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Macular changes resulting from papilloedema

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SUMMARY Six cases are presented with macular changes in association with papilloedema; 4 suffered permanent visual loss. The present paper emphasises this previously infrequent finding and discusses the haemodynamic and mechanical factors responsible. The macular changes consisted of haemorrhages situated in front, within, or behind the retina, and occasionally the results of neovascular membrane formation produced secondary visual loss. Changes in the pigment epithelium were seen in 3 cases associated with choroidal folds. Macular stars rarely produce visual loss. Recognition of these changes is important in the assessment of the visual loss in papilloedema.

Visual loss as a result of papilloedema is usually due to atrophy and degeneration of optic nerve fibres and macular involvement as a cause has been neglected. This neglect is reflected in the paucity of well documented reports and the absence of any correlation with visual loss.

Schmidt and Wegener[1] described speckling of the macula in 2 patients with papilloedema due to intracranial tumours. Paton[2] in a review of patients with papilloedema at the National Hospital mentions that small white dots seen in a parabolic shape occurred in 5 cases. Similar observations were made by Colrat,[3] and both authors emphasised the absence of visual deterioration. One report of a macular hole and papilloedema[4] may represent only a chance finding.

The aetiology of cystic changes at the macula in post-mortem cases of disc oedema was discussed by Samuels,[5] though he failed to differentiate papilloedema from local causes of disc oedema. The absence of recent reports suggests that ophthalmologists should be consulted when visual deterioration is a feature of papilloedema.

This paper presents 6 selected cases of macular changes occurring in the presence of papilloedema (the term papilloedema being restricted to cases of disc oedema due to raised intracranial pressure) studied at the National Hospital by fluorescein angiography.

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Case reports

CASE 1

A 56-year-old woman was admitted to the National Hospital under the care of Dr Gooddy on 18 April 1973 with a long history of headaches of increasing severity over the last year and associated with gradual visual loss.

On examination she had a mild right spastic hemiplegia with bilateral extensor plantar responses.

Investigations included a plain skull *x*-ray, left carotid arteriogram, right vertebral arteriogram, ventriculogram, air encephalogram, and Myodil ventriculogram. These showed severe symmetrical hydrocephalus, without evidence of a tumour and a ventriculo-atrial shunt was inserted.

Ocular examination. Visual acuity was 6/24 in the right eye and counting fingers in the left eye with a large central scotoma. There was bilateral papilloedema with narrowing and sheathing of vessels and multiple small infarcts surrounding the discs. A massive preretinal haemorrhage was visible at the left macula (Fig. 1a), and then gradually absorbed with a resultant improvement in vision (Fig. 1b).

Conclusion. Temporary visual loss due to a preretinal macular haemorrhage occurred in a patient with bilateral chronic papilloedema.

CASE 2

This 55-year-old woman was admitted to the Brook General Hospital on 13 May 1970 under the care of

Dr Hierons with a 3-month history of frequent obscurations.

At that time the only finding was bilateral papilloedema. Skull *x*-rays, EEG, gamma scan, and examination of the cerebrospinal fluid were normal. A ventriculogram showed no enlargement of the ventricles, and a provisional diagnosis of benign intracranial hypertension was made.

In October 1970 the visual acuity in the left eye suddenly deteriorated and perimetry showed a central scotoma. On 9 March 1971 she was admitted to the National Hospital under the care of Professor Gilliatt, and a lumboperitoneal shunt was inserted by Professor Logue. This had to be revised in 1972. After this procedure her symptoms have improved with resolution of the obscurations, and the disappearance of papilloedema.

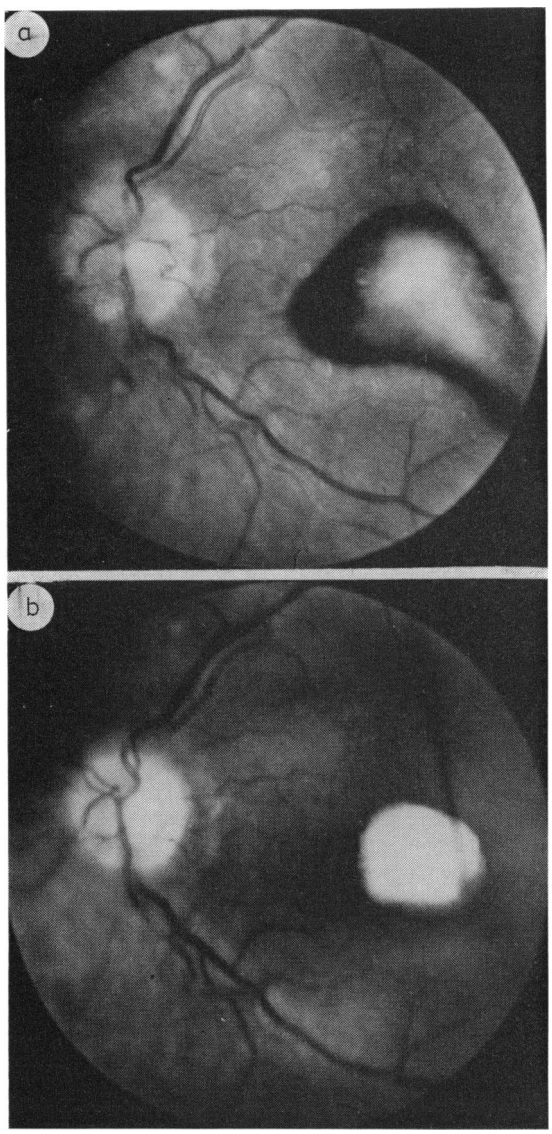

Fig. 1 *Case 1:* (a) *Copy of colour photograph showing a large preretinal haemorrhage at the macula,* (b) *which 5 weeks later had largely resolved.*

Ocular examination. Her visual acuity was 6/5 in the right eye and 6/60 in the left eye. There was bilateral enlargement of the blind spots and a left central scotoma. The fundi showed bilateral papilloedema with an extensive subretinal haemorrhage in the left eye which encircled the optic disc to involve the macula, together with a large subpigment epithelial haemorrhage supero-temporal to the disc which produced displacement of the retinal vessels.

Fluorescein studies showed (Figs. 2a and b) dilatation of the peripapillary plexus, tortuosity of the retinal vessels superotemporally, and traction on the macula. Choroidal fluorescence was visible above, but obscured below by the large subretinal haemorrhage. Repeat studies 9 months later showed restriction of the leakage to the vascular membrane.

Conclusion. Permanent visual loss resulted from a choroidal and subretinal haemorrhage in the macular region in a patient with benign intracranial hypertension.

CASE 3

A 53-year-old Hungarian was admitted to the National Hospital under the care of Dr Blau on 9 March 1972 with a history of severe episodic headaches for many years associated with a recent deterioration of vision. On admission he had an occipital bruit but no other neurological signs. Carotid and vertebral angiography revealed a large occipital dural arteriovenous malformation.

In April 1972 after a subarachnoid haemorrhage the feeding vessels to the malformation were ligated.

Ocular examination. Visual acuity was 6/18 in each eye, and visual fields showed large blind spots with a relative central scotoma in each eye. Fundus examination showed there was bilateral papilloedema with linear choroidal folds encircling the disc and radiating across the macular area. The choroidal folds persisted after treatment, and fluorescein angiography demonstrated their distortion of the macular area. Leakage of dye into the retina was visible in relation to a linear fold immediately above the macula in the left eye (Fig. 3) and similar distortion though with less leakage was visible in the right eye.

Conclusion. Bilateral visual loss resulted from macular distortion and choroidal folds in a patient with raised intracranial pressure due to an occipital dural arteriovenous malformation.

CASE 4

A 26-year-old man was first admitted to the National Hospital on 3 November 1971 under the care of Dr Denis Williams with a history of unsteadiness on his feet for 4 months, nausea, occipital headaches, and an inability to look to the right. He had obscurations of vision for 1 month before admission.

On admission he had a left cerebellar ataxia, left sided weakness, and a right VIth nerve palsy. A posterior fossa tumour was diagnosed by vertebral angiography and ventriculography.

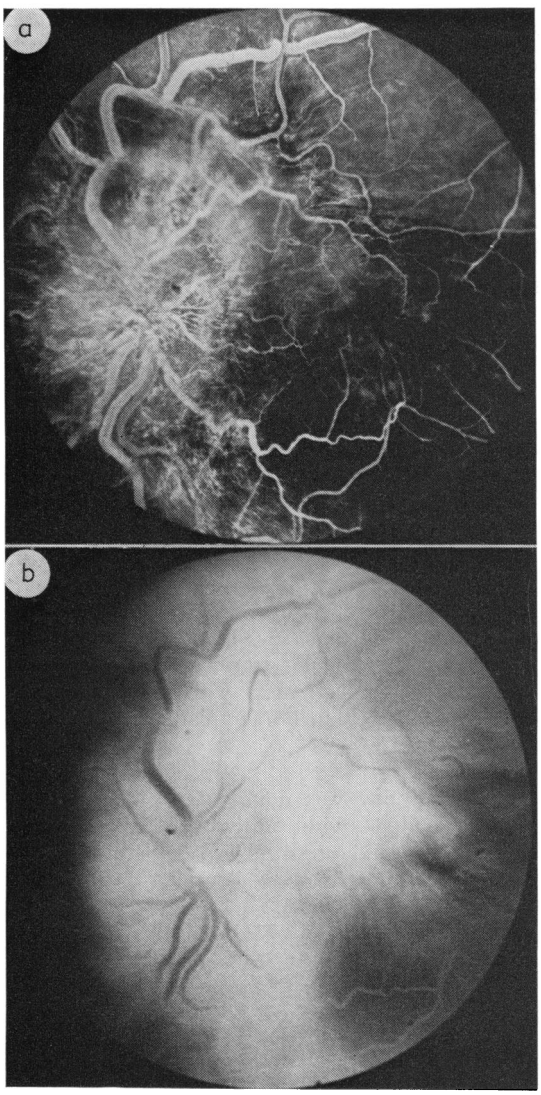

Fig. 2 *Case 2:* (a) *Fluorescein angiogram in the venous phase showing dilatation of the peripapillary capillaries with vascular distortion due to macula traction. The choroidal fluorescence is obscured below by an extensive subretinal haemorrhage.* (b) *Residual picture taken at 10 minutes to show extensive leakage at the optic disc extending superotemporally towards the macula.*

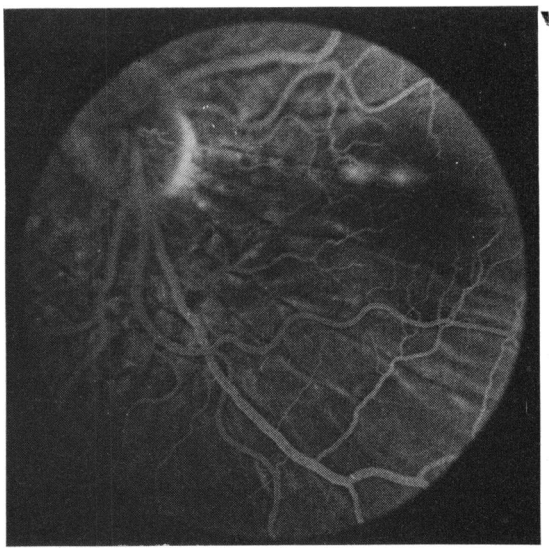

Fig. 3 *Case 4: Fluorescein angiograms showing the choroidal folds extending through the macula with hyperfluorescence occurring over one of the folds. Right eye in a venous phase.*

On 6 November 1971 a cystic cerebellar astrocytoma was removed from the left cerebellum. Postoperatively he made a good recovery.

Ocular examination. At admission his visual acuity was 6/18 in the right eye and 6/36 in the left eye, with a left temporal field loss and a right paracentral loss. Examination of the fundi showed gross papilloedema with dilatation of the superficial papillary and peripapillary plexuses. The peripapillary retina was swollen, with a well defined macular star and numerous haemorrhages and microaneurysms.

Fluorescein study showed an abnormally dilated superficial capillary plexus extending into the retina above and below the disc. This appeared to fill from the retinal system. Numerous microaneurysms were present on the disc surface, and there was gross leakage of dye from both discs in the late stages.

Visual acuity improved to 6/6 and 6/36 postoperatively.

Conclusion. Bilateral macular stars were associated with papilloedema due to a cerebellar astrocytoma. Visual reduction in the left eye was attributed to amblyopia.

CASE 5

A 54-year-old woman with symptomless papilloedema was referred by Dr Mackenzie for fluores-

cein angiography. No other neurological abnormality was found, and investigations which were normal included skull *x*-ray, brain scan, EEG, carotid angiogram, and a ventriculogram. In view of these findings a diagnosis of benign intracranial hypertension was made, and after a course of steroid therapy resolution of her papilloedema occurred.

Ocular examination. The visual acuity in each eye was 6/24 and perimetric examination showed enlarged blind spots. There was bilateral chronic papilloedema and in the right eye multiple lipid exudates condensed to form a macular star (Fig. 4a).

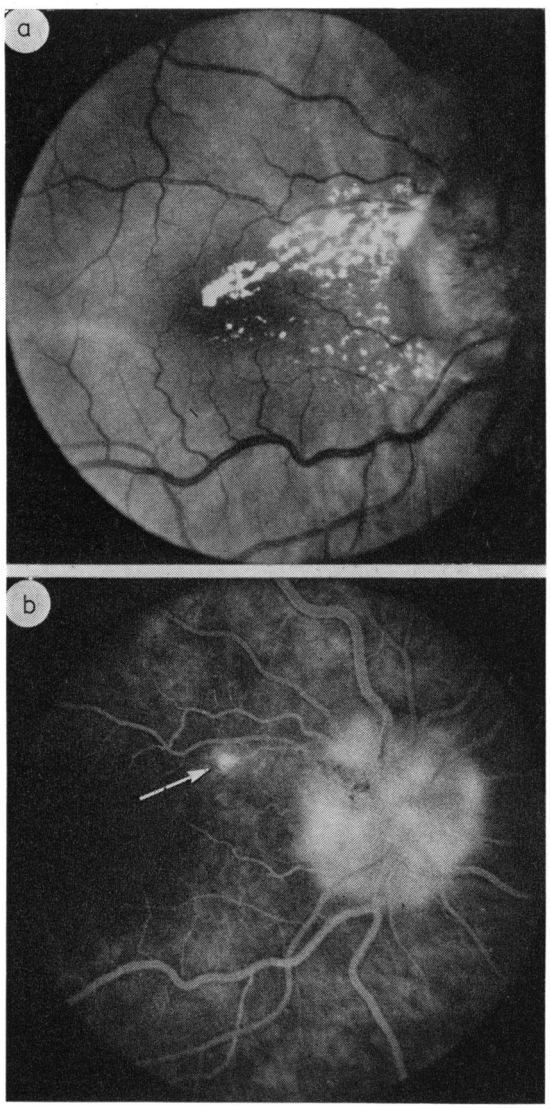

Fig. 4 *Case 5: Copy of a colour photograph of the right eye showing a macular star, and fluorescein angiography.* (b) *Showed leakage of dye at the disc with a smaller area of focal leakage superotemporal to the disc.*

Fluorescein angiography showed dilatation of the papillary plexus with extravasation of dye. In the right eye there was an area of focal leakage superotemporal to the disc (Fig. 4b, arrow).

Conclusion. A macular star with reduced visual acuity occurred in a patient with bilateral papilloedema due to benign intracranial hypertension.

CASE 6

A 6-year-old boy was admitted to the National Hospital in 1944 with otitic hydrocephalus. His cerebrospinal fluid pressure was 300 mmHg, and massive macular stars were described in both eyes by Mr Williamson Noble.

Ocular examination in 1971. His visual acuity was 6/9 in the right eye and 6/36 in the left eye, and his visual fields had central scotomata together with moderate enlargement of his blind spots. The optic discs were pale, the retinal vessels showed tortuosity at the disc margin, and pigment epithelial defects were visible at both maculae.

Fluorescein angiography showed attenuation of the arteries with linear pigment epithelial defects around the disc. In the macular regions punctate and nodular areas of hyperfluorescence were visible (Fig. 5).

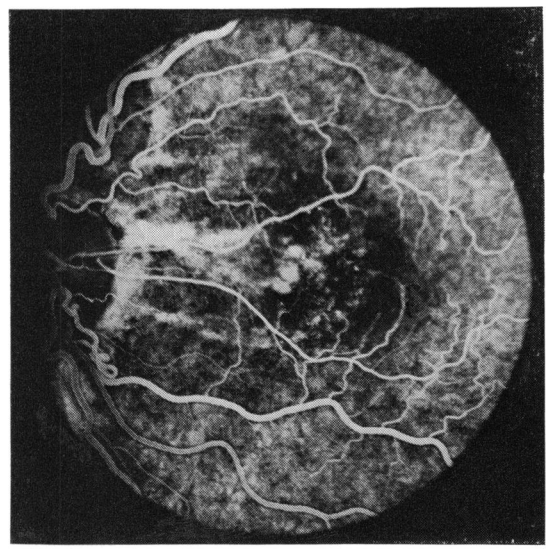

Fig. 5 *Case 6: Fluorescein angiogram in the late phase shows linear transmission defects extending from the disc to the macula with diffuse nodular areas of hyperfluorescence at the macula.*

Conclusion. Visual loss resulted from pigment

epithelial changes at both maculae in a patient with otitic hydrocephalus.

Discussion

PATHOPHYSIOLOGY OF PAPILLOEDEMA

Raised intracranial pressure produces papilloedema if the intracranial and perioptic nerve sheaths are in communication. Incision of the optic nerve sheath in experimental animals[6] and in man[7–11] produces resolution of disc oedema. Elevation of the CSF pressure in the optic nerve subarachnoid sheath produces both haemodynamic and mechanical changes at the optic disc and peripapillary region. The haemodynamic changes include delayed choroidal filling of the optic disc and peripapillary region, with compensatory dilatation of the retinal papillary and peripapillary plexus. Fluorescein angiography demonstrates the delayed choroidal filling, and the early filling of retinal peripapillary capillaries suggests that the vessels are functioning as compensatory shunt vessels.[12] The raised pressure in the optic nerve sheaths transforms this structure into a rigid tube, which indents the posterior globe, producing acquired hypermetropia and choroidal folds.[13]

The extension of both the haemodynamic and mechanical effects to the macular area when the papilloedema is either severe or long standing is therefore not surprising, and the cases selected for this report indicate the varieties of macular involvement.

HAEMODYNAMIC EFFECTS

Haemorrhages. Haemorrhages in the nerve fibre layer are a constant feature of fully developed papilloedema. Situated normally in relation to the optic disc, they may become preretinal or spread into the vitreous.[2] Their occurrence at the macula is rare, but is facilitated by marked dilatation of the radial peripapillary plexus, by impaired function of the endothelial cells as in the elderly, and when ocular perfusion is labile. The prognosis for vision is good, as resolution usually ensues.

Peripapillary haemorrhages in the subretinal layer are often seen in papilloedema, but ophthalmoscopic recognition is difficult owing to obscuration by the overlying disc tissue. They presumably represent abnormal permeability of the choroidal capillaries. Haemorrhages deep to the pigment epithelium (case 2) produce a major destruction of the retina with scarring and gliosis resulting in retinal distortion and traction. The prognosis for vision is poor but this manifestation is fortunately rare.

Macular star. The constellation of hard exudates that represents a macular star is more frequently seen in accelerated hypertension with disc oedema than in papilloedema. Gross papilloedema with massive fluorescein leakage at the optic disc is usually seen without macular exudates. The pathophysiological requirements for the development of a macular star are not known, but the following factors are probably contributory: (1) impaired retinal and choroidal perfusion to an area with high metabolic needs; (2) increased capillary permeability with leakage of large molecular weight lipoproteins.

The disc fluorescence rarely extends to the macular region even in the most severe cases, and macular fluorescence is not a concomitant finding in star formation. One patient (case 5), however, showed a focal area of leakage from a retinal vessel which may have been contributory.

MECHANICAL FACTORS

Distension of the distal optic nerve sheaths with CSF under high pressure may indent the posterior globe, with resultant hypermetropia, and permanent pigment epithelial changes due to folding or wrinkling of the choroid which was first described by Nettleship.[14] The mechanism was discussed by Bird and Sanders,[13] who described 2 main types: (1) the broad folding of the choroid with hyperfluorescent crests and troughs which show reduced fluorescence; (2) pigment epithelial wrinkling.

COMBINATION OF HAEMODYNAMIC AND MECHANICAL FACTORS

The combination of impaired peripapillary choroidal perfusion and the indentation by the distended nerve sheath may produce severe degenerative and hypoxic changes in the peripapillary choroid (case 6). The macula may therefore be involved at a preretinal, retinal, or pigment epithelial level.

We thank the consultants of the National Hospital for referring these patients to the Neuro-ophthalmology Unit, Miss Gaye Hopkins for secretarial assistance, and Mr Rolf Sennhenn for the photography.

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