

Swollen optic discs

The need for timely diagnosis

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The differential diagnosis for swollen optic discs covers a wide range of conditions, from normal variants to ophthalmic and neurological causes. Investigation of the cause is needed in every patient with disc swelling.

The optic disc is the anteriorly visible portion of the optic nerve within the eye. Pathological changes seen at the optic disc fall into three broad categories: cupping, pallor and swelling.

Swelling of the optic disc occurs when there is a hold-up of the axoplasmic flow along the optic nerve at the level of the lamina cribrosa for any reason (Figure 1). Therefore, all cases of disc swelling will need some form of investigation to determine the cause. None of the different causes of true optic disc swelling can be diagnosed clinically (Figure 2).

The term papilloedema is reserved for cases where the disc swelling is known to be due to raised intracranial pressure.

Causes of optic disc swelling

There are many causes of optic disc swelling; this topic alone could fill a textbook. This article discusses the broad principles of diagnosis and the most common causes.

Normal anatomical variants

There are some normal variants of optic disc appearance that can mimic disc swelling, including:

- long-sightedness (look to see whether the patient's spectacles cause magnification)
- optic disc drusen
- myelinated nerve fibres (Figure 3).

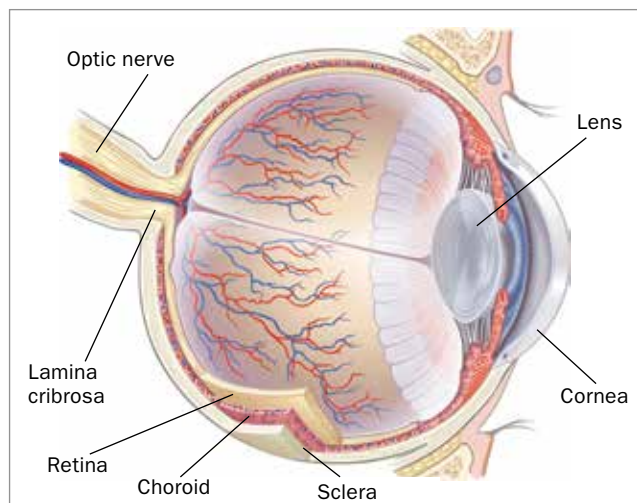


Figure 1. Illustration of the normal eye anatomy showing the position of the lamina cribrosa.

Patients with such normal variants have 'pseudopapilloedema' and are typically not at risk of vision loss. The exception is optic disc drusen, which rarely can cause changes in peripheral vision over the long term; however, this diagnosis can be very difficult to make when using a direct ophthalmoscope.

Ophthalmic causes

Not all disc swelling is neurological. Ophthalmic conditions associated with a swollen optic nerve include:

- ocular inflammation such as uveitis, posterior scleritis or retinitis
- central retinal vein occlusion
- hypotony (very low eye pressure).

Raised intracranial pressure

Disc swelling can occur due to raised intracranial pressure of any cause, including:

- a space-occupying lesion
- dural venous sinus thrombosis
- meningitis.

Malignant hypertension also falls into this category and can result in swollen optic discs with associated retinal vascular changes, including arteriolar narrowing, arteriovenous crossing changes, haemorrhages and cotton-wool spots.

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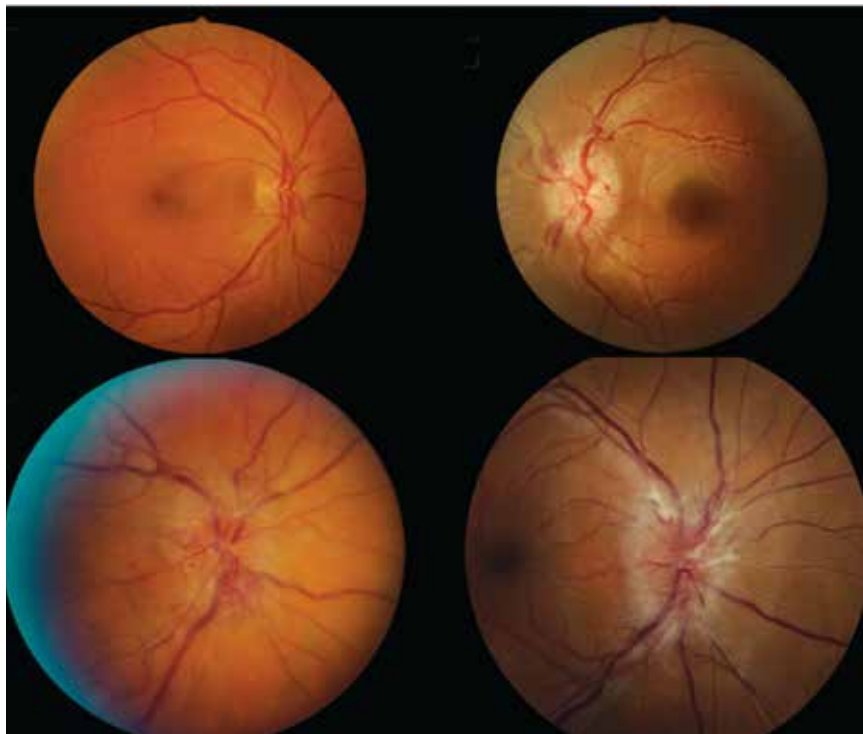


Figure 2. Fundus photographs of swollen optic discs with different causes. None of the swollen discs are clinically distinguishable; all required investigation for diagnosis. a (top left). Early papilloedema from idiopathic intracranial hypertension. b (top right). Severe papilloedema from dural venous sinus thrombosis. c (bottom left). Nonarteritic anterior ischaemic optic neuropathy in a 'disc at risk'. d (bottom right). Syphilitic optic neuritis.

In pseudotumour cerebri intracranial pressure is raised secondary to medication use (e.g. tetracyclines), anaemia or obstructive sleep apnoea, to name just a few causes.

Idiopathic intracranial hypertension is typically seen in women of childbearing age who are overweight, but is a diagnosis of exclusion.

Optic nerve inflammation or infiltration

Optic neuritis occurring in young white patients is typically associated with multiple sclerosis. If optic neuritis occurs in patients of Asian or African descent then other causes such as sarcoidosis or neuro-myelitis optica must be considered. Syphilis, tuberculosis and *Bartonella henselae* infection (cat scratch disease) are some of the most common infective causes of optic neuritis. Rarely, a malignant infiltration from leukaemia or lymphoma can cause

disc swelling and present similarly to optic neuritis.

Optic disc ischaemia

A patient with anterior ischaemic optic neuropathy (AION) presents with a swollen optic disc caused by ischaemia of the optic disc microcirculation. The swelling may be segmental, for example affecting either the superior or inferior disc. AION can have either an arteritic cause such as giant cell arteritis or a nonarteritic cause such as nocturnal hypotension in a 'disc at risk', or 'crowded' disc, defined as a disc with a small optic cup to disc ratio. Nonarteritic AION is not an embolic phenomenon.

How the patient presents

Most optometrists will refer patients with suspected pseudopapilloedema directly to an ophthalmologist. If the optometrist suspects true disc swelling then patients

may be advised to consult a GP for systemic assessment or for ongoing referral. In this case the optometrist may provide the GP with information about the patient's vision and often visual field test results.

Some patients with headache, neurological symptoms or vision loss may present initially to a GP. If disc swelling is found on direct ophthalmoscopy then it is useful to categorise patients into one of two broad categories by testing central visual acuity: disc swelling with vision loss or disc swelling with normal central vision.

Disc swelling with vision loss

Disc swelling with vision loss is most likely due to a direct optic nerve abnormality such as:

- optic neuritis (subacute loss of vision over days, with pain on eye movement)
- infiltration (painless loss of vision over days to weeks; may have associated symptoms of malignancy)
- ischaemia:
 - arteritic AION (symptoms suggestive of giant cell arteritis; see the Box)
 - nonarteritic AION (painless loss of vision typically noticed on waking, in a patient with vasculopathic risk factors or sleep apnoea or with use of erectile dysfunction medication).

Any ophthalmic causes of disc swelling will also affect vision.

Disc swelling with normal central vision

Disc swelling with normal central vision is classically seen in patients with raised intracranial pressure. Associated symptoms include:

- a headache that is worse when lying flat
- pulse-synchronous rumbling or whooshing tinnitus
- transient vision loss on bending forward
- nausea and vomiting
- focal neurology.



Figure 3. Photographs of the optic disc in patients with pseudo disc swelling. a (left). Small crowded optic disc in a long-sighted patient. b (middle). Optic disc drusen with peripapillary atrophy. c (right). Myelinated nerve fibres superior to the optic disc.

Important symptoms

Patients must be assessed for associated neurological symptoms and signs, in particular any fevers, neck stiffness, severe or new headache, nausea or vomiting. These are all features of meningitis or a sudden rise in intracranial pressure and the patient must be investigated immediately. Any focal neurological deficits, such as double vision, hearing loss or changes in arm and leg strength or sensation must raise concerns regarding a possible space-occupying lesion.

Raised intracranial pressure with accompanying vision loss may indicate severe papilloedema. Patients with both conditions require combined ophthalmic and neurological assessment that is best performed at a teaching hospital. Patients in rural areas should be transferred to a large centre if possible.

All patients with disc swelling are at risk of vision loss without timely and appropriate treatment.

Initial investigations

Office tests

All patients should have their blood pressure measured to check for malignant hypertension. Body temperature measurement is important if meningitis is suspected.

If the patient has not seen an optometrist, visual acuity testing and confrontation visual field testing should help to narrow

the differential diagnosis to one of the two broad categories above.

Blood tests

Any patient over 50 years of age with disc swelling and vision loss must be presumed to have giant cell arteritis until proven otherwise. Therefore, urgent measurement of the erythrocyte sedimentation rate and C-reactive protein level should be ordered. A full blood count to check for thrombocytosis, and liver function tests to check levels of other acute-phase reactants are also helpful.

If giant cell arteritis is considered a possibility and the patient has vision loss, then they should be given oral prednisolone 1 mg/kg immediately (in the office) and the nearest ophthalmologist or hospital with emergency facilities should be contacted to make appropriate follow-up arrangements.

Neuroimaging

Most patients with disc swelling will need some form of neuroimaging. A standard brain MRI without gadolinium is not adequate in any of these scenarios. If, as a GP, you are unable to request an MRI of the brain and orbits with gadolinium it is best to liaise first with the ophthalmologist or neurologist.

The specific scans required include fine-slice axial and coronal fat-suppressed

post-gadolinium views of the orbits. Patients in whom raised intracranial pressure is a concern will also need a contrast-enhanced MR venogram to exclude dural venous sinus thrombosis.

An MR angiogram with contrast would only be required if there was concern about a cerebral vasculitis.

GP review

GP follow up is required for all patients to make sure that these investigations are performed and the referral plan is followed appropriately in a timely manner. An example where timely referral did not occur was a young girl referred to me with disc swelling who had failed to attend an ophthalmologist appointment as requested

FEATURES OF GIANT CELL ARTERITIS

Ophthalmic features

- Transient visual obscurations
- Vision loss
- Double vision (involvement of cranial nerves III, IV or VI)

Systemic features

- New temporal headache
- Jaw claudication
- Scalp tenderness
- Fevers or chills
- Limb girdle muscular pain

TABLE. DISTINGUISHING TRUE FROM PSEUDO DISC SWELLING

True	Pseudo
Blurred disc margin	Vessels visible crossing disc margin
Vessels obscured crossing optic disc margin	Anomalous branching of vessels
Hyperaemia	No microvascular congestion
Loss of spontaneous venous pulsations (note: absent in 20% of the general population)	

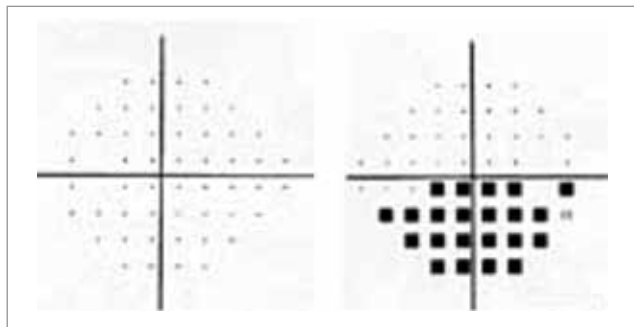


Figure 4. Results of visual field testing showing a right inferior altitudinal defect in a patient with nonarteritic anterior ischaemic optic neuropathy.

six months earlier. She was eventually diagnosed with idiopathic intracranial hypertension and started on treatment, but that six-month delay could have resulted in severe visual field loss.

Referral to a specialist

All patients with previously undiagnosed disc swelling should be seen by an ophthalmologist, or an optometrist who can perform visual acuity and visual field tests and optical coherence tomography. Deciding whether a patient has true or pseudo disc swelling can be a diagnostic challenge, even with all of the equipment available in an ophthalmic practice.

Making a diagnosis

Several key features help distinguish true from pseudo disc swelling, as shown in the Table.

Patterns of vision loss and visual field loss are helpful in making the diagnosis, as noted below.

- Normal central vision with an enlarged blind spot or peripheral field constriction is seen in patients with raised intracranial pressure. The diagnosis is confirmed after an MRI scan with a lumbar puncture to measure opening pressure.
- Optic neuritis typically causes a central loss of vision with colour vision loss out of proportion to the loss of visual acuity. A gadolinium-enhanced MRI of the optic nerves

may show the thickening and inflammation that confirm the diagnosis. Other MRI features suggestive of multiple sclerosis will also help the diagnosis.

- Typically, ischaemic optic neuropathy causes an altitudinal loss of visual field (Figure 4). If giant cell arteritis is suspected then a definitive diagnosis can be made with a temporal artery biopsy. Nonarteritic ischaemic optic neuropathy is typically a clinical diagnosis that can only be confirmed once the disc swelling settles. Treatment options will depend on the underlying cause.

Collaborative care with GPs

Many patients with disc swelling caused by optic neuritis or giant cell arteritis are treated with oral corticosteroids. Collaborative care with a GP is needed, with help monitoring for corticosteroid side effects. Attention to gastrointestinal and bone protection, and metabolic management of weight gain and possible diabetic complications are crucial.

Patients with idiopathic intracranial hypertension secondary to weight gain will need a significant amount of care to help them with ongoing weight-loss strategies, with the aim of helping them lose a minimum of 10% of their starting body weight.

In patients with nonarteritic AION, addressing vascular risk factors such as

hypertension, cholesterol and sleep apnoea is important to reduce the risk of vision loss in the other eye.

Conclusion

The differential diagnosis for optic disc swelling covers a wide range of conditions, from normal variants to ophthalmic and neurological causes. All patients with swollen discs will need some investigation, the first of which can be determined by whether or not the disc swelling occurs with central vision loss. Collaborative care between the GP and eye specialist is needed to arrive at the correct diagnosis and monitor the patient during future treatment. MT

Further reading

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COMPETING INTERESTS: None.



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