



Pseudotumour cerebri

Idiopathic intracranial hypertension, Benign intracranial hypertension

DESCRIPTION

Pseudotumour cerebri denotes raised intracranial pressure (ICP) without other evidence of intracranial pathology. It is strictly a diagnosis of exclusion, requiring the following:

- Signs and symptoms of raised ICP, such as headache and optic nerve swelling
- Elevated cerebrospinal fluid (CSF) opening pressure (>250mmH₂O), with normal composition
- Normal neuroimaging studies
- No unexplained abnormalities on neurological examination. The exception is palsy of the sixth cranial nerve; this is a common, non-specific feature of raised ICP.

These criteria exclude a range of diseases including intracranial space-occupying lesions, hydrocephalus and various inflammatory and infective processes. The raised ICP in pseudotumour cerebri has therefore been described as 'idiopathic'. However, it has a tendency to affect obese women in the child-bearing years, suggesting that abnormal hormonal activity may contribute to the syndrome. Several medication classes have also been implicated, including tetracycline, nalidixic acid, oral contraceptives, lithium, corticosteroids and amiodarone.

SYMPTOMS

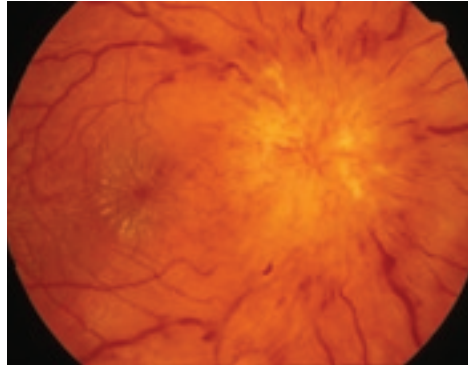
The classical symptoms of raised ICP are headache, nausea and vomiting, often worse in the morning. Sixth nerve palsy (eg lateral rectus muscle palsy) occurs in approximately 30 per cent of patients. Patients may describe transient visual 'blackouts', dizziness, tinnitus or other intracranial noises such as 'whooshing'.

SIGNS

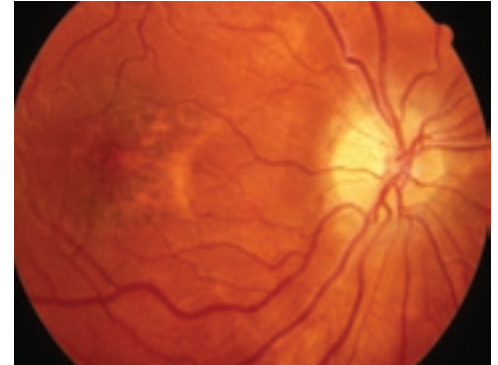
Signs are usually bilateral. Visual acuity may be reduced, but is often normal, particularly early in the disease course. Colour vision and contrast sensitivity may be impaired. The blind spot is often enlarged. On fundoscopy, the optic disc is typically swollen and hyperaemic, with indistinct margins. Other signs include loss of venous pulsation, peripapillary flame-shaped haemorrhages and cotton-wool spots. With advanced, chronic optic nerve damage, the disc becomes atrophic, with pale indistinct margins.

PREVALENCE

Rare (approximately one per 100,000 in the general population); 20 times more



Marked disc swelling (papilloedema) with cotton-wool spots, flame-shaped haemorrhages and macular star



Same eye, three years later: optic atrophy; chorioretinal atrophy beneath the macula; sheathing of retinal veins

common in overweight women of child-bearing age.

SIGNIFICANCE

Some of the causes of raised intracranial pressure and papilloedema are potentially life-threatening. The consequences of untreated idiopathic raised intracranial pressure may include severe headache, vomiting and double vision, and occasionally severe and permanent loss of vision.

SEE ALSO

Papilloedema, Hypertensive retinopathy, Glaucoma classification.

MANAGEMENT

Urgent

Urgent imaging of the brain and orbit is needed to exclude intracranial pathology. If imaging is normal, lumbar puncture is performed for CSF opening pressure and laboratory analysis. Formal optic nerve evaluation, including visual fields, is required to assess optic nerve function and response to treatment.

Oral medications

When possible, medications associated with pseudotumour cerebri are ceased. Weight loss is critical in the medium to long term. Concurrent treatment of increased intraocular pressure and systemic hypertension will minimise further damage to the optic nerve.

Additional treatment is indicated for intractable headache or progressive decline in optic nerve function. A common initial medical treatment is acetazolamide, a carbonic anhydrase inhibitor and diuretic. Corticosteroids have been used in the setting of acute

visual loss, but may increase fluid retention, systemic hypertension and intraocular pressure.

Incisional surgery

Symptom relief is achieved in some patients with serial therapeutic lumbar punctures, to reduce CSF pressure to normal levels. Optic nerve sheath decompression or fenestration surgery is performed when visual loss progresses despite maximum medical therapy. When headache is a dominant symptom, lumboperitoneal shunt creation may be considered.

Review

Review is conducted at two-week intervals initially, with attention to symptoms, visual acuity and fields, fundoscopy and weight loss. Subsequent review intervals are determined by the patient's progress. Regular review and appropriate treatment may prevent permanent loss of vision.

The full series of these articles will be available in the book *Posterior Eye Disease and Glaucoma A-Z* by Bruce AS, O'Day J, McKay D and Swann P. £39.99. For further information click on the Bookstore at opticianonline.net

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