### Intracranial Hypertension, Idiopathic

**Alternative Names**
- IIH
- Pseudotumor Cerebri
- Benign Intracranial Hypertension
- BIH

**Record Category**
- Disease phenotype

**WHO-ICD**
- Diseases of the nervous system > Other disorders of the nervous system

**Incidence per 100,000 Live Births**
- 2-5

**OMIM Number**
- 243200

**Mode of Inheritance**
- Autosomal recessive

**Gene Map Locus**
- N/A

**Description**
Idiopathic Intracranial Hypertension, as the name suggests, is a condition characterized by an abnormal elevation in the intracranial pressure, in the absence of any expansive lesion or ventricular dilation. Since the signs and symptoms of the condition are very similar to that of brain tumor, the disease is also known as pseudotumor cerebri (PC). Most of the symptoms are non-specific, such as headache, nausea, transient visual obstructions and double vision, rhythmic sound in the ears (pulsatile tinnitus), lethargy, mood change and dizziness. The primary hallmark of IIH is bilateral, asymmetric, or unilateral papilloedema, caused by atrophy of the optic nerve. If left untreated, the papilloedema may result in blindness. The pathophysiology of the disease is unclear. However, it is proposed that increased resistance to absorption of CSF across the arachnoid villi may be the causative factor of the increased fluid pressure.

This disease predominantly affects obese women of child bearing age, or reaching menarche. Diagnosis involves imaging techniques like CT and MRI scans of the brain, along with orbital ultrasound. The primary aim of treatment is to reduce the intracranial pressure. Diuretics and hyperosmotic drugs are administered to promote loss of fluid. Due to the condition’s relationship with obesity, weight loss is a necessary part of the treatment strategy. A majority of the patients respond to medication within three to five months, failing which surgical intervention is considered. Surgery involves therapeutic shunting, wherein the spinal fluid is made to drain into the abdominal cavity via a tube.

**Molecular Genetics**
A genetic predisposition to the disease is likely; especially considering the reports of familial IIH. However, the gene or the locus responsible for the condition has not been identified.

**Epidemiology in the Arab World**

**Jordan**
Ahmed and Shaaban (1989) described the case of a 33-year old Jordanian male patient, who presented with bilateral papilloedema, blurring of vision, and a three –month history of headache. The patient’s visual acuity was 6/9 and CSF pressure more than 40cms of water. CT scan of the brain was normal, except for slit like lateral ventricles. A large polyp in the floor of the left maxillary antrum was seen in the radiographs of the paranasal sinuses. Even though lumbar puncture was performed on the patient eight times to remove the fluid, the papilloedema and headache persisted, and the CSF pressure remained persistently high. Medication (dexamethasone, and acetazolamide) also did not seem to have any effect on the patient’s condition. Since intra-ventricular
tube insertion was difficult in the presence of slit like ventricles, a James lumbar-peritoneal shunt was inserted at L2/L3 level in the subarachnoid space to drain the fluid. Following the surgery, the patient’s CSF pressure came down to normal, and the headaches and papilloedema resolved within two weeks.

**Libya**

Radhakrishnan et al. (1993) conducted an epidemiologic survey of idiopathic intracranial hypertension (IIH) in Benghazi, Libya, over a period from September 1982 through August 1989. The group was comprised 81 patients (76 females and 5 males). Ages ranged from 8 to 55 years. The average crude annual incidence rates for IIH per 100,000 persons were 2.2 for the total and 4.3 for females for all ages. In females aged 15-44 years, IIH occurred at a rate of 12.0 per 100,000 per year; for those defined as obese, the rate rose to 21.4. Moderate to severe visual loss occurred as a sequela in 20% of the patients. The extent of visual loss did not correlate with age at diagnosis, duration of symptoms, degree of obesity, use of oral contraceptive pills, cerebrospinal fluid (CSF) opening pressure, steroid treatment, or recurrence. Radhakrishnan et al. (1993) found no correlation between CSF protein and opening pressure. Radhakrishnan et al. (1993) also conducted a case-control study on 40 consecutive female incident IIH patients and 80 age-matched female control subjects. Obesity and recent weight gain occurred more frequently in patients. More patients were married and more had irregular menses.

**Oman**

Koul et al. (1994) reported two female patients, who presented with headache and vomiting. Case 1 (nine years old) had her symptoms along with giddiness for a year with gradual onset. Case 2 had her symptoms for 20 days with the headache being mostly occipital and of maximum intensity in the morning. There was no history of drug ingestion or any illnesses in both cases. Their weights were both below the 10th percentile. Case 1 had bilateral papilloedema with visual aquity of 6/6 (right eye) and 6/9 (left eye). CT scan of the brain showed evidence of brain edema. Blood counts, immunological, and endocrinological investigations were normal. Electroencephalogram (EEG) and visual evoked potentials (VEP) were normal. Examination of case 2 showed little thyroid swelling and bilateral papilloedema with normal visual aquity and ocular movements. Routine blood, immunological, and endocrinological investigations as well as VEP were found to be normal. Her brain CT scan showed edema with no mass lesion and no obstruction to the CSF flow. Both patients were diagnosed with idiopathic intracranial hypertension, and were started on acetazolamide (for both patients), as well as a loop diuretic (only for case 1). Three months later, the patients’ symptoms were found to improve. Koul et al (1994) emphasized the importance of considering the diagnosis of pseudotumor cerebri in children presenting with headache, vomiting, and features of increased intracranial pressure. In 2002, Koul et al. described an 11-year-old girl who presented with headache of 3 months’ duration. There was bilateral disc edema. The cerebrospinal fluid pressure was 50 cm of water with normal cerebrospinal fluid cytology and biochemistry. She developed severe headache (different and disabling), dizziness, vomiting, and backache on sitting up 6 hours after lumbar puncture, and lying supine relieved all of her symptoms. Intravenous fluids, analgesics, and complete bed rest did not relieve her symptoms over the next 72 hours. Magnetic resonance imaging (MRI) done 96 hours after lumbar puncture revealed the entire dura overlying the brain, including the posterior fossa, showing intense enhancement on contrast injection with leak at the lumbar puncture site. Oral caffeine (coffee, three times a day) was advised over 1 week. The patient remained asymptomatic, and a repeat MRI scan after 10 days showed complete clearing of the cerebrospinal fluid leak with no dural enhancement.

Knox-Macaulay et al. (2002) reported pseudotumor cerebri in a young Omani female diagnosed with acute promyelocytic leukemia (APL), before starting treatment for her malignancy. The 16-year old patient had had a history of episodic attacks of headache 10-years ago, which worsened five days before her hospital admission. There was no previous or family history of neurological or ophthalmic disorder or history of drug ingestion. The patient showed no evidence of obesity, or any other disease associated with pseudotumor cerebri. Examination revealed fever, ecchymoses on limbs and back, hepatomegaly, but no splenomegaly or lymphadenopathy. She was found to have a visual aquity of 6/60 in both eyes, severe bilateral papilloedema and fundal hemorrhages on ophthalmic examination. Chest X-ray and CT scan of the brain and both orbits were normal with normal sized ventricles, no space occupying lesion, and no intraorbital or intracranial bleeding. The diagnosis of pseudotumour cerebri was made on the basis of her symptoms, ophthalmic evaluation and normal CT
scan and confirmed by CSF examination (elevated CSF opening pressure of 420 mmH2O with normal proteins and glucose concentration and no cells). Her management included ATRA therapy for a total of 60 days, three cycles of combination cytotoxic consolidation chemotherapy, followed by maintenance chemotherapy. Her headache and right eye visual aquity (vision in left eye remained poor) improved with no evidence of papilloedema.

**United Arab Emirates**

A 25-year old Arab woman, who presented with headache, visual obscuration and dizziness, one month after delivering a full term baby, was examined by Ahmed and Shaaban (1989). The patient was found to be obese, and had bilateral papilloedema. An enlarged pituitary fossa was visualized in her skull radiographs. Multiple lumbar punctures were performed on the patient. However, both the punctures as well as the medication administered failed to alleviate the symptoms. Surgery was performed to insert a James lumbar-peritoneal shunt via the lumbar route. Both the papilloedema and the headache were found to have been resolved. [See also: Jordan > Ahmed and Shaaban, 1989].

**References**


**Related CTGA Records**

Leukemia Acute Myeloid

**External Links**

http://www.emedicine.com/neuro/topic329.htm
http://www.ihrfoundation.org/
http://www.pseudotumorcerebri.com/

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