Papilloedema with retinal haemorrhages in idiopathic intracranial hypertension

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DESCRIPTION

A woman in her 20's was admitted with a 2-week history of severe headaches associated with vomiting, tinnitus, diplopia and transient visual obscurations. Her medical history was significant for obesity and oestrogen oral contraceptive use. On examination, her blood pressure measured 135/75 mm Hg. Neurological examination was unremarkable. Fundoscopy revealed papilloedema and retinal bleeding. A full blood count, clotting, renal, liver and bone profiles were all within normal limits. Cerebral MRI and CT venography were unremarkable.

On day 4 of admission, she was referred for ophthalmological assessment. Best corrected visual acuity was 6/19 in both eyes. Pupils, tonometry and ocular motility were normal in both eyes. Dilated retinal examination confirmed bilateral Frisén Grade 5 disc oedema, retinal vein tortuosity with congestion and widespread retinal haemorrhages (figure 1).

A subsequent lumbar puncture measured an opening pressure of 69 cm H₂O, which was reduced to 35 cm H₂O. The cerebrospinal fluid (CSF) fluid constituents were normal. CSF viral studies and oligoclonal band analysis were negative. We performed additional investigations for serum B12 and folate, antinuclear antibody, dsDNA, anti-cardiolipin antibodies, Haemoglobin A1c and protein electrophoresis, all of which demonstrated no further abnormalities.

A new diagnosis of idiopathic intracranial hypertension (IIH) was made. Oral acetazolamide was commenced at 250 mg two times a day. At 2-month

Figure 2 Colour fundus photographs and corresponding visual fields of the right eye (A) and left eye (B). There is bilateral atrophic papilloedema with visual field constriction.

follow-up, she was asymptomatic. Visual acuity improved to 6/9.5 in the right eye and remained static at 6/19 in the left eye. The retinopathy had resolved and the optic discs developed atrophic papilloedema (figure 2). Humphrey visual fields demonstrated marked constriction bilaterally.

We describe a patient presenting with IIH and haemorrhagic retinopathy, which is an unusual combination. The differential diagnoses included bilateral central retinal vein occlusions, systemic hyperviscosity and hypercoagulability syndromes, and blood dyscrasias. The improvement in the haemorrhages with reduction of the intracranial pressure indicated that IIH was the cause. Further investigations did not reveal an underlying systemic disorder. There are few case reports of retinal vascular changes in association with IIH, some due to central retinal vein occlusion,^{1 2} and others due to pre-retinal and vitreous haemorrhage.³ It has been speculated that an acute rise in intracranial pressure compresses the central retinal vein in the region of the optic nerve head.⁴ This mechanism would explain the observed venous tortuosity and distribution of haemorrhages along the vascular arcades.

Our patient presented with a reduced visual acuity. This point is significant, because acute papilloedema is often associated with preserved vision. We attributed the acute visual loss to haemorrhagic retinopathy. Other retinal causes of visual loss in IIH include macula oedema, subretinal fluid, retinal artery and vein occlusion and choroidal infarction.⁵ Despite the normal retinal appearances at follow-up, our patient's visual acuity remained limited. The development of bilateral disc atrophy and concentric visual field loss were consistent with optic neuropathy. This reflects the differing aetiologies of visual loss in acute and chronic IIH.

Figure 1 Colour fundus photographs of both eyes. (A) right and (B) left severe optic disc swelling with peripapillary haemorrhages and cotton wool spots. (C) right and (D) left engorged, tortuous retinal veins with retinal haemorrhages.

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The diagnosis of IIH relies on skilled direct ophthalmoscopy. Clinicians' lack of confidence with using the direct ophthalmoscope has been well documented.⁶ Retrospective studies suggest that up to 20% of IIH diagnoses are made without funduscopy.⁷ Retinal changes may be an under-reported aspect of diagnosis.

The combination of papilloedema and retinal vascular changes from IIH is rare. This may cause acute visual loss, but is reversible with timely reduction of raised intracranial pressure. Patients, however, may be left with long-term sequelae of optic atrophy. We emphasise the importance of direct fundus examination in all cases of suspected IIH.

Learning points

- ▶ Idiopathic intracranial hypertension (IIH) may present with papilloedema and haemorrhagic retinopathy.
- ▶ IIH presenting with a reduced visual acuity may be unrelated to papilloedema, emphasising the importance of a direct fundus examination.
- ▶ Timely reduction of raised intracranial pressure can relieve the vascular haemorrhage.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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