THE SWOLLEN DISC

Valerie Biousse, MD
Emory University School of Medicine
Atlanta, GA

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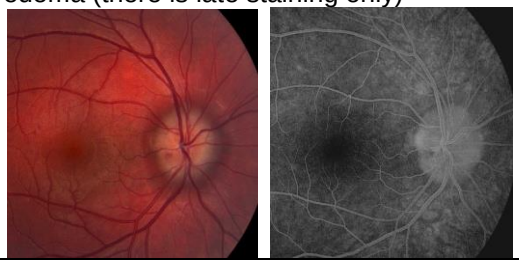
Edema of the optic nerve head (disc edema, swollen disc) is a non-specific term describing localized swelling anterior to the lamina cribrosa.

Mechanisms of optic nerve edema include:
-Local optic nerve injury such as from:
.Inflammation (anterior optic neuritis or papillitis)
.Ischemia (anterior ischemic optic neuropathy)
.Fluctuations in intraocular pressure (high, as in acute glaucoma, or low, as in ocular hypotony)
.Toxicity
-Blockage of retrograde axonal transport from:
.Optic nerve compression (optic nerve tumor or orbital mass)
.Raised intracranial pressure (papilledema)

Differentiating *true optic nerve head edema* from *pseudo-edema* is essential. In most cases, pseudo-disc edema appearance results from a congenital anomaly of the optic nerve and does not require any work-up, while true disc edema is associated with numerous disorders.

Characteristics of true disc edema vs pseudo-disc edema

True disc edema	**Pseudo-disc edema**
-Elevated optic nerve -Margins blurry -Vessels obscured -Venous dilation and tortuosity -± Peripapillary hemorrhages and exudates -Leakage on fluorescein angiogram	-Elevated optic nerve -Sharp margins -Vessels not obscured -Absence of central cup -Anomalous retinal vasculature (arterial branching) -No leakage on fluroscein angiogram
Leakage on fluorescein angiography in true disc edema (late phases) 	No leakage on fluorescein angiography in pseudo disc edema (there is late staining only) 

THE DIFFERENTIAL DIAGNOSIS OF THE "SWOLLEN DISC"

Disc elevation without true swelling:

Optic disc anomalies
- Drusen
- Tilted disc
- Crowded disc
- Myelinated nerve fibers

Optic disc infiltration

Leber hereditary optic neuropathy

True disc swelling:

Elevated intracranial pressure (papilledema)

Inflammatory optic neuropathy
- Demyelinating
- Sarcoidosis or other inflammatory diseases
- Infectious

Neuroretinitis

Vascular optic neuropathy
- Anterior ischemic optic neuropathy
  - Nonarteritic
  - Arteritic
- Diabetic papillopathy
- Central retinal vein occlusion
- Carotid-cavernous fistula
- Malignant systemic hypertension

Compressive optic neuropathy
- Neoplastic
  - Meningioma
  - Hemangioma
  - Lymphangioma
- Non-neoplastic
  - Thyroid ophthalmopathy
  - Orbital inflammatory pseudotumor

Infiltrative optic neuropathy
- Neoplastic
  - Leukemia
  - Lymphoma
  - Glioma
- Non-neoplastic
  - Sarcoidosis

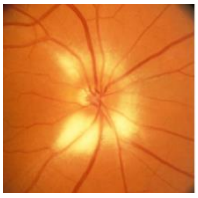
Toxic

Metabolic / Nutritional deficiencies

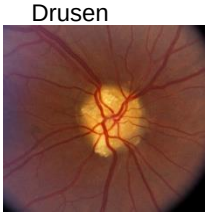
Traumatic optic neuropathy

Intraocular hypotony (low pressure)

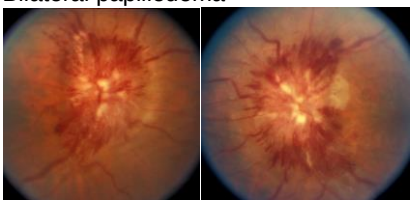
Myelinated nerve Fibers



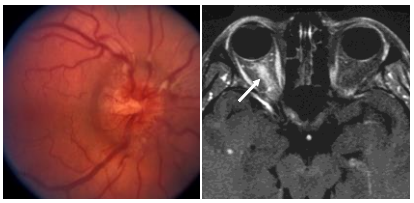
Optic nerve head Drusen



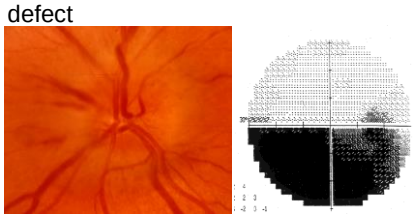
Bilateral papilledema



Right anterior optic neuritis. Optic nerve enhancement on T1-MRI (arrow)

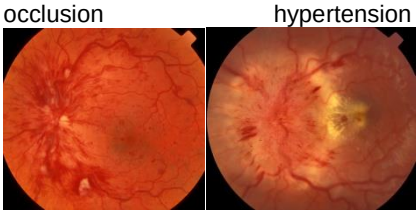


Anterior ischemic optic neuropathy: Disc edema and inferior altitudinal visual field defect

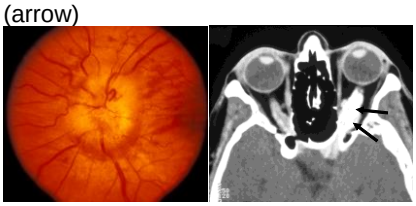


Central retinal vein occlusion

Malignant systemic hypertension



Left optic nerve sheath meningioma with disc edema and enhancement on CT (arrow)



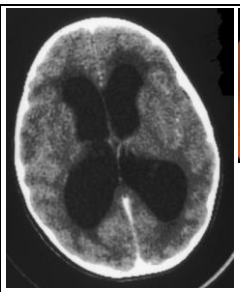
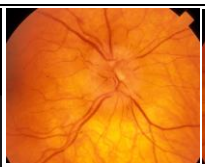
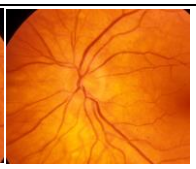
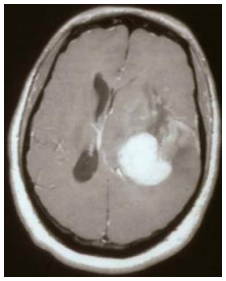
EVALUATION OF THE PATIENT WITH DISC EDEMA:
Once true disc edema is confirmed (see above), it should be determined whether the disc edema is related to an optic nerve disorder or to raised intracranial pressure (papilledema).

Optic neuropathy vs papilledema

Characteristics of disc edema from anterior optic neuropathy vs raised intracranial pressure

Optic neuropathy with disc edema	Papilledema (raised ICP)
-Decreased visual acuity -Decreased color vision -Central, arcuate, or altitudinal visual field defect -Disc edema more often unilateral -Often isolated (or associated with symptoms or signs related to underlying disease)	-Normal visual acuity (until late) -Normal color vision -Enlarged blind spot, nasal defects, constriction of visual fields -Disc edema almost always bilateral -Other symptoms and signs of raised ICP (headache, nausea, diplopia from 6th nerve palsies, pulsatile tinnitus, transient visual obscurations) -Focal neurologic symptoms if focal intracranial process

Mechanisms responsible for raised intracranial pressure:

-Intracranial mass Tumor, abscess Intracerebral hemorrhage Subdural/epidural hemorrhage Large vascular malformation -Hydrocephalus -Meningeal process Infectious Inflammatory Neoplastic -Increased venous pressure -Cerebral venous thrombosis -Idiopathic intracranial hypertension	   

-Most disorders producing raised intracranial pressure are life-threatening emergencies: the finding of papilledema should prompt immediate work-up, ideally in a specialized center with up-to-date neuro-imaging, as well as neurological and opthalmological consultations. Cerebral venous thrombosis is a classic cause of raised intra-cranial pressure. Patients may present with isolated raised ICP (thereby mimicking idiopathic intracranial hypertension). Early recognition may prevent a devastating stroke and visual loss from chronic papilledema

-The work-up of a patients with papilledema should include:
.Looking for an underlying neurological process
.Careful evaluation of the visual function (visual acuity and formal visual field testing), since papilledema can result in permanent visual loss from secondary optic atrophy.
.Blood pressure (severe systemic hypertension (or malignant hypertension) may produce bilateral disc edema that mimics papilledema).

Although papilledema is a reliable sign of raised intracranial pressure, the absence of disc edema does not rule-out an intracranial process in a patient presenting with headache.

Evaluation of a patient with presumed papilledema (raised intracranial pressure):
-Neuro-imaging needs to be obtained emergently to rule-out an intracranial process.
A brain MRI with contrast is the ideal test and is the most sensitive to detect intracranial masses, infiltrative and meningeal processes, as well as cerebral venous thrombosis. A head CT without contrast – which is often the test of choice in the Emergency Room – is in most cases useless in these patients. It is only helpful to detect intracranial hemorrhages, hydrocephalus, and large mass lesions, but does not rule-out any of the other intracranial lesions. Patients with a normal head CT should be investigated further with a brain MRI.
-A normal brain MRI in the setting of papilledema suggests either a meningeal process, venous hypertension, or idiopathic intracranial hypertension as the cause of raised intracranial pressure.
-A lumbar puncture with measurement of the CSF opening pressure and CSF analysis should always be performed.

Visual loss from papilledema:

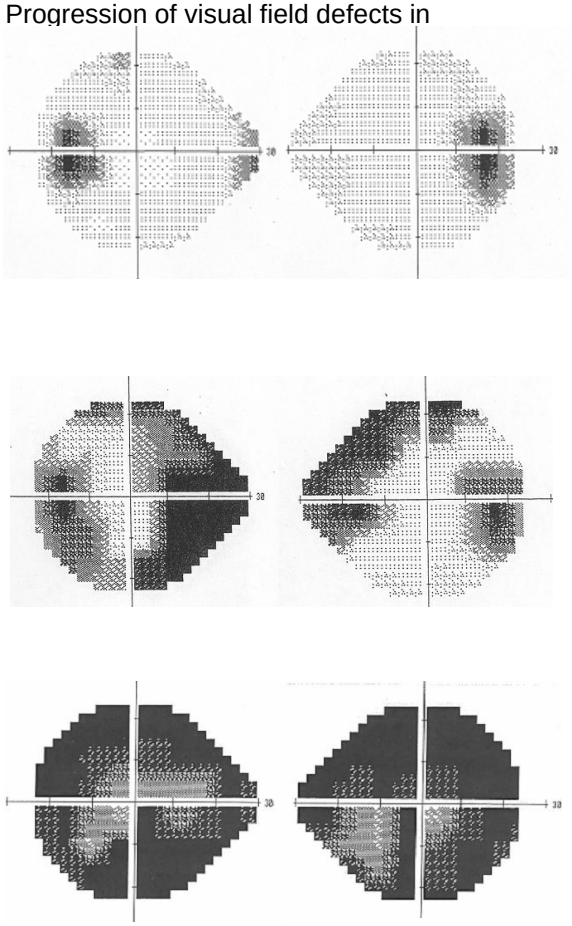
-Patients with papilledema often have no visual symptoms initially.
-They may complain of “flashing lights” or transient visual obscurations (brief episodes of visual loss occurring in one or two eyes, often precipitated by changes in posture such standing up after bending over).
-Untreated chronic papilledema results in visual loss: central visual acuity is normal until late and patients develop insidious progressive visual field constriction.

Visual loss from papilledema happens with any cause of papilledema. Hence, it is very important to look for papilledema in all patients with headache, known hydrocephalus, brain tumor or meningitis, and to prevent visual loss when papilledema is present.

Visual field defects in papilledema

Formal visual field testing (Humphrey perimetry shown here) is often abnormal in papilledema.
-Blind spot enlargement and nasal defects are common initially (top)
-They may progress usually circumferentially to involve the central 30 degrees of visual field (middle).
-Severe devastating visual field loss (bottom) is often permanent if raised intracranial pressure is not promptly treated (note that even with the severe visual field loss seen in the bottom example, visual acuity was still relatively preserved at 20/25 OD and 20/40 OS).

Progression of visual field defects in



Idiopathic intracranial hypertension (IIH) (previously called pseudotumor cerebri):

- Defined as increased intracranial pressure with normal imaging and normal CSF contents.
- By definition, papilledema from a meningeal process or cerebral venous thrombosis should not be classified as IIH.
- Patients with IIH have symptoms and signs of raised intracranial pressure such as headaches, nausea, pulsatile tinnitus, papilledema (and visual loss), and diplopia from unilateral or bilateral sixth nerve palsy.
- The management of the disease is based on the severity of headaches and the presence of visual loss, specifically visual field deficits.

Criteria for the diagnosis of idiopathic intracranial hypertension

1. Signs and symptoms of raised intracranial pressure.
2. No localizing neurologic signs, in an alert patient, other than abducens nerve paresis.
3. Normal neuroimaging studies except for small ventricules or empty sella (neuroimaging should include a good quality MRI +/- MRV or CTV to rule-out cerebral venous thrombosis).
4. Documented increased opening pressure (250 mm of water or more in adults; 280 mm of water or more in children) but normal cerebrospinal fluid composition.
5. Primary structural or systemic causes of elevated intracranial venous sinus pressure excluded (for example, chronic meningitis or cerebral venous thrombosis).

The cause of this disorder is unknown. It involves most often young obese women and may be associated with other factors.

Management of idiopathic intracranial hypertension

- Goal of the management:
  .To relieve the headaches and diplopia
  .To preserve visual function.
- There is a high spontaneous remission rate.
- The lumbar puncture performed as part of the work-up is usually the first step of the treatment as it immediately decreases the intracranial pressure (at least transiently). Headaches that do not improve (at least transiently) after lumbar puncture are unlikely to be entirely the result of raised intracranial pressure.
- Discontinuation or treatment of presumed precipitating factors is recommended, including weight loss, discontinuation of tetracyclines, or vitamin D, treatment of anemia, and of sleep apnea.
- Acetazolamide is classically prescribed at a dose of 500mg to 4 grams a day.
- Topiramate is also often used to control headaches in IIH.
- In rare cases, patients develop rapidly progressive visual loss that requires emergent surgical treatment, sometimes preceded by a lumbar drain. A brief course of intravenous steroids is sometimes helpful in this setting, but steroids should not be prescribed routinely or chronically in IIH (because of weight gain and rebound effect).
- Surgical treatments are necessary in fulminant IIH or in patients worsening despite appropriate medical management.
  CSF shunting procedures (performed by neurosurgeons): derivation of CSF drainage into the peritoneum"
  - Lumbo-peritoneal shunt or ventriculoperitoneal shunt
  - Preferred when headaches are severe
  - Obstruction or disconnection require a revision in about 50% of lumboperitoneal shunts

  Optic nerve sheath fenestration (performed by ophthalmologists): decompression of the optic nerve by making a window into its dural sheath from a transconjunctival medial or lateral approach.
  - Done on the eye with worst visual function first – often needs second eye surgery
  - Preferred when visual loss is predominant and headaches mild
  - The fenestration fails in 1/3 of cases within 3 years

Endovascular venous stenting for transverse sinus stenosis remains debated and should only be used in very selected cases in specialized centers.

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