The swollen optic disc

Introduction

A swollen optic disc can be an ominous neuro-ophthalmological sign. In some patients it indicates raised intracranial pressure as a result of serious neuropathology, but in other cases it is caused by local pathology within the eye or orbit. Recognizing the presence of optic disc oedema is a key skill for all doctors, which is occasionally required not only in clinical practice but also in postgraduate exams. This article summarizes the main indications for examining the optic disc (*Table 1*), signs to look for and what to do next if you suspect a patient has a swollen optic disc.

Appearance of the disc

The normal optic disc lies nasal to the fovea and is an orange-pink colour (*Figure 1*). The disc margins (M) are normally sharply demarcated. In the centre of the optic disc there is usually a pale depression or 'cup' (C) surrounded by a darker neuroretinal rim (R); the cup is variable in size but usually takes up less than two thirds of the total disc diameter. The central retinal artery and vein emerge from the nasal edge of this central cup and branch before spreading out to supply the four retinal quadrants. In many (but not all) healthy subjects the largest retinal veins show spontaneous pulsation as they cross the neuro-retinal rim.

The cardinal features of any swollen optic disc are elevation of the optic nerve head (i.e. above the level of the surrounding retina), and blurring of its margins (*Figure 2*). In addition a number of other features may be present and can help with the diagnosis (*Table 2*).

Table 1. When to examine the optic disc

Headache worse on waking, leaning forward, sneezing, coughing, straining and lying flat
Headache associated with vomiting
Headache associated with reduced level of consciousness
Headache associated with fever
Diplopia (especially when caused by an abducens nerve palsy)
Systolic blood pressure >190 mmHg
Head trauma
Acute or subacute loss of vision

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Causes of disc swelling

The causes of a swollen disc can broadly be divided into three groups:

1. Papilloedema (swollen disc associated with raised intracranial pressure)
2. Pseudopapilloedema (an 'elevated' disc which is not swollen, i.e. not associated with any true oedema)
3. Disc oedema caused by local pathology within the eye or optic nerve.

The management of patients in each group varies considerably. The simplest way to distinguish between these groups is to measure visual acuity: a patient with preserved visual acuity is more likely to have papilloedema (in its early stages) or pseudopapilloedema, whereas a patient with reduced visual acuity is more likely to have disc swelling caused by local pathology within the eye or optic nerve.

Figure 1. A healthy optic disc with clear margins (M) and normal cup (C) to disc ratio. R = rim.

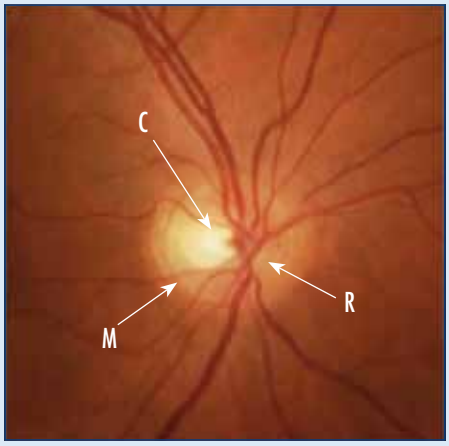


Figure 2. A swollen optic disc, which appears elevated with blurred margins. Venous congestion and retinal haemorrhages can also be seen.

Papilloedema

This is defined as optic disc swelling secondary to raised intracranial pressure and represents a medical emergency. The upper limit of intracranial pressure in healthy adults is 250 mmH_2O. *Table 3* lists some features of the history and examination that should alert the clinician to the possibility that a

Table 2. Features of a swollen optic disc

Cardinal features	Blurred margins
	Elevation
Other changes	Hyperaemia
	Haemorrhages
	Absence of a central cup
	Absence of spontaneous venous pulsations
	Signs of venous congestion (peripapillary haemorrhages, cotton wool spots, engorged veins)
	Retina: folds concentric to disc margins (Paton's lines)
	Choroid: folds horizontally across macula
	Macula: subretinal fluid and 'star' of hard exudates
Chronic changes	Pale disc
	Peripapillary gliosis
	Sheathing of retinal vessels

Table 3. Clinical features suggesting raised intracranial pressure

Symptoms	Headache (worse on lying flat, leaning forward, coughing, straining)
	Nausea or vomiting
	Diplopia
	Transient visual obscurations
	Tinnitus
	Reduced level of consciousness
Signs	Bilateral disc swelling
	Absence of spontaneous retinal venous pulsations
	Preserved vision
	VI cranial nerve palsy (unilateral or bilateral)

Table 4. Causes of raised intracranial pressure

Intracranial	Space-occupying lesion (tumour, aneurysm, abscess)
	Hydrocephalus
	Diffuse brain disease (encephalitis, oedema)
	Diffuse meningeal disease (infection, infiltration)
	Cerebral venous sinus disease (thrombosis, stenosis)
Systemic	Obstructive sleep apnoea
	Systemic hypertension
	Hypercapnia
	Anaemia
Other	Idiopathic intracranial hypertension

swollen disc is being caused by raised intracranial pressure. *Table 4* lists some of the main causes of raised intracranial pressure.

The disc swelling in papilloedema may have a variety of different appearances (*Figure 3*). In early papilloedema (*Figure 3a*), the nasal disc margin is blurred and there is mild elevation and hyperaemia of the neuro-retinal rim. If the intracranial pressure rises suddenly there may be signs of acute papilloedema (*Figure 3b*) with florid congestive vascular signs. A slower and more protracted rise in intracranial pressure leads to chronic papilloedema (*Figure 3c*) in which the disc is markedly elevated ('like a champagne cork') without vascular signs. Eventually there may be irreversible damage to the optic nerve fibres leading to atrophic papilloedema (*Figure 3d*); this is the only stage of papilloedema in which there is significant permanent loss of sight, although patients with earlier stages of papilloedema sometimes report briefly losing the sight in one or both eyes for a few seconds, particularly when bending down (transient visual obscurations).

In most cases, the disc changes associated with raised intracranial pressure are bilateral; however, unilateral swollen discs have been reported (Lepore, 1992; Wattamwar et al, 2010), and in these rare cases it is assumed that the 'normal' disc is protected from the raised intracranial pressure by compartmentation of the subarachnoid space. In addition, patients with anterior cranial fossa tumours may develop Foster Kennedy syndrome, which is optic atrophy of one disc as a result of direct compression of that optic nerve, and swelling of the contralateral disc as a result of raised intracranial pressure (Pastora-Salvador and Peralta-Calvo, 2011).

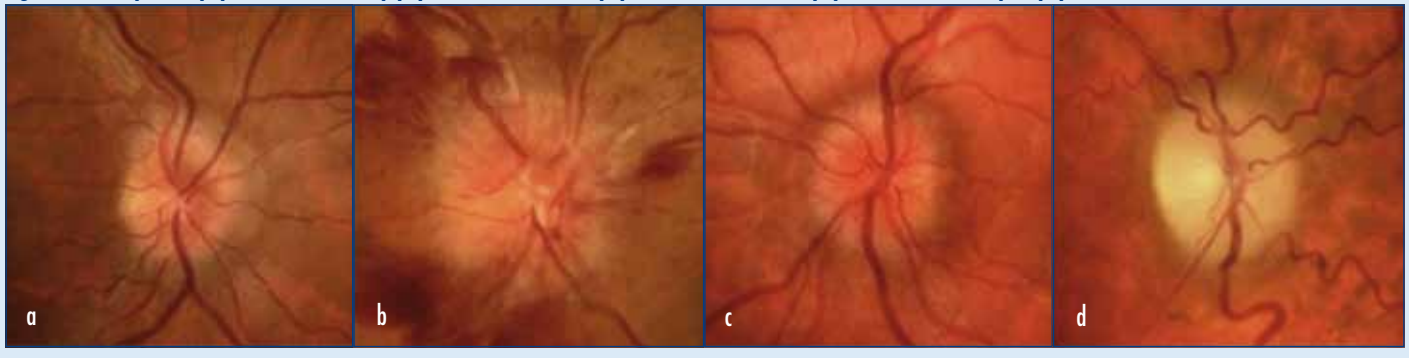
A useful sign in distinguishing true papilloedema from other causes of disc swelling is the absence of spontaneous venous pulsations, which are present in 70–80% of normal subjects (Hu et al, 2008). As the intracranial pressure rises, it becomes equal to the intraocular pressure, causing the spontaneous venous pulsations to stop. This usually occurs when the intracranial pressure rises above 204 mmH_2O (Walsh et al, 1969) and is a sensitive marker of raised intracranial pressure (Jacks and Miller, 2003). However, false positive (absent pulsation when intracranial pressure is normal) and false negative (visible pulsation when intracranial pressure is raised) cases do occur so this sign should always be interpreted in the context of other clinical findings.

How good is the correlation between optic disc appearance and raised intracranial pressure? As noted above, some patients with raised intracranial pressure only show disc swelling in one eye, and there are other examples where both optic discs remain normal in appearance. In one paediatric study, the presence of disc swelling was found to be highly predictive of raised intracranial pressure in children above the age of 8 years (specificity 98%) but the sensitivity was much lower (32%). This may be in part explained by the fact that papilloedema takes several days to develop so if clinical examination is performed early on in the disease, papilloedema may not be apparent (Tuite et al, 1996).

Pseudopapilloedema

This may be defined as an apparent elevation of the optic nerve head without the presence of true oedema. In some cases this is a result of congenital anomalies in the local anatomy of the optic nerve head, e.g. small crowded discs (*Figure 4a*), tilted discs or myelinated nerve fibres (*Figure 4b*), and in others there are acquired changes such as optic disc drusen, either 'exposed', i.e. visible on the surface (*Figure 4c*) or 'buried' (*Figure 4d*). Some causes of pseudopapilloedema are listed in *Table 5* (Fuentes-Peliera and Hodelín-Tablada, 2010).

Figure 3. Examples of papilloedema. a. Early papilloedema. b. Acute papilloedema. c. Chronic papilloedema. d. Atrophic papilloedema.



In all cases, regardless of cause, patients with pseudopapilloedema do not complain of visual symptoms. The abnormal disc appearance may be an incidental finding or it may be noticed when examining a patient who is complaining of headache, in which case it is important to distinguish these discs from true papilloedema.

Local causes of disc swelling

Pathology within the eye or optic nerve may also cause disc swelling. In most cases the disc swelling is unilateral and associated with reduced vision. The patient may also complain of peri-ocular pain. There are many types of local pathology that may cause disc swelling, including inflammation, infection, ischaemia and compression, and some of the main aetiologies are summarized in *Table 6*. Examples of local causes of disc swelling are showed in *Figure 5*.

Epidemiology

How often do these different types of disc swelling occur? In one study of 100 consecutive patients presenting with disc swelling (Binder et al, 1986), raised intracranial pressure was the commonest cause (32%), followed by vascular disease (22%) and inflammation (18%). Another study in Korean subjects showed that the commonest diagnosis in patients with unilateral disc swelling was non-arteritic anterior ischaemic optic neuropathy (40%), followed by optic neuritis (32.5%). However, when considering only patients with bilateral disc swelling, raised intracranial pressure was the commonest cause (44.4%) (Jung et al, 2011).

Table 5. Causes of pseudopapilloedema

Congenital	Small crowded optic disc (hyperopes)
	Tilted disc (astigmats)
	Dysplastic disc
	Myelinated nerve fibres
Acquired	Optic disc drusen: buried or exposed

Completing the examination

The correct interpretation of a swollen optic disc depends critically on the rest of the clinical assessment. In addition to testing visual acuity, it is important to also assess colour vision and the field of vision. The pupillary responses to light should be examined, and a swinging flashlight test performed to look for a relative afferent pupil defect. The orbits and ocular motility should also be examined, particularly looking for any weakness of abduction that may suggest VI cranial nerve palsies.

Table 6. Local causes of optic disc swelling

Pathology in the optic nerve	Inflammatory	'Optic neuritis' (demyelinating, granulomatous, autoimmune)
	Infectious	Bacterial (syphilis, Lyme, tuberculosis)
		Viral (HIV, varicella, cytomegalovirus, hepatitis A/B)
		Fungal (cryptococcus, aspergillus, mucor)
	Ischaemic	Arteritic (especially giant cell arteritis) or non-arteritic
	Compressive	Orbital tumours
	Infiltrative	Leukaemia, lymphoma
Pathology in the eye	Vascular	Central retinal vein occlusion, vasculitis
	Inflammatory	Posterior uveitis

In true papilloedema there is enlargement of the physiological blind spot, but otherwise acuity, colour and visual field are all preserved (except at the late atrophic stage). However, there may be uni- or bilateral abduction weakness (a false localizing sign) and in some cases there may also be

Figure 4. Examples of pseudopapilloedema. a. Small crowded disc. b. Myelinated nerve fibres. c. Exposed optic nerve drusen. d. Buried optic nerve drusen.

Figure 5. Examples of local causes of disc swelling. a. Optic neuritis associated with multiple sclerosis. b. 'Non-arteritic' anterior ischaemic optic neuropathy. c. Optic neuritis associated with sarcoidosis. d. 'Arteritic' anterior ischaemic optic neuropathy associated with giant cell arteritis.

other neurological deficits, which help localize a focal lesion.

In disc swelling caused by local disease in the eye or optic nerve there is usually a significant impairment of sight associated with a relative afferent pupil defect (unless the pathology is bilateral and symmetric), and there may be additional signs when examining the eye or orbit to indicate the likely underlying aetiology.

In pseudopapilloedema vision is preserved, there is no relative afferent pupil defect, and the rest of the ocular orbital and motility examination are unremarkable. A quick look at the patient's spectacles may confirm an associated refractive error.

The pathophysiology of optic disc swelling

The pathogenesis of optic disc swelling is not fully understood but disturbance of axoplasmic transport is the final common mechanism in most causes of disc oedema (Van Stavern, 2007). This mechanism has been demonstrated in monkey models of raised intracranial pressure (Hayreh, 1977) and optic nerve head ischaemia (McLeod et al, 1980). In cases of raised intracranial pressure, it is likely that the high CSF pressure in the optic nerve sheath causes axoplasmic flow stasis, resulting in swelling of axons (Hayreh, 1977).

Initial investigations and management

If raised intracranial pressure is suspected, urgent neuroimaging is first needed to rule out an intracranial mass, hydrocephalus or a cerebral venous sinus thrombosis. The most useful study is a gadolinium-enhanced magnetic resonance imaging and magnetic resonance venography, but in some centres a computed tomography scan of the head is easier to arrange in the first instance. If no structural abnormality is found, then a lumbar puncture should be performed, both to measure the CSF pressure and to evaluate the CSF constituents. Blood tests should be sent off from the outset, including a full blood count, renal function, clotting screen and general inflammatory markers (e.g. erythrocyte sedimentation rate or C-reactive protein) (*Table 7*). If raised intracranial pressure is confirmed but the scans show no cause then more detailed evaluation is needed, necessitating referral to neurologists or neuro-ophthalmologists.

Prompt treatment of raised intracranial pressure is needed otherwise permanent damage may occur to the optic nerves and sight. As well as addressing the primary pathology (when this can be identified), the intracranial pressure may be lowered to a safe level medically (with diuretics such as acetazolamide) or surgically (by diverting CSF using a shunt).

In patients with headache whose optic discs are 'equivocal', but who have no other neurological abnormalities and are otherwise well, it is sensible to first obtain an opinion about the optic disc appearances from an ophthalmologist before requesting extensive investigations for possible raised intracranial pressure.

When the initial clinical assessment reveals poor sight and a probable local cause for the disc swelling, then the patient should be urgently referred to an ophthalmologist for further management. If the patient is elderly and gives a history of acute loss of vision associated with significant headache or pain around the eye then consider a diagnosis of giant cell arteritis, send off blood to measure the erythrocyte sedimentation rate and C-reactive protein, and start high-dose steroids (1 mg/kg) without waiting for the results of a temporal artery biopsy.

Conclusions

A swollen optic disc is occasionally found when performing a funduscopic examination and should be looked for in any patient presenting with new onset headache, diplopia or loss of sight. The most important condition to urgently rule out is raised intracranial pressure. This should be investigated first with neuroimaging; if this shows no abnormality then the patient needs a lumbar puncture to measure the intracranial pressure and examine the CSF constituents. In cases of unilateral disc swelling, a local pathology within the eye, optic nerve or orbit should be considered (usually associated with significant loss of sight) or pseudopapilloedema (where sight is usually unaffected) and refer to an ophthalmologist for further management. BJHM

Table 7. Investigations if papilloedema is suspected

Blood tests: full blood count, coagulation, inflammatory markers
Gadolinium-enhanced magnetic resonance imaging and magnetic resonance venography
Lumbar puncture: measure CSF pressure and send off for analysis of constituents

Conflict of interest: none.

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KEY POINTS

- Papilloedema is an emergency.
- Both eyes must be examined to detect a unilateral swollen disc.
- A normal funduscopic examination does not rule out raised intracranial pressure.
- Neuroimaging and lumbar puncture are the key investigations to diagnose raised intracranial pressure.
- Reduction of intracranial pressure can be achieved medically or surgically in addition to targeting the underlying pathology.