Pseudotumor Cerebri: A Review*

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ABSTRACT

Pseudotumor cerebri (PTC) is a syndrome seen more frequently in young, obese women defined by increased intracranial pressure without obstruction of the ventricular system. Normal neurodiagnostic studies, normal cerebrospinal fluid (CSF) contents, papilledema in one or both eyes, and normal mental status should also be recognized. Although the disorder has been associated with drugs, endocrinopathies, compromised nutritional status and other disease states, no underlying disorder is uniformly present.

Introduction

The syndrome complex of increased intracranial pressure, normal cerebrospinal fluid (CSF) contents, normal brain and ventricular size, and clinical signs of increased intracranial pressure (papilledema) has been referred to by many names. "Idiopathic intracranial hypertension," coined by Quincke, was the first term used to describe this syndrome. In 1904, the term "pseudotumor cerebri" was suggested by Nönné. Later, the description "benign intracranial hypertension" was proposed by Foley in 1955.

Clinical Features

The symptoms of pseudotumor cerebri are quite variable, the most common complaints being headache and visual problems. Headaches, reportedly seen in 94 percent of reported cases, are nonspecific and variable in presentation. They are usually worse in the morning and increased with the Valsalva maneuver. Aching neck, arms, and back have also been reported as presenting symptoms. Nausea is another frequently seen symptom.

Visual disturbances are also common. Transient visual obstructions (TVO) are seconds of visual blackouts thought to be secondary to hypertension in the visual pathway caused by changes in intracranial pressure. These TVO are not predictive of permanent visual loss, and they seem to revolve with or without reduction in CSF pressure. Horizontal diplopia may occur secondary to increased pressure on the sixth cranial nerve. Visual field defects and decreased visual acuity occur in half of the patients. Papilledema is usually present; however,
cases without papilledema have been described. Hemorrhages and exudates around the disc may also be seen. Retrobulbar pain with eye movement has been described.

**Etiology and Associations**

The etiology of PTC is undetermined. Many theories try to explain intracranial hypertension. Interstitial and/or intracellular edema of the brain parenchyma has been reported on brain biopsy. While the etiology of this change is unclear, it may prevent ventriculomegaly.

Another theory involves CSF hypersecretion. Donaldson suggested PTC may be induced by elevated estrone levels, with the estrone inducing a hypersecretion of CSF. As estrone is released by adipocytes, estrone levels are increased in obese patients commonly seen with this disease. This theory could also explain the association of PTC with patients taking oral contraceptives and girls undergoing menarche. Critics of this theory feel some of the patients may have cerebral sinus thrombosis, a well known complication of oral contraceptives and pregnancy.

A genetic predisposition to this disease has been proposed. Both autosomal dominant and recessive modes of inheritance have been suggested. Individuals with the proposed genetic defects were all obese, raising the question of an hormonal etiology as well. In children, mastoiditis, otitis media, head injury, and lateral sinus thrombosis may precipitate PTC. Endocrine conditions including hypoparathyroidism, Addison’s disease, hypothyroidism with thyroid hormone replacement, pituitary neoplasms, and anabolic steroids have all been associated with PTC.

While there are many drugs implicated in PTC, much of the evidence is unsubstantiated. Certain classes of drugs appear to be more frequently associated with PTC including nalidixic acid, tetracyclines, vitamin A, danazol, steroids, ethinylestradiol, thyroid hormone, and lithium. The list, however, does not lend itself well to common mechanism of disease induction. Nutritional deficiencies of vitamins A and D, while rare, have been described. Additionally, malnourished children can develop PTC upon being renourished. Iron deficiency (anemia) may fall into this category as well; however, it is an unusual cause of PTC. Autoimmune diseases have been implicated. There are a multitude of other disease associations, none of which seem terribly convincing.

Despite the lengthy list of potentially causative agents (table I), the only entity with a statistically significant association with PTC has been obesity.

**Comment**

This diagnosis of PTC is one of exclusion. Computerized tomography scanning or magnetic resonance imaging (MRI) of the head should be performed initially to rule out intracranial mass lesions. Only if these studies are normal may a lumbar puncture be performed to diagnose increased intracranial pressure. An opening pressure greater than 200 mm of water is diagnostic in non-obese patients, and greater than 250 mm of water in obese patients. The CSF fluid studies will be normal. Cerebral spinal fluid protein has been reported to be lower with more increased CSF pressures; however, repeat studies did not confirm this. Currently, there are no definitive CSF proteins that are specific for diagnosing PTC. Some investigators
who further confuse the criteria for diagnosing PTC suggest the inclusion of diseases with structural defects, or include diseases in which no increase in intracranial pressure is documented. This further complicates establishing firm diagnostic criteria.

Treatment of PTC involves evaluating the patient for a causative agent. Removal of offending drugs or resuming steroids that have been withdrawn while treating another condition should be done. Treating a patient's obesity or underlying disease condition is also helpful. A majority of patients require nonspecific means for controlling intracranial pressures. Multiple modes of therapy have been used, including repeated therapeutic lumbar punctures, dexamethasone, carbonic anhydrase inhibitors, and furosemide. In patients refractory to medical management, optic nerve sheath fenestration and lumboperitoneal shunting have been performed. Surgery should be considered with continued reduction in visual acuity and visual fields.2

References


15. **Peng, F.:** Verbal Communication PTC. April, 1993.

