

Optic nerve swelling – your survival guide (part 2)

BY ALI YAGAN

In this second article we will discuss bilateral optic nerve swelling, its aetiology, various investigations and possible treatments. We will also discuss various protocols used in the management of suspected optic nerve swelling cases.

Bilateral optic nerve swelling [1,2]

When faced with bilateral optic nerve swelling it is important to differentiate true bilateral optic nerve swelling from unilateral but sequential optic nerve swelling as these will usually have a different aetiology, investigations and treatment options.

Swelling of the optic nerve due to raised intracranial pressure (ICP) (papilloedema)

Causes:

- Idiopathic
- Secondary
 - Brain tumours
 - Trauma
 - Venous thrombosis
 - Medical conditions such as kidney failure
 - Medications such as vitamin A, tetracycline, contraception, lithium.

Case example

- Female 45-years-old
- Routine visit to optician
- Suspected optic nerve swelling
- Normal visual functions
- Subtle optic nerve swelling on OCT
- CT / MRI scan

Idiopathic intracranial hypertension (IIH)

General features: This condition usually affects females in their reproductive years that are usually overweight.

Signs and symptoms: General symptoms include headaches and pulse synchronised tinnitus. Ophthalmic symptoms include transient visual loss and visual obscurations and various visual fields defects. Fundus examination reveals optic nerve swelling which is usually bilateral but could be asymmetrical. Frisen grading is sometimes used in assessing the optic nerve swelling.

Investigations: It is important to take a good history to try to rule out possible secondary causes. It is also necessary to arrange urgent neuro imaging such as CT scan and CT venogram to rule out compressive lesions

and sinus thrombosis. Once these are done, lumbar puncture to assess cerebrospinal fluid (CSF) opening pressure as well as constituents is needed to confirm the diagnosis.

Treatment options: It is important to remember that we are not treating the pressure but treating the patient and protecting their visual functions. Mild cases of IIH could be monitored without any treatment. Other cases are usually treated medically to reduce ICP and first line medication is acetazolamide tablets with a dose dependent on the pressure level, headaches and degree of optic nerve swelling, as well as visual functions. Minority of cases that are refractory to medical treatment or with aggressive vision threatening IIH will need surgical intervention with CSF divergent procedures such as ventriculoperitoneal (VP) shunts or optic nerve fenestration.

Difficulties with IIH diagnosis

These include: getting the wrong diagnosis (not true optic nerve swelling or not IIH) and that will lead to inappropriate invasive

procedures. It is also important to recognise that patients usually have difficulty losing weight.

As most visual function tests are subjective and performance related, it is very important to be accurate and certain before taking any major decisions that can have permanent effects on patient health and vision. Most patients with IIH have several psychosocial issues and a degree of functional visual loss that can affect their performance and assessments.

Refractory headaches are usually an issue in some patients and chronic migraines are a typical feature of patients with long-term raised ICP.

It is also important to remember that various treatment options for IIH will have side-effects and it is important to counsel patients regarding these in order to maintain compliance with treatment. Due to the fact that IIH affects females in their reproductive years, it is very important to inform patients about relationships between IIH and its treatments with pregnancy.

Modified Frisen papilloedema grading [3]

0 (Normal optic disc)

Prominence of the retinal nerve fibre layer at the nasal, superior, and inferior poles in inverse proportion to disc diameter. Radial nerve fibre layer striations, without tortuosity.

1 (Minimal degree of oedema)

C-shaped halo that is subtle and grayish with a temporal gap; obscures underlying retinal details. Disruption of normal radial nerve fibre layer arrangement striations. Temporal disc margin normal.

2 (Low degree of oedema)

Circumferential halo. Elevation (nasal border). No major vessel obscuration.

3 (Moderate degree of oedema)

Obscuration of one segment of major blood vessels leaving disc. Circumferential halo. Elevation (all borders). Halo (irregular outer fringe with finger-like extensions).

4 (Marked degree of oedema)

Total obscuration on the disc of a segment of a major blood vessel on the disc. Elevation (whole nerve head, including the cup). Border obscuration (complete). Halo (complete).

5 (Severe degree of oedema)

Obscuration of all vessels on the disc and leaving the disc.

Diagnostic criteria for IIH:

Papilloedema.

Normal neurological examination except IV, VI, VIII palsies.

Normal brain scan but can see: distended optic nerve sheath, narrowing of transverse sinus, empty sella, slit ventricles, flattening of posterior globe.

Normal CSF constituents but raised pressure more than 25cm of H₂O.

THE MANCHESTER EXPERIENCE**1. The one stop optic nerve head clinic:**

In response to the increased referrals of suspected swollen optic nerves.

Orthoptic led investigations	VA
	CV
	Fields 30-2
	OCT scan
Neuro ophthalmology fellow	Assess results
	Perform optic nerve ultrasound scan
Discuss with consultant	Diagnosis
	Investigations / referral / discharge

Case

- 19-year-old female
- Suffering from headaches
- Seen her optician
- Suspected swollen optic nerve, sent to Emergency Eye Department
- Bilateral optic nerve swelling was noted
- Urgent neuro imaging: Normal CT / CTv
- Sent for LP: raised ICP
- Diagnosed with IIH

2. Protocols:

Optic neuritis	Typical optic neuritis	Explain to patient the condition and possible link to MS
		MRI scan (urgent within two to three weeks) to assess demyelination and refer if positive
		Bloods for Aquaporin 4
		OCT of disc and macula (useful baseline test)
		No treatment generally
	Atypical optic neuritis	Blood for autoimmune and infectious screen
		Neuro imaging
		Consider treatment
AION	Suspect GCA (new onset headaches, over 50, raised ESR and/or CRP, temporal tenderness, weight loss, night sweats, jaw claudication etc.)	Start patient on prednisolone (1 mg/kg) with lansoprazole 30mg o.d. and calcium supplements
		Perform OCT scan of RNFL thickness for baseline
NAION	Book temporal artery biopsy to be done within 2/52 of treatment	
	After biopsy if no contraindications, commence aspirin 75mg o.d.	
	Refer to rheumatology if appropriate	
	Patient is usually elderly with microvascular risk factors	
	Deterioration of visual acuity	
	Typically an altitudinal field defect with RAPD and disc swelling	
	Usually painless (90%)	
	Other risk factors to query:	Obstructive sleep apnoea, phosphodiesterase inhibitors (Viagra), amiodarone
		• Rule out GCA
		- Bloods: BP / ESR / CRP / FBC / glucose / lipids
Bilateral disc swelling	Neuro imaging	
	Perform OCT scan for baseline documentation	
	Suspected raised ICP	
	VA, CV, fields	
	OCT, USS	
	Urgent neuro imaging	CT / CTv
	LP	
	Look for signs of optic nerve functions compromise	
	Treatment if raised ICP: Acetazolamide	

References:

1. Chan J. *Optic Nerve Disorders, Diagnosis and Management*. 2nd edition. Springer-Verlag: Berlin Heidelberg; 2014.
2. Miller N, Newman N. *Walsh and Hoyt Clinical Neuro Ophthalmology*. 6th edition. Lippincott Williams & Wilkins: Philadelphia, USA; 2005.
3. Scott CJ, Kardon RH, Lee AG, et al. Diagnosis and grading of papilledema in patients with raised intracranial pressure using optical coherence tomography vs clinical expert assessment using a clinical staging scale. *Arch Ophthalmol* 2010;**128**(6):705-11.

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