

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 age¹
5. Chronically elevated intracranial pressure (ICP) and papilledema are primary issues¹
6. Can lead to blindness if not treated¹

Pathophysiology

1. Pathophysiology not clear, but current theories suggest increased resistance to cerebrospinal fluid (CSF) outflow, which may produce IIH^{2,3}
2. Incidence (in United States) 0.9 cases per 100,000⁴
3. Female-to-male ratio 8:1⁴
4. Risk Factors: Female sex, reproductive age group, obesity⁴
5. Morbidity: 5-30% of IIH patients will experience permanent vision loss from progressive visual field loss^{4,5}

Diagnostics

1. History⁴
 - Most common complaint is headache (usually generalized and worse in the a.m.)(94% of patients)
 - Visual symptoms (with and without papilledema)
 - Usually preceded by headache and occur in 40-70% of patients
 - Transient disturbances in visual clarity most common
 - 6th nerve palsy only common focal neurological deficit
 - Less common symptoms:
 - pulsatile tinnitus (58%)
 - retrobulbar pain (44%)
 - neck/back/shoulder pain
 - nausea and vomiting
 - dizziness
2. Physical Examination
 - Fundoscopy looking for papilledema
 - Neurological exam
3. Laboratory evaluation⁴
 - CSF studies
 - CBC
 - BUN/Cr
 - TSH
 - Serum iron and TIBC
 - Thrombophilia screen
4. Diagnostic imaging (primary role is to exclude other conditions)^{2,6}
 - 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 cerebri⁶

- MR Venography (rule out dural venous sinus thrombosis)
 - CT scan (rule out large tumors/brain lesions)
 - Ultrasound (identify intracranial hypertension by measuring diameter of optic nerve sheath; >5mm correlated with intracranial pressure greater than 20 cm H₂O)⁷
5. Lumbar puncture⁴
 - 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 satisfied⁴
 - Normal brain imaging
 - Raised CSF pressure with normal CSF constituents
 - Exclusion of other causes of raised CSF pressure

Differential Diagnosis⁴

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^{4,8,9,10}

1. Management focuses on preservation of visual function and symptom relief
2. Serial lumbar punctures (relieves increased ICP)⁴
 - Temporary relief; not a chronic treatment plan

3. Mild or No Visual Loss⁴
 - 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 Loss⁴
 - 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 Headaches⁴
 - 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. Children⁸
 - 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. Pregnancy⁹
 - 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-Up⁴

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 outcomes¹⁰:
 - 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 loss⁵
4. Small 10-year follow up study revealed 55% of patients with papilledema remained stable; 45% worsened¹¹
5. One study showed 96% of patients with IIH had some vision loss¹²
 - 60% improved with treatment, while 10% deteriorated, over an average of 12 months

Prevention

1. Weight loss if obese

Patient Education

1. <http://www.ninds.nih.gov/disorders/pseudotumorcerebri/pseudotumorcerebri.htm>

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