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Idiopathic Intracranial Hypertension, Empty Sella Turcica And Polycystic Ovary Syndrome - A Case Report

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ABSTRACT

Permanent visual loss is a well established major sequela of idiopathic intracranial hypertension (IIH). It is often insidious and frequently unnoticed by patients with IIH. It is vital to monitor these patients with serial perimetric and visual acuity tests because visual loss can be halted and occasionally reversed if treatment is begun early. We report a case of IIH with an empty sella turcica and polycystic ovary syndrome who developed visual field loss over ten years. This report illustrates the importance of close ophthalmic monitoring and detailed neurological and endocrinological evaluation to prevent complications in such patients.

Keywords: benign intracranial hypertension, empty sella turcica, idiopathic intracranial hypertension, polycystic ovary syndrome, visual loss

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INTRODUCTION

Idiopathic intracranial hypertension (IIH) is commonly known as benign intracranial hypertension. However, the term "benign" is a misnomer as permanent visual loss is an established major sequela of IIH[1-6]. An extensive ophthalmic review of 57 patients with IIH followed up from five to 41 years revealed that the incidence of detectable visual field or visual acuity loss may reach 49%[2]. We report a case of visual loss in a patient with IIH who also had an empty sella turcica and polycystic ovary (PCO) syndrome.

CASE REPORT

A 34-year-old, obese Eurasian female with a history of "essential" hypertension since the age of eighteen years presented with a four-month history of sharp generalised headache, blurring of vision in the left eye and diplopia on extreme lateral gaze. Her headache was aggravated by changes in posture, sneezing and coughing. She also had occasional episodes of transient visual obscuration and photopsia for a few years. Her menstrual periods were irregular for the preceding five months and hirsutism over the chin was present for the past year. There was no history of head injury, seizures, tinnitus, sensory or motor disturbances, exposure to heavy metals or ingestion of oral contraceptives, steroids or tetracyclines.

The patient was previously seen in another ophthalmic department ten years prior for bilateral optic disc swelling. Her Goldmann visual fields were normal then. She was lost to follow-up.

Ophthalmic examination revealed that her best-corrected visual acuity was 6/6 bilaterally. Her pupils were equally reactive to light and no relative afferent pupillary defect was present. Colour vision tested with Ishihara charts was normal. Extraocular movements appeared full but there was mild diplopia at the extreme lateral gaze. No other neurological abnormality was detected clinically. Indirect ophthalmoscopy revealed bilateral papilloedema (Fig 1). Haemorrhages and exudates were absent.

Goldmann visual fields showed a slightly enlarged blind spot and irregular constriction of the nasal field bilaterally and a small paracentral scotoma in the visual field of the left eye (Fig 2). Magnetic resonance imaging of the brain showed an empty sella turcica without other abnormality. Endocrinological evaluation revealed an elevated serum luteinising hormone (LH)/follicle stimulating hormone (FSH) ratio of more than 2.5 and raised serum testosterone and dihydroepiandrosterone. Other pituitary function tests were normal. Both ovaries were normal on ultrasonography. Cerebrospinal fluid pressure during lumbar puncture was more than 40 cm H_2O. Biochemical, microbiological and cytological examination of the cerebrospinal fluid were normal. Screening tests for connective tissue diseases were negative.

DISCUSSION

This patient had visual field defects due to IIH. Estimates of visual loss in IIH as measured by the presence of visual field defects range from 27% to 50% in retrospective studies[1-3]. Visual loss in IIH commences with the loss of peripheral nerve fibres. This causes constriction of the visual field which progresses to nasal depression, nasal steps and overt arcuate defects. Ultimately, loss of central nerve fibres may lead to reduced acuity. These field changes probably result from chronic ischaemia of the optic disc[4]. Peripapillary subretinal neovascularisation complicating papilloedema may also reduce visual acuity[4]. As visual loss is often insidious and frequently unnoticed by the patient, patients with IIH should be monitored carefully with frequent perimetric and visual acuity tests. It has been suggested that contrast sensitivity tests may demonstrate visual loss when no changes are detected in visual acuity or visual field tests[7].

Systemic hypertension has been found to be a

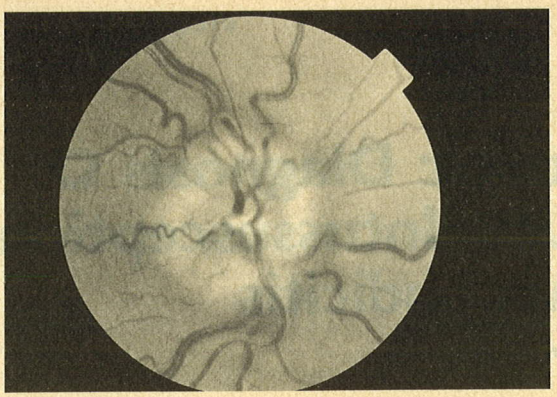

Fig 1a - Papilloedema (right eye).

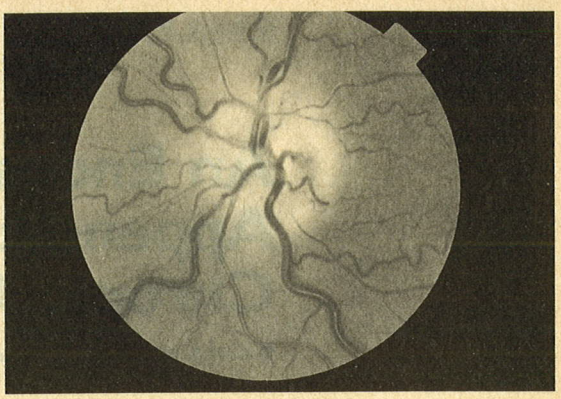

Fig 1b - Papilloedema (left eye).

significant risk factor for visual loss in patients with IIH[2]. The fact that this patient has had hypertension since the age of 18 years may have accelerated her visual loss. Her empty sella turcica is a recognised association in patients with IIH[4].

The elevated serum testosterone, dihydroepiandrosterone and LH/FSH ratio of more than 2.5 in the absence of adrenal dysfunction suggests the diagnosis of PCO syndrome. The increased androgen production explains the hirsutism in the patient. There is a continuum of ovarian volume ranging from normal to substantially enlarged in PCO syndrome[8]. To the best of our knowledge, the association of PCO syndrome with IIH has not been previously reported. The treatment for PCO syndrome includes weight reduction which is also part of the management for IIH.

Various forms of treatment have been described for IIH but none has been proven to be consistently effective[1]. Treatment can prevent further visual loss and reverse some of the visual defects if commenced early[1]. Repeated lumbar puncture is usually the initial mode of treatment and a weight reduction programme is begun concomitantly. Acetazolamide to decrease cerebrospinal fluid secretion or a short course of corticosteroids may be tried. If medical treatment fails and there is progressive visual loss, surgical procedures such as ventriculoperitoneal or lumboperitoneal shunt and optic nerve sheath fenestration may be attempted[1].

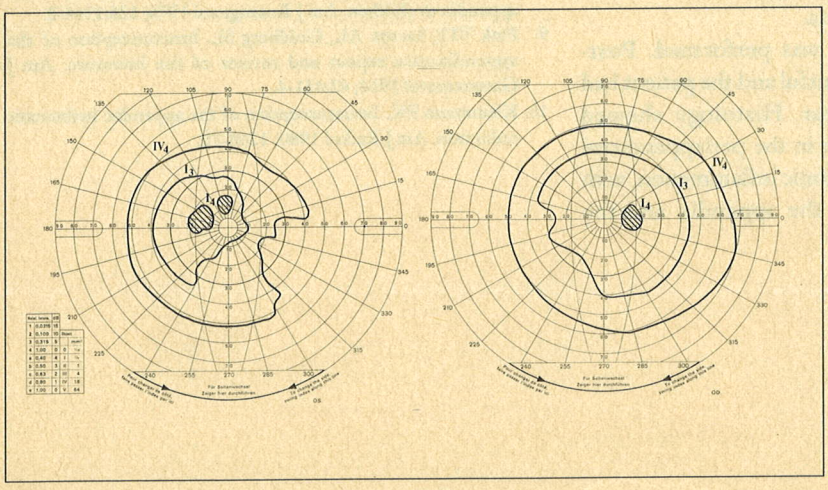

Fig 2 - Goldmann visual fields showing visual field loss.

This patient had six lumbar punctures over a period of four months. About 20 mLs of cerebrospinal fluid was removed during each lumbar puncture. The opening pressure was initially greater than 40 cm H_2O but eventually decreased to 28 cm H_2O. During this time, the patient reported subjective improvement in her symptoms. She also started on a weight reduction programme.

CONCLUSION

As permanent visual loss is a major sequela of IIH, the importance of close ophthalmic monitoring as part of the total management of these patients is emphasised. Detailed perimetric and visual acuity tests should be done serially to detect early visual loss so that treatment can be instituted promptly. In the presence of an associated empty sella turcica, tests of pituitary function should be performed.

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