Peripheral retinal haemorrhages with papilloedema

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SUMMARY Two cases are described with severe intracranial hypertension, papilloedema, and a hitherto unreported haemorrhagic peripheral retinopathy. The marked disc swelling in these patients has probably contributed to a venous occlusive element resulting in the haemorrhagic retinopathy.

Haemorrhages on the disc and in the peripapillary region remain one of the diagnostic features of papilloedema. In very severe cases haemorrhages may be seen more extensively, but it is exceptional to see them beyond the radius of the macular region.¹ However, the vascular retinopathies such as those associated with systemic hypertension, diabetes mellitus, and retinal vein occlusion characteristically display diffuse retinal changes.²

We report 2 cases of papilloedema with extensive peripheral retinal haemorrhages due to raised intracranial pressure alone. We are not aware of previous reports of such a picture.

Case reports

CASE 1

The patient was a 30-year-old multiparous Libyan woman. She complained of throbbing occipito-frontal headache for 3 years. Initially the pain was intermittent and frequently woke her at night. After 2 years the headache subsided during her tenth pregnancy, but recurred shortly after delivery of the child, some 6 months prior to referral. For 2 months she had noticed deterioration of vision in both eyes. This was initially gradual but then accelerated, leading to complete blindness a month before her admission. Her only other symptoms were mild bilateral tinnitus and transient rotational vertigo when the headache was severe. There was no vomiting, and menstruation was normal. She denied drug ingestion apart from aspirin. Her mother suffered from hypertension.

On examination she was grossly obese. Her blood pressure was 150/90 mmHg, and the heart, chest, and abdomen were clinically normal. There was no perception of light in either eye. The pupils were

equal at 4 mm and fixed. The optic discs were pale and swollen (Fig. 1), and there was a diffuse haemorrhagic retinopathy with numerous peripheral haemorrhages (Fig. 2). Intraocular pressure was normal, and ophthalmodynamometry showed pressures of 60/30 right and 60/20 left. Upward gaze was limited to 10 degrees; the eye movements were otherwise normal. There was a first-degree end point horizontal nystagmus to right and left. The remaining cranial nerves, including caloric responses, were normal. In the limbs there was no weakness, the reflexes were symmetrically sluggish, and the plantar responses were flexor. Sensation was normal. Her gait was slightly unsteady but not unduly so considering her blindness.

Investigations

Skull x-ray showed a thin and slightly eroded lamina dura of the dorsum sellae compatible with raised intracranial pressure. Computerised tomography showed normal ventricular size and position with no evidence of space occupation. Right carotid angiogram, vertebral angiogram, and lumbar air encephalogram were normal. An EEG was of low voltage with poorly developed alpha rhythm and no detectable responses to flicker. Lumbar puncture showed an opening pressure of 460 mm of water, rising to 500 mm on jugular compression. The CSF contained 114 red and 3 mononuclear cells $\times 10^9/l$ and protein 55 mg/100 ml (0.55 g/l); CSF Lange curve, IgG, sugar, Wassermann reaction, and cytology were normal. The ESR was recorded at 26 and 5 mm in 1 hour. The following investigations were normal: haemoglobin, PCV, white cell count and differential, platelet count, serum B₁₂ and folate, blood urea, electrolytes, sugar, calcium, phosphate, liver function tests, protein electrophoresis, autoantibodies, urine analysis, chest x-ray, and electrocardiogram.

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Fig. 1 (Case 1). Copy of colour photograph to show bilateral atrophic papilloedema with multiple punctate retinal haemorrhages in both eyes: a, right; b, left.

Management

After a short course of steroids her CSF pressure fell to 340 mm water. In view of the visual status, with little hope of recovery, and in the absence of any remediable lesion the steroids were discontinued.

CASE 2

This patient was a previously fit nulliparous housewife aged 21 years. She had been taking a low-oestrogen oral contraceptive for 6 months. Twelve

days before admission she had an upper respiratory tract infection that subsided in 48 hours. Two days later she noticed aching in both her shoulders, stiffness in her neck, and swelling of the second joint in her right hand. The following day she developed nausea and vomiting and became aware of frequent brief, jagged yellow flashes of light before both her eyes. She noticed intermittent double vision and that this was associated with her left eye turning inwards. She attended a general practitioner at this stage and migraine was diagnosed.

With simple analgesics her pains eased, but on the sixth day she developed intermittent tingling paraesthesiae in the face and hands. Over the next 48 hours she had recurrent episodes of giddiness, unsteadiness of gait, and a single brief period of loss of consciousness without convulsion and heralded by visual impairment. On the 11th day her vision deteriorated dramatically. Transient obscurations were followed by persisting upper field loss and finally a deterioration to perception of light bilaterally.

Examination showed her to be an obese young woman whose blood pressure was 140/80 mmHg. The heart and chest were normal and there were no abdominal masses. The visual fields were constricted to small central islands where acuity was reduced to counting fingers. There were frequent obscurations when she sat upright. She had gross bilateral papilloedema with infarcted discs and multiple peripapillary haemorrhages (Fig. 3). There were in

Fig. 2 (Case 1). Periphery of left eye to show punctate haemorrhages.

Fig. 3 (Case 2). Copy of colour photograph of both eyes to show disc swelling and peripapillary cotton-wool spots, with haemorrhages extending to the periphery: 3a, right; 3b, left.

Peripheral haemorrhages with a perivenous distribution in left eye (3c) and a late fluorescein angiogram of the right eye (3d) to show diffuse perivenous leakage of dye and residual disc leakage.

addition obvious peripheral haemorrhages in both fundi. These were associated with considerable venous leakage on fluorescein angiography. The pupils were widely dilated with a poor light response. She had a partial right sixth nerve weakness. The cranial nerves were otherwise normal. The reflexes, with the exception of ankle jerks, were absent and the plantar responses were bilaterally extensor.

There were no abnormal sensory or cerebellar signs.

Investigations

An EMI scan showed small ventricular size and no evidence of a space occupying lesion. Lumbar CSF pressure was in excess of 400 mm of water. The fluid was clear and contained 1 red and 3 mono-

nuclear cells, a protein of 18 mg/100 ml (0.18 g/l) and a normal sugar. The EEG showed a mild diffuse abnormality with a slowing of dominant rhythm to 7 c.p.s. and a moderate excess of theta activity. There was no focal lesion. Blood count showed a mildly elevated haemoglobin at 17G (PCV 49%: RBC 5.4×10^6). There was a polymorphonuclear leucocytosis. Platelet count was normal. The remaining investigations were normal. These included: ESR, sugar urea, electrolytes, osmolalities, Astrup test, calcium, phosphate, proteins, liver function, lead, autoantibodies, blood and CSF viral studies, Wassermann reaction, and culture of blood, stools and sputa, x-rays of chest, skull, and cervical spine, motor nerve conduction velocity, and sensory nerve action potentials.

Course and progress

The raised intracranial pressure was initially treated by repeated lumbar puncture, dexamethasone, acetazolamide, and glycerol. Her systemic symptoms resolved and her visual acuity improved to 6/6 right and 6/9 left, but still with marked field constriction and occasional postural obscurations. The retinal haemorrhages resolved and the discs developed marked optic atrophy. Despite the repeated lumbar punctures and drug therapy, her CSF opening pressure remained elevated at 300 mm of water. Because of this and increasingly Cushingoid features a thecoperitoneal shunt was inserted 10 weeks after her admission. Her CSF pressure settled to normal after this and remained so when steroid therapy was discontinued.

Discussion

In their classic paper Paton and Holmes¹ postulated that papilloedema was due to a rise in optic nerve sheath pressure producing compression of the intravaginal portion of the central retinal vein. They also attributed obstruction of lymph drainage from the papilla to an effect of the elevated sheath pressure. The haemorrhages were attributed to venous engorgement and the stretching and rupture of small disc capillaries.

For over a decade Hayreh^{3,4} has performed a series of experiments on rhesus monkeys and demonstrated that optic disc swelling is the first change in disc oedema in raised intracranial pressure. Elevated optic nerve sheath pressure causes a rise in optic nerve tissue pressure, stasis of axoplasmic flow,⁵ and swelling of the nerve fibres in the optic nerve head.⁶ This swelling then compromises the low pressure, predominantly venous, fine blood supply to the prelaminar region, and initiates a

vicious circle of oedema, congestion, and further oedema.⁷ The vascular changes on the nerve head are secondary to and not the cause of the disc oedema.⁷

Our cases show papilloedema in conjunction with peripheral retinal haemorrhages and a markedly elevated intracranial pressure. The clinical diagnosis in case one is 'benign' intracranial hypertension. Case 2 displays many of the features of this condition, but the diagnosis is not tenable in the context of a febrile illness, loss of tendon jerks, and extensor plantar responses, and remains one of raised intracranial pressure of uncertain aetiology.

The appearance of retinal haemorrhages varies according to their site in the retina, but the underlying mechanism remains identical with abnormal endothelial function due to anoxia. Anoxia at a capillary level can be readily understood in the small vessel disease of hypertension and diabetes mellitus. The combination of stasis, anoxia, and venous congestion in central retinal vein occlusion leads to profuse and characteristically peripheral retinal haemorrhage. Other stasis retinopathies behave similarly. The engorged veins, stasis, and superficial and deep haemorrhages of polycythaemia also occur with venous congestion and anoxia. In dysproteinaemia there is venous congestion and segmentation of blood flow, again conducive to anoxia, and fluorescein studies show circulatory delay and increased capillary permeability. There was no evidence of hyperviscosity or polycythaemia in either of these patients.

Paton and Holmes¹ demonstrated dilatation of the whole intraneural portion of the central retinal vein and also showed venous and capillary congestion in the retina, especially in the nerve fibre layer, for a short distance from the disc margin. It should be remembered that their pathological study was performed on the eyes of patients who had died from end stage, presumably very high, elevation of intracranial pressure. We feel that the marked elevation of pressure demonstrated in our patients led to a similar venous congestion and that, although this was not florid, it was enough to produce the retinopathy when acting together with the anoxic factor. The appearances we report are highly unusual, but recognition may be diagnostically important and alert the ophthalmologist to the occurrence of a haemorrhagic retinopathy with extreme elevation of intracranial hypertension.

We would like to thank Dr J. Morgan-Hughes and Dr R. Kocen for allowing us to use their cases.

We thank Mr Rolf Sennhenn for the photography and Mrs Gaye Jurewicz for secretarial assistance.

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