BENIGN INTRACRANIAL HYPERTENSION

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BENIGN INTRACRANIAL HYPERTENSION

Give clinical presentation, causes, and diagnostic modality and treatment options in benign intracranial hypertension. (2+3+2+3) D2013

Definition

- Benign intracranial hypertension (BIH) is an idiopathic condition associated with increased CSF pressure, normal or small ventricles, normal CSF composition and papilledema, usually bilateral, which may be very severe.
- The basic cause is decreased absorption of CSF.
- The peak incidence is in the 3rd decade of life, especially obese females in the reproductive age group.
- The female to male ratio is about 2:1.
- Ninety percent of patients are women and 90% are obese.
- The disease is rare in prepubertal children (with obesity less a factor) and in lean adults.
- Common field defects include an enlarged blind spot.

Symptoms

- Generalized headache worse in the mornings aggravated by Valsalva's maneuver.
- Transient visual disturbances usually preceded by the headache.
- Headache and nausea are common.
- Diplopia (secondary to abducens nerve paresis),

Signs

- Visual field loss
- Pulsatile tinnitus (pulse synchronous bruit).
- Almost all patients with IIH have papilledema.
- Other neurologic abnormalities other than abducens palsy are not associated with IIH.
- As discussed above for acute clearly visible overlying the disc papilledema, early IIH shows normal visual acuity and enlarged blind spots on perimetry testing.
- Optic nerve function may deteriorate in long-standing, untreated, or severe cases.
- 50 percent of causes occur as isolated conditions and remaining 50 percent are associated with other conditions.

Associations of BIH

a. *Obstruction or impairment of cerebral venous drainage*,

• Tumors,

• Septic thrombi,

• Radical neck dissection.

b. *Endocrine and metabolic dysfunction*

• Elevated estrogen levels

• During pregnancy (especially the 2nd &5th month) there is decrease in levels of adrenal corticoids & increased estrogen

• Hypoparathyroidism and associated hypocalcemia interferes with transport of CSF through arachnoid granulation.

c. *Exogenously administered agents*

• Systemic corticosteroid therapy leads to suppression of the adrenal cortex

• Withdrawal of use from corticosteroids.
•Antibiotics, e.g. tetracycline, nalidixic acid, doxycycline, minocyline
• Retinoic acid, lithium
• Anti-inflammatories like indomethacin, ketoprofen
• Vitamin A 1 lakh units day for few months
• Lead encephalopathy caused cerebral edema and increased ICT.

d. *Systemic illness*

• Meningitis, encephalitis, lead to blockage of the ventricular system.
• Status epilepticus, leads to cerebral hypoxia and cerebral edema.
• Vascular hypertension.
• Thrombocytopenic purpura.
• Chronic respiratory insufficiency, hypercapnia, reduced blood oxygen, polycythemia, elevated venous pressure and ICT.

e. *Familial BIH is also a well-known entity*

Differential diagnosis

- Such conditions may resemble IIH

1. Cerebral venous disorders such as cerebral venous obstruction resulting from
    - Trauma,
    - Childbirth,
    - Hypercoagulable state
    - Middle ear infection
2. Systemic or localized extracranial venous obstruction
    - After radical neck dissection
3. Dural arteriovenous malformation or systemic vasculitis
    - May lead to decreased venous outflow and thus increased ICP.

Investigations and diagnosis

1. suspected IIH should undergo not only neuroimaging with magnetic resonance imaging to rule out
    i. Tumor,
    ii. Hydrocephalus,
    iii. Meningeal lesion.
2. Magnetic resonance venography (MRV) to assess for venous sinus occlusion.
3. Characteristic MRI findings of intracranial hypertension include
    i. Flattening of the globe,
    ii. Enlarged optic nerve sheaths,
    iii. Partially empty sella,
    iv. Narrowing of the distal transverse sinus.
4. Lumbar puncture should always be performed to confirm elevated ICP and to rule out infectious or inflammatory processes.
5. Visual field

Diagnostic criteria

- The original **criteria for BIH by Dandy** in 1937 comprised

1. Clinical evaluation of a patient with signs and symptoms of increased ICP and CSF pressure of more than 25 mm Hg.

2. No localizing signs with the exception of abducens nerve palsy,
3. Normal CSF composition.
4. Normal to slit ventricles with no intracranial mass on imaging by ventriculography.

- This was modified by Smith in 1985 and by Digre and Corbett subsequently in 2001, and the revised criteria were put up as the **modified Dandy criteria** comprising in addition to the initial criteria,

1. Evaluation by computed tomography instead of ventriculography,
2. The presence of an awake and alert patient.
3. Exclusion of venous sinus thrombosis.
4. This was in addition to the requirement that no other cause for the raised ICP be found.

Table 4-3 Criteria for Diagnosis of Idiopathic Intracranial Hypertension

Documented elevated ICP, typically ≥25 cm H_20 in adults during lumbar puncture measured in the lateral decubitus position
No evidence of ventriculomegaly, mass, structural, or vascular lesion on MRI scan or contrast-enhanced CT scan for typical patients, and MRI scan and magnetic resonance venography for all others
No other cause (including medication) of intracranial hypertension identified
Normal CSF composition
Signs representing increased ICP or papilledema
Symptoms, if present, representing increased ICP or papilledema

CSF = cerebrospinal fluid; CT = computed tomography; ICP = intracranial pressure; MRI = magnetic resonance imaging.

Management

➢ **Follow up and examination**

1. The ophthalmologist plays a crucial role in the management of IIH.
2. Careful long-term follow-up is essential to ensure that papilledema resolves.
3. Regularly scheduled examinations should include testing of visual acuity, color vision, and quantitative perimetry to document the level of optic nerve function.
4. Stereophotographs of the optic nerve are essential to obtain during patient follow-up.
5. The frequency of visual field testing depends on the severity of papilledema, the level of optic nerve dysfunction, and the response to treatment.

➢ **Treatment -** It depends on symptomatology and vision status.
➢ The disease may be self-limited.
➢ If headache is controlled with minor analgesics and optic nerve dysfunction is absent, no therapy may be required.
➢ However, the natural history of IIH may result in severe vision loss; 26% of patients in a long-term study eventually had visual acuity worse than 20/200 in at least 1 eye.

➢ **For obese patients**,

1. Weight loss can be an effective treatment and is always recommended, as weight loss alone can lead to resolution of IIH.
2. In some cases, bariatric surgery has been considered.

➢ **For patients requiring medical therapy**,

1. Acetazolamide is usually the first choice;
2. Topiramate has also been used with success. It has multiple beneficial effects—
   - ✓ Headache control,
   - ✓ Appetite suppression,
   - ✓ Carbonic anhydrase inhibition.
3. Furosemide is frequently used in patients intolerant of acetazolamide or topiramate.
4. The use of corticosteroids is controversial.
   - ✓ ICP can improve with use of corticosteroids, recurrence can occur commonly during corticosteroid taper.
   - ✓ Corticosteroid withdrawal is a documented cause of IIH.
   - ✓ A short course of high-dose intravenous corticosteroids may benefit the patient with fulminant papilledema and severe vision loss.

- **Surgical management**
- **In cases of intractable headache or progressive vision loss** despite maximally tolerated medical therapy, surgical therapy is recommended.
- In some patients with severe vision loss and papilledema from markedly elevated ICP, surgical intervention may be considered without waiting for definite evidence of progression.
- The primary surgical options are
  - ✓ Optic nerve sheath fenestration (ONSF)
  - ✓ CSF diversion procedure (lumboperitoneal or ventriculoperitoneal shunt).
- **ONSF**
  - ✓ **In the presence of substantial loss of vision without prominent headache**, ONSF may represent the preferred surgical option because it directly protects the optic nerve and has lower morbidity than that associated with shunting.
  - ✓ ONSF carries a 1%–2% risk of vision loss from optic nerve injury, central retinal artery occlusion (CRAO), or central retinal vein occlusion (CRVO).
  - ✓ It does not lower ICP and thus often does not treat headache.
  - ✓ A reduction of papilledema in the contralateral eye can occur, but bilateral ONSF may be required.
  - ✓ The long-term success rate of ONSF remains unclear.
  - ✓ Repeat ONSF may be performed but is technically more difficult because of scarring.
- **Lumboperitoneal or ventriculoperitoneal shunting**
  - ✓ These procedures effectively lower ICP, with improvement of headache, abducens palsy (if present), and papilledema; moreover, shunting entails no direct risk to the optic nerve.
  - ✓ A shunt, however, may become occluded, infected, or altered in position, requiring reoperation in many cases.
- **Gastric bypass**
  - ✓ Among patients with morbid obesity, gastric bypass surgery can effectively reduce both weight and ICP.

Pediatric IIH

- IIH also occurs in the pediatric population, but the criteria for pediatric IIH remain controversial.
- Although the term pediatric typically refers to children under the age of 18 years, some authors believe it should be reserved for prepubescent children.
- IIH appears to be a different disorder in prepubertal children, with a predilection for boys and nonobese children.

- Unlike in adult IIH, several cranial neuropathies have been associated with pediatric IIH that reverse with lowering of the ICP; these include neuropathies of cranial nerves (CNs) III, IV, VI, VII, IX, and XII. In 2010 and 2011, studies showed that normal opening pressure among children is higher than previously believed and is similar to that in adults.
- Papilledema without headache or visual symptoms is more common in younger patients.
- The treatment for pediatric IIH is similar to that for adult IIH