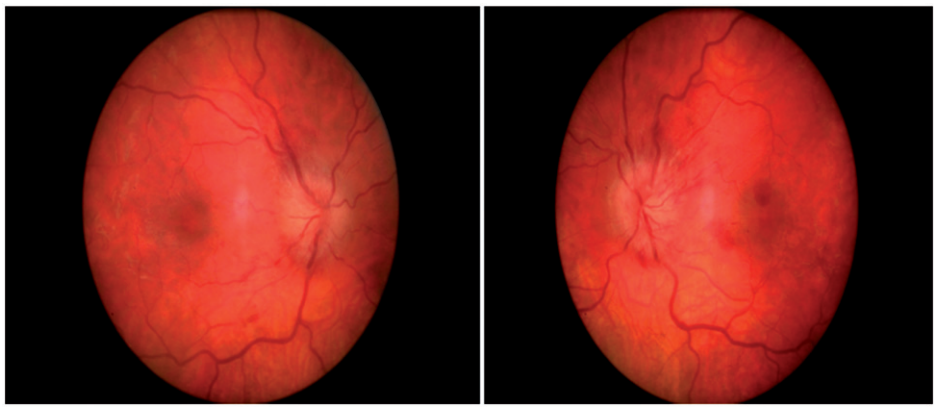

FIGURE 1 Bilateral optic disc swelling and retinal haemorrhages (***Note***: Figure 1 of this article is available in colour online at www.informahealthcare.com/oph).

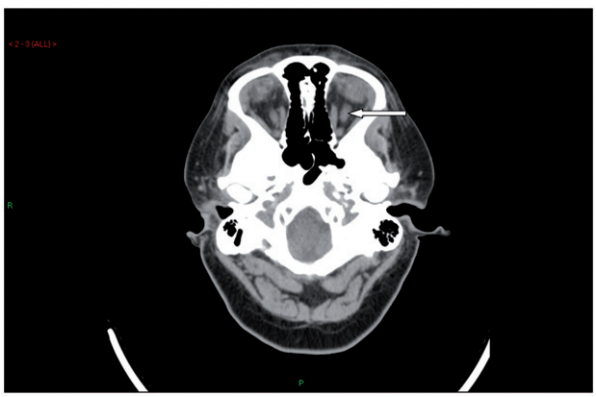

FIGURE 2 Initial CT head showing optic nerve sheath distension (arrow).

malignant hypertension was made and the optic nerve swelling was attributed to this. Ramipril, amlodipine, and doxazosin were commenced to treat the hypertension. A computed tomography (CT) brain was reported as normal, but on review of images retrospectively it showed evidence of optic nerve sheath swelling bilaterally (Figure 2). Magnetic resonance imaging (MRI) of the heart and MR angiogram of aorta and renal arteries were normal. Serum aldosterone was 1121 pmol/L (normal range: 100–400 pmol/L) and renin was raised at 95 mIU/L (normal range: up to 40 mIU/L). These results were compatible with secondary hyperaldosteronism caused by reduced renal perfusion from systemic vasoconstriction (in primary hyperaldosteronism plasma renin is suppressed). No secondary cause for her hypertension was identified.

Despite a gradual improvement in blood pressure, she continued to notice worsening of vision. She initially described difficulty seeing things in the periphery and then her vision became ''grey'' bilaterally. Soon after discharge from hospital, 13 days after her admission, she experienced a further deterioration in vision when ''everything went black.'' She was then seen by neuro-ophthalmology where her visual acuity was reduced to hand movements (RE) and perception of light only (LE). Pupils measured 7 mm bilaterally and were unreactive to light. Extensive swelling of optic discs bilaterally was noted with haemorrhages and exudates. A left sixth cranial nerve palsy was also observed. Following a repeat CT brain, a lumbar puncture was performed showing an opening pressure of 64 cm cerebrospinal fluid (CSF) (normal range: 10–20 cm CSF). The CSF protein count was 0.83 g/L (normal range up to 0.51 g/L). A CT venogram was normal.

Subsequent lumbar punctures performed over the next 2 days showed opening pressures of 24 and 16 cm CSF, respectively. MRI brain displayed resolution of optic nerve sheath dilatation. She was commenced on acetazolamide and underwent further lumbar punctures over the following 6 months. There was some visual improvement with visual acuities, 9 months later, of 6/24 RE and 6/60 LE, although she is registered blind. There is bilateral optic disc pallor. Visual field perimetry results are shown in Figure 3. Blood pressure has remained well controlled on her antihypertensive regimen.

DISCUSSION

Severe visual loss in IIH is usually a result of papilloedema and is reported in approximately 10–20% of cases, most often in longer-standing cases as a result of axonal death leading to optic atrophy.[3] A small number of patients with IIH have a fulminant course, with an early onset of visual loss. The pathogenesis of abrupt visual loss in IIH is poorly understood but may be related to axoplasmic stasis and optic nerve head ischaemia in the setting of an acute rise in intracranial pressure.[4] Axoplasmic stasis is fundamental in the pathogenesis of papilloedema and is thought to arise either from a direct compression of axons (mechanical theory) or secondary to reduced perfusion of axons (ischaemic theory).[5] In support of the ischaemic theory of visual loss in

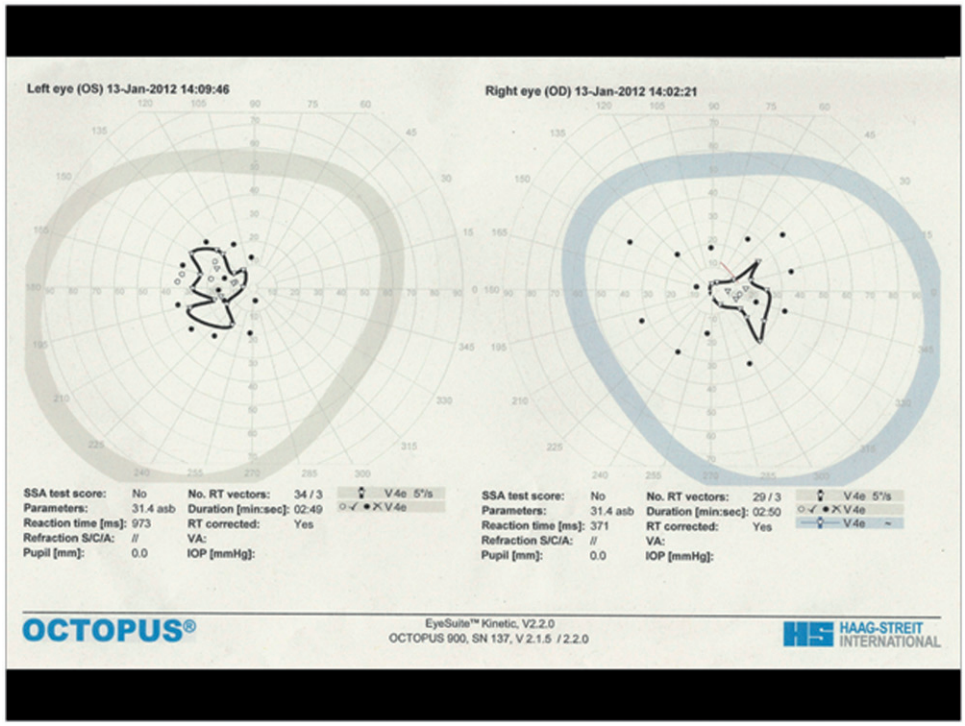

FIGURE 3 Automated visual field perimetry 10 weeks after presentation, when first able to perform test.

papilloedema are the observations that fluorescein angiography shows delayed filling of the optic disc and peripapillary choroidal vessels in papilloedema,[6] coupled with the fact that those with arteriosclerosis are especially prone to optic neuropathy in IIH.[7]

Malignant hypertension may cause hypertensive retinopathy, a choroidopathy and a hypertensive optic neuropathy. Hypertensive optic neuropathy is characterized by optic disc oedema. This has been proposed as representing a form of anterior ischaemic optic neuropathy[8] with vasoconstriction and choroidal ischaemia contributing to the disc oedema. The retinal photographs of our case do not show the extensive cotton wool spots and exudates normally expected in malignant hypertension, pointing more to papilloedema as the cause of the disc oedema.

Obesity predisposes to systemic hypertension, which therefore, unsurprisingly, is prevalent in the IIH population. Hypertension has been cited in the medical history of between 20%[9] and 32%[10] of patients with IIH, although blood pressure recordings at presentation were not given. Obesity and female sex have also been identified as risk factors for the development of hypertensive crises.[11] We propose that a combination of the two diagnoses of IIH and systemic hypertension led to an acceleration of visual loss and suggest that treating systemic hypertension without lowering raised intracranial pressure may compound a critical ischaemia at the level of the ciliary arterial circle. This circle is vulnerable to acute hypotension (and therefore perhaps a rapid decrease in blood pressure in a previously hypertensive patient), as it competes with the choroidal circulation. A change in circulation dynamics may lead to a ''choroidal steal'' phenomenon.[5] In addition, certain antihypertensive agents (for example, calcium channel antagonists and angiotensin-converting enzyme [ACE] inhibitors), whilst effectively treating systemic hypertension, may dilate the cerebral vasculature and increase cerebral blood volume, thus potentially increasing intracranial pressure.[12] In retrospect, a beta blocker may have been a wiser choice for initial antihypertensive therapy.

It is possible that the secondary elevation in serum aldosterone observed in our case may contribute to the intracranial hypertension. Intracranial hypertension has been previously reported in patients with primary hyperaldosteronism and CSF aldosterone correlates with serum levels.[13,14] The choroid plexus contains binding sites for mineralocorticoids, including aldosterone, and it is conceivable that raised CSF aldosterone may have an effect on CSF production, although this remains speculative.[13]

An additional possibility to consider is that the coexistence of malignant hypertension and IIH is not coincidental. The Cushing reflex is an intracranial baroreflex that attempts to preserve cerebral blood flow in the context of raised intracranial pressure (ICP). Although traditionally it has been viewed as a pre-terminal response in the context of severe head injury, recent work has highlighted it may be relevant in less catastrophic circumstances.[15] The raised intracranial pressure evident in our case may have precipitated systemic hypertension, and some support

for this comes with the observation that blood pressure control has not been a problem during follow-up and with a reduction in antihypertensive agents.

Less likely is the possibility that the raised ICP observed here may have been a result of early hypertensive encelphalopathy. Although initially ICP was quick to settle, later lumbar punctures continued to show ongoing raised ICP despite normal blood pressure.

In conclusion, systemic hypertension in IIH may be under-recognized and a small minority of patients with IIH may present in the context of a hypertensive crisis. Systemic hypertension may increase the potential for visual loss in IIH. It is important to exclude IIH when patients with systemic hypertension present with optic disc swelling.[16] Recognizing and treating both conditions appropriately may require joint care between those with an expertise in both conditions as treating either condition in isolation may not safeguard against possible complications.

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

***Note*:** Figure 1 of this article is available in colour online at www.informahealthcare.com/oph.

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