Malignant pseudotumor cerebri

Report of two cases

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Two young Arab women presented with a very rapid loss of vision, ophthalmoplegia, florid papilledema, areflexia of the lower limbs, and normal mentation. Lumbar puncture pressure was above 60 cm H₂O, but no intracranial structural lesion was found in either patient. An exhaustive evaluation as to an etiology was negative in both. Under continuous lumbar cerebrospinal fluid drainage and administration of steroids, furosemide, and acetazolamide, both patients had significantly improved vision and ocular movement. In both, lumboperitoneal shunting was considered but only one eventually underwent this procedure. These two patients with pseudotumor cerebri are unique in their fulminant clinical course and severely increased intracranial pressure. Virtually inevitable blindness was prevented by timely intervention.

KEY WORDS • blindness • lumboperitoneal shunt • ophthalmoplegia • pseudotumor cerebri • intracranial pressure

Pseudotumor cerebri is a widely described "benign" entity of obscure etiology, involving increased intracranial pressure (ICP). The patients are usually obese young women complaining of headaches, blurred vision, transient visual obscurations, and diplopia. Typically, papilledema and abducens palsies are the only neurological deficits found, and elevated ICP up to 40 cm H₂O is the only positive auxiliary examination. Although the etiology of pseudotumor cerebri is unknown, many conditions have been associated with this entity: vitamin metabolism disorders, drug reactions, venous drainage obstruction, endocrine dysfunctions, and hematological disorders are among the more prominent. The usual spontaneous resolution of this disorder, leaving few sequelae, has been the source of the adjective "benign" in its description.

Two patients with pseudotumor cerebri are presented whose unique and fulminant clinical course merits the definition "malignant" pseudotumor cerebri.

Case Reports

Case 1

This 18-year-old obese unmarried Arab woman was hospitalized due to repeated vomiting, dizziness, and an apparent single grand mal seizure with ensuing total loss of vision, all on the day of admission. She showed symptoms typical of a viral upper respiratory tract infection 10 days prior to admission.

Examination. The patient was fully alert, had marked neck stiffness, total blindness, florid papilledema with multiple retinal hemorrhages, complete ophthalmoplegia on the right, and global ophthalmoparesis on the left with bilaterally dilated, nonreactive pupils. The corneal reflexes were diminished bilaterally. The lower-limb tendon reflexes were absent with intact muscle strength. The rest of the neurological and general physical examination was noncontributory.

The erythrocyte sedimentation rate was 50 mm/hr, the peripheral white blood count 11,900/cu mm, and hemoglobin 10.5 gm%, with hypochromic microcytic indices; the platelet count and coagulation profile were normal. The serum biochemical profile was normal. Computerized tomography (CT) and magnetic resonance imaging did not demonstrate any intracranial pathology. Four-vessel cranial angiography was normal, except for a very slow vascular passage time, appropriate for severely increased ICP, without any evidence of venous obstruction. Lumbar puncture yielded clear fluid that spurted out of the top of a glass column 60 cm in height. The cerebrospinal fluid (CSF) contained 600 erythrocytes/cu mm without white cells, 20 mg% protein, and 85 mg% glucose; it was sterile and negative serologically for cryptococci and viruses. The electroencephalogram was normal, and no visual evoked
potentials were elicited over the occipital cortex. An extensive search was made for vasculitis, other inflammatory disorders, an infectious pathogen, a neoplasm, endocrinopathology, or a metabolic derangement but was negative (except for hyperlipidemia type II). There was no evidence supporting exposure to toxins or drugs. The human lymphocyte antigen type was A30 (W19) B49(21) CW7. The patient was not pregnant.

**Course.** The treatment that was initiated included dexamethasone 16 mg/day, furosemide up to 120 mg/day, and acetazolamide up to 1.5 gm/day. Several days later lumbar puncture still yielded excessive pressure, and therefore a continuous lumbar drainage of CSF was begun, draining approximately 300 cc of fluid daily. Under this therapy both vision and ocular movements improved, and after 1 week the lumbar catheter was removed. During the following days the patient suffered from increasing dizziness, and 17 days following hospitalization she developed bilateral facial palsy. Repeated lumbar punctures demonstrated a tendency for pressure to increase up to 33 cm H2O. At the same time, the development of medication side effects, including hepatic function disturbances, hypokalemia, and clinical arthropathy, necessitated the cessation of all medication. The combination of clinical deterioration and medical side effects dictated the implantation of a lumboperitoneal shunt. During the ensuing weeks the ocular movements returned to normal, facial paresis disappeared, and vision improved significantly.

**Case 2**

This 26-year-old obese Arab mother of three was hospitalized due to increasing headache and loss of vision a few days following a spontaneous abortion.

**Examination.** On admission, the patient was fully conscious, blind, had neck stiffness, prominent global bilateral ophthalmoplegia, bilateral florid papilledema, and hyporeflexia of the lower limbs. The rest of the physical examination was unremarkable.

The complete blood count and serum biochemical studies were normal. Computerized tomography of the head and four-vessel cerebral angiography were normal except for a slow vascular passage time. The venous system was well demonstrated without any signs of venous obstruction. Lumbar puncture pressure exceeded 60 cm H2O. The CSF was xanthochromic with microscopic fields full of crenated erythrocytes, 60 mg% protein, and 55 mg% glucose. The fluid was serologically negative and sterile. The electroencephalogram showed diffuse nonspecific slowing. An extensive general laboratory evaluation as in Case 1 yielded negative results.

**Course.** Medication similar to that administered in Case 1 was initiated. Three days following hospitalization transient unilateral peripheral facial paresis appeared. Following failure to decrease the lumbar puncture pressure, continuous lumbar drainage was begun. The ophthalmoparesis gradually resolved, the papilledema regressed, and vision improved to finger counting at 3 m. The lumbar drainage was stopped after 1 week. One month later, while still under medication, the patient again complained of blurred vision and a repeat lumbar puncture elicited a pressure of 34 cm H2O. A lumboperitoneal shunt was contemplated, but was not placed for technical reasons. The patient was lost to follow-up review.

**Discussion**

These two patients had remarkably similar presentations, consisting of acute severe loss of vision, ophthalmoplegia involving the third, fourth, and sixth cranial nerves, nonreactive pupils, florid papilledema, and normal mentation. Both patients had ICP's exceeding 60 cm H2O with no etiology being found in CT, angiography, or magnetic resonance imaging of the brain. Extensive additional laboratory studies were noncontributory. These findings direct us to a diagnosis of pseudotumor cerebri. Additional supportive evidence of this diagnosis are the patients' age, female sex, and one patient having just undergone an abortion. The outstanding features of both patients that are not typical of pseudotumor cerebri are the acute visual loss with virtual blindness, ophthalmoplegia, and facial nerve paresis. Unusual cases of pseudotumor cerebri were described by Rush et al. and Corbett, et al., with what was considered a rapid loss of vision: a significant decrease in visual acuity within 1 week. McCammon, et al.,11 and Snyder and Frenkel described oculomotor paresis, and Halpern and Gordon reported trochlear paresis in pseudotumor cerebri. Abducens paresis is a well-known feature of this condition. Facial nerve paresis has also been reported, usually in children. Although obviously uncharacteristic, all of these features have been associated with pseudotumor cerebri and do not negate this diagnosis. To our knowledge, the occurrence of all of these features in a single patient has not been described previously.

Both patients had decreased lower limb reflexes verging on areflexia. This finding, combined with increased ICP and ophthalmoplegia, raises the possibility of the variant of acute idiopathic polyneuropathy described by Fisher. The normal CSF protein level and normal neuromuscular electrodiagnostic evaluation preempt this diagnosis. The hyporeflexia may possibly be related to the patients' obesity.

The above considerations establish the diagnosis of pseudotumor cerebri in these two patients, although their clinical course was unusual in its fulminant evolution and combination of deficits. It is evident that their outcome would have been catastrophic without intervention, and the adjective "benign" is certainly inappropriate for these two patients with increased ICP. Sklar has also noted that the clinical course of many patients with pseudotumor cerebri can hardly be pronounced "benign" due to a significant loss of vision. For these unusual patients we propose the term "malignant" pseudotumor cerebri.
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References


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