PSEUDOTUMOR CEREBRI

Background
1. Also known as idiopathic intracranial hypertension (IIH)
2. Unknown etiology
3. Diagnosis of exclusion
4. Primarily found in obese women of childbearing age
5. Chronically elevated intracranial pressure (ICP) and papilledema are primary issues
6. Can lead to blindness if not treated

Pathophysiology
1. Pathophysiology not clear, but current theories suggest increased resistance to cerebrospinal fluid (CSF) outflow, which may produce IIH
2. Incidence (in United States) 0.9 cases per 100,000
3. Female-to-male ratio 8:1
4. Risk Factors: Female sex, reproductive age group, obesity
5. Morbidity: 5-30% of IIH patients will experience permanent vision loss from progressive visual field loss

Diagnostics
1. History
   o Most common complaint is headache (usually generalized and worse in the a.m.)(94% of patients)
   o Visual symptoms (with and without papilledema)
     ▪ Usually preceded by headache and occur in 40-70% of patients
     ▪ Transient disturbances in visual clarity most common
   o 6th nerve palsy only common focal neurological deficit
   o Less common symptoms:
     ▪ pulsatile tinnitus (58%)
     ▪ retrobulbar pain (44%)
     ▪ neck/back/shoulder pain
     ▪ nausea and vomiting
     ▪ dizziness
2. Physical Examination
   o Fundoscopy looking for papilledema
   o Neurological exam
3. Laboratory evaluation
   o CSF studies
   o CBC
   o BUN/Cr
   o TSH
   o Serum iron and TIBC
   o Thrombophilia screen
4. Diagnostic imaging (primary role is to exclude other conditions)
   o MRI (rule out meningeal infiltration and/or dural venous sinus thrombosis)
   ▪ If present, flattening of posterior globe is the only sign that strongly suggests pseudotumor cerebri

Pseudotumor Cerebri
5. Lumbar puncture
   - Performed in lateral decubitus position
   - For diagnosis of IIH, CSF opening pressure must be >250mm of water

6. Diagnosis of IIH requires 3 components to be satisfied
   - Normal brain imaging
   - Raised CSF pressure with normal CSF constituents
   - Exclusion of other causes of raised CSF pressure

Differential Diagnosis

1. Key Differential Diagnoses:
   - Dural venous sinus thrombosis
   - Hydrocephalus
   - Cerebral mass lesions
   - Hypertensive encephalopathy
   - Optic disc anomalies
   - Intracranial hypertension secondary to medications and/or systemic diseases

2. Systemic Diseases associated with intracranial hypertension:
   - SLE
   - Behcet’s disease
   - Uremia
   - Iron deficiency anemia
   - Addison's disease
   - Hypothyroidism
   - Polycystic ovarian disease

3. Medications associated with intracranial hypertension:
   - Tetracyclines
   - Nalidixic acid
   - Fluoroquinolones
   - Oral contraceptive pills
   - Danazol
   - Progesterone
   - Lithium
   - Vitamin A, isotretinoin
   - Sulfamethoxazole
   - Steroid withdrawal
   - Growth hormone

Therapeutics

1. Management focuses on preservation of visual function and symptom relief
2. Serial lumbar punctures (relieves increased ICP)
   - Temporary relief; not a chronic treatment plan
3. Mild or No Visual Loss
   - Weight loss and sodium restriction
     - Resolution of papilledema and reduced ICP reported with as little as 6% weight loss
     - Consider dietician consult
   - Acetazolamide (reduces CSF production by decreasing sodium ion transport across choroidal epithelium)
     - Dosing: Start 250mg BID; increase slowly to maintenance dose of 1000-2000mg daily
   - Furosemide (20mg BID to 40mg TID) or topiramate can be used as alternative
   - Corticosteroids no longer recommended due to side effects; may be used preoperatively prior to shunting

4. Moderate to Severe Visual Loss
   - Some recommend early surgery vs. medical trial
   - Maximum medical therapy
     - 1 gram acetazolamide/day with gradual increase to maximum tolerated dose
     - Can consider adding furosemide TID
   - Proceed to surgical management
     - Optic nerve sheath fenestration (ONSF) to improve papilledema and headaches
     - Shunt (LP/VP) to drain excess CSF and decrease ICP

5. Persistent Headaches
   - Only 50% relieved with surgery; analgesic and caffeine rebound may coexist
   - Trial of standard prophylactic vascular headache remedies
     - Avoid hypotensive causing agents (beta or calcium channel blockers)
     - Tricyclic antidepressants at low dose (may cause weight gain)
     - Topiramate

6. Children
   - Incidence same in boys and girls up to puberty
   - Obesity not a significant contributor to pathology or treatment
   - Repeat lumbar punctures discouraged
   - Acetazolamide 15mg/kg divided BID or TID until headache, disc swelling and visual symptoms abate (usually 3-9 months)
   - Furosemide, topiramate, zonisamide are alternatives
   - Acute vision loss - intravenous acetazolamide and methylprednisolone can be used pending surgical evaluation

7. Pregnancy
   - Same treatment as non-pregnant women with the exception of more modest weight loss recommendations
   - Acetazolamide in the first trimester potentially teratogenic (Category C); is used only after appropriate informed consent
     - Consider high-risk obstetrics consult prior to starting in first trimester
Follow-Up

1. Best to be followed up by Neurology and Ophthalmology jointly
   - Newly diagnosed and those with significant visual impairment every 2-4 weeks until stabilized
   - Stabilized patients every 3-6 months
   - Assessments should include:
     - Visual acuity
     - Color vision
     - Visual fields
     - Optic disc exam with photography
     - CSF opening pressure unreliable as measure (can use if symptomatic without visual field defects or papilledema)

2. Admit to Hospital
   - Rarely necessary
   - Admissions usually due to intractable headache or for surgical interventions

Prognosis

1. Most important factor is vision loss
2. Risk factors associated with worse outcomes:
   - Male gender
   - African American race
   - Morbid obesity
   - Anemia
   - Obstructive sleep apnea
   - Acute onset of symptoms plus signs of raised intracranial hypertension
3. 5-30% of patients will have permanent significant visual field loss
4. Small 10-year follow up study revealed 55% of patients with papilledema remained stable; 45% worsened
5. One study showed 96% of patients with IIH had some vision loss
   - 60% improved with treatment, while 10% deteriorated, over an average of 12 months

Prevention

1. Weight loss if obese

Patient Education


References


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