CLINICAL STUDY

The contribution of macular changes to visual loss in benign intracranial hypertension

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Abstract

Purpose **To describe the significance of macular changes to visual outcome in benign intracranial hypertension (BIH).**
Method **The clinical and photographic records of 24 patients with BIH who required optic nerve sheath fenestration were analysed.**
Results **Macular changes were found in 21 of 48 (44%) eyes. These were: choroidal folds 9; circumferential lines (Paton's lines) 4; nerve fibre layer haemorrhage 3; macular stars 5; macular oedema 6; retinal pigment epithelial changes 4; subretinal haemorrhage leading to a macular scar 1. Significant visual loss attributable to the macular changes was found in 5 eyes in the short term and 3 in the long term. The 2 eyes that improved had macular stars. Of the 3 eyes that did not improve, 2 eyes had retinal pigment epithelial changes and 1 a large subretinal haemorrhage that led to a macular scar. These 3 cases had long-standing BIH.**
Conclusions **The majority of macular changes resolve and do not add to visual loss from optic nerve damage. Patients with marked macular oedema are at most risk of permanent visual loss and should be considered for early treatment such as optic nerve sheath fenestration.**

Key words Benign intracranial hypertension, Macular, Optic nerve sheath fenestration

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Macular abnormalities associated with papilloedema have not often been documented as a source of visual loss. Most descriptions concentrate on optic nerve damage and emphasise that macular changes resolve. Permanent macular changes can, however, occur. Gittinger and Asdourian[1] reported mottled macular pigmentation with preservation of normal vision in 3 cases of resolved papilloedema due to benign intracranial hypertension (BIH). Morris and Sanders[2] presented 6 patients with macular changes associated with papilloedema due to various causes, 4 of whom suffered permanent visual loss. Their findings were preretinal, intraretinal and subretinal haemorrhages, choroidal neovascularisation, choroidal folds, macular stars and retinal pigment epithelial changes. Subhyaloid and vitreous haemorrhage has also been reported and is thought to be due to forward dissection of severe peripapillary haemorrhage. Scattered haemorrhage can also occur due to an associated central retinal vein occlusion.[3]

We describe the frequency of macular changes in a series of patients with chronic papilloedema due to BIH who required optic nerve sheath fenestration and describe the incidence of visual loss due to these changes. We suggest macular abnormalities should be another indicator for early treatment such as optic nerve sheath fenestration.

Method

We carried out a retrospective analysis of the records of 24 patients with papilloedema due to BIH who required optic nerve sheath fenestration. There were 19 female and 5 male patients with an age range of 23–57 years (average 40 years). The majority of patients had presented elsewhere and had been followed up for a variable period prior to our assessment. The average follow-up by us was 2½ years (range 3 months to 8 years). The decision to carry out optic nerve sheath fenestration was made when medical or neurosurgical treatment had failed to control the intracranial pressure and prevent progressive visual deterioration. The visual acuity, visual fields (Goldmann), colour vision (Ishihara) and presence or absence of a relative afferent pupil defect had been documented and a clinical description and colour photographs recorded. Photographs were taken pre- and post-operatively in all cases and at various stages of follow-up. This information was used to record the frequency and type of macular changes and their effect on visual function. The pattern of visual field loss, assessment of colour vision and the pupil

Eye (1998) 12, 806–808

responses were used to assess the relative contributions of macular and optic nerve changes to visual loss. To be recorded as a macular lesion the lesion had to be more than one disc diameter from the temporal edge of the disc, i.e. in the parafoveal zone.

Results

We found macular changes in 21 of 48 (44%) eyes. These were: choroidal folds 9; circumferential lines (Paton's lines) 4; nerve fibre layer haemorrhage 3; macular stars 5; macular oedema 6; retinal pigment epithelial changes 4; subretinal haemorrhage leading to a macular scar 1.

Sixteen eyes had a best corrected visual acuity of <6/9 at presentation. Thirteen eyes retained a best corrected visual acuity of <6/9, but the average visual acuity was better in this group and the visual fields had improved. Macular changes contributed to the visual loss in 5 eyes in the short term and 3 in the long term (3/13, 23%). The 2 eyes that improved had macular stars that resolved on treatment. Of the eyes that did not improve, 2 eyes of the same patient had retinal pigment epithelial changes (Fig. 1) and one eye of a different patient a large subretinal haemorrhage that led to a macular scar (Fig. 2). Retinal pigment epithelial changes occurred in 4 eyes. In 2 the vision was 6/6; however, in 2 others it was reduced to 6/24 with preservation of good fields. These patients were known to have had BIH for a long period of time. The patient with reduced vision had variably attended for follow-up over a 16 year period prior to referral to us and optic nerve sheath fenestration. The macular pigmentation was present before surgery. The large subretinal haemorrhage was probably due to a choroidal neovascular membrane. A fluorescein angiogram was not carried out as the patient had a strong history of allergy.

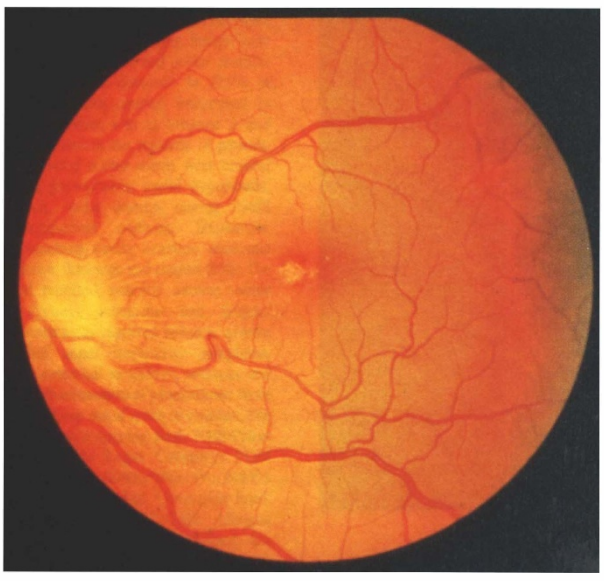

Fig. 1. *Retinal pigment epithelial changes with foveal atrophy and choroidal folds in a patient with long-standing BIH.*

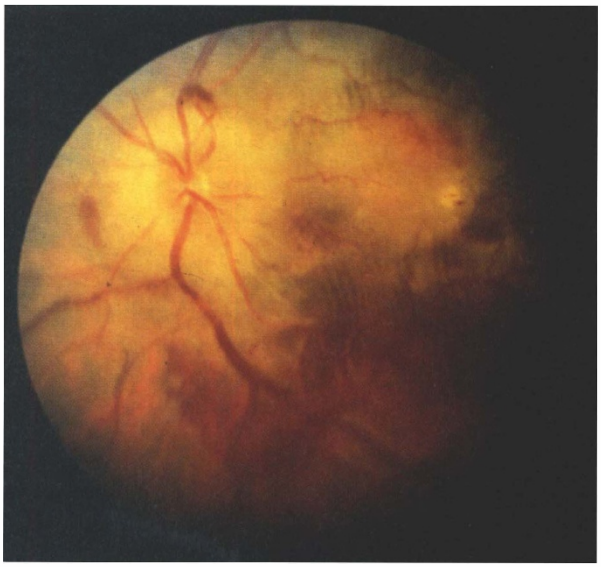

Fig. 2. *Large subretinal haemorrhage in a patient with long-standing BIH.*

Discussion

Raised intracranial pressure causes raised pressure in the optic nerve sheath. This produces both mechanical and haemodynamic changes at the optic nerve head and in the peripapillary region. If these changes are severe and long-standing they may spread to the macular region.

Choroidal folds occur due to indentation of the globe by a distended optic nerve sheath[4] and the subretinal accumulation of fluid.[5] The presence of choroidal folds has been associated with a persistently enlarged blind spot,[6] although such a finding can be explained by a hypermetropic shift induced by elevation of the circumpapillary retina.[5] Choroidal folds can lead to a permanent disturbance of the retinal pigment epithelium[1] and visual loss.[2]

Macular oedema can cause disruption of the retinal pigment epithelium leading to mottling of the retina.[1] Choroidal folds are a risk factor for choroidal neovascularisation and possibly central retinal artery occlusion.[7]

A macular star occurs due to leak of lipoprotein resulting from impaired retinal and choroidal perfusion, subsequent hypoxia and increased capillary permeability.[2] In itself it may not cause visual loss but is an indicator of disturbance of retinal function that may subsequently lead to permanent damage. Preretinal, intraretinal and subretinal haemorrhages in the macular region usually resolve but can lead to retinal scarring and damage to the retinal pigment epithelium.

Optic nerve sheath fenestration itself could lead to damage of nerve fibres from the macular region. In BIH the nerve sheath is usually separated from the nerve fibres by the surrounding excess cerebrospinal fluid, so the risk of damage to the nerve fibres is small. In a report by Spoor *et al.*[8] of 101 operations, 1 patient suffered a peripapillary haemorrhage that had no lasting effect on visual function.

The cohort of patients studied had optic nerve sheath fenestration because of progressive deterioration of visual function. This surgery stabilises or improves visual acuity and field.[8] We have shown that in this group with vision-damaging BIH macular changes are common, and a contributor to the reduction in visual function. Most of the macular changes are reversible on surgical relief of the effects of raised pressure on optic nerve head function. In a minority of patients permanent visual loss was attributable in part to macular damage. We therefore suggest that macular changes should be an indicator for intervention to normalise intracranial pressure at the optic nerve head, and prevent permanent visual loss from this source.

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