

Paradoxical Transtentorial Herniation in a Patient with Pseudotumor Cerebri

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Pseudotumor cerebri, or idiopathic intracranial hypertension, is a disorder characterized by symptoms and signs of a space-occupying intracranial mass such as headache, transient visual obscurations, and papilledema, without evidence of a mass or ventricular obstruction. Lumbar puncture reveals elevated intracranial pressure. Cerebrospinal fluid components are normal [1]. The treatment of pseudotumor cerebri is aimed at reducing intracranial pressure to prevent visual loss and headaches.

Medical treatment includes carbonic anhydrase inhibitors, loop diuretics, steroids and digitalis. Surgical treatment is required in patients unresponsive to medication. Surgical options include optic nerve sheath fenestration, lumboperitoneal shunt, ventriculoperitoneal shunt, and subtemporal decompression (subtemporal craniectomy) [1]. Subtemporal decompression is used when fenestration or shunting have failed. We report the first reported case of pseudotumor cerebri in which subtemporal decompression performed after lumboperitoneal shunt resulted in transtentorial herniation.

Patient Description

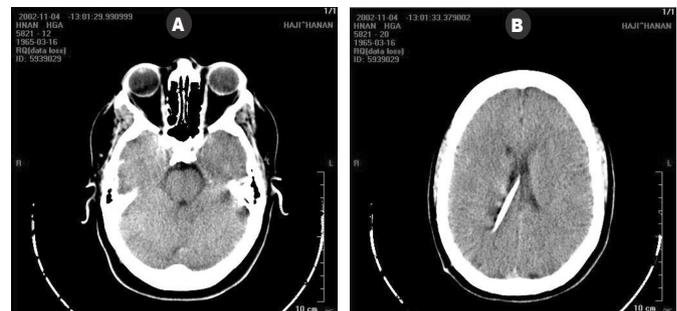
This 37 year old female patient was diagnosed with pseudotumor cerebri in 1998. She had a typical history of severe headaches and progressive visual loss. Having failed medical treatment and optic nerve sheath fenestration, she was referred for lumboperitoneal shunting. She underwent several shunt revisions due to

obstruction and/or migration of the catheter. A right ventriculoperitoneal shunt was inserted due to persistent symptomatic disease, although further deterioration of vision, which included an enlargement of the blind spot of the right eye, was demonstrated at her previous neuro-ophthalmologic follow-up visit, and it was the reason for her repeat admission. The patient complained of headaches and occasional visual obscurations.

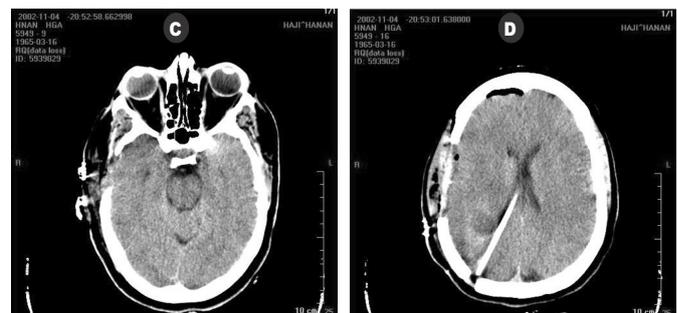
On examination, she was morbidly obese and fully alert. Her neurologic examination was normal. Attempts at lumbar puncture failed to retrieve CSF. The decision was taken to proceed with a right subtemporal decompression [Figure A-D]. Forty-eight hours following the procedure the patient complained of severe, not positional, headaches and became

CSF = cerebrospinal fluid

progressively drowsy. On examination she was obtunded but arousable; her right pupil was dilated and sluggishly reactive to light. Computerized tomography of the head demonstrated transtentorial herniation with a midline shift from right to left [Figure E,F]. She was treated with mannitol and furosemide for 36 hours with little improvement in her clinical condition. Repeat CT scan did not show major changes. The patient was taken to the operating room for ligation of the distal



[A,B] Axial CT of head before sub-temporal decompression demonstrates small ventricles and no midline shift.



[C,D] Axial CT of head immediately after sub-temporal decompression

catheter of the lumboperitoneal shunt. Following this procedure she gradually recovered full consciousness within a few hours. Her pupils became equal and reactive to light. Postoperative CT scan demonstrated complete resolution of the transtentorial herniation and the midline shift [Figure G,H]. A neuro-ophthalmologic examination before discharge and at follow-up 2.5 years later demonstrated no change and no further deterioration.

Comment

Surgical intervention in patients with pseudotumor cerebri is aimed at preventing visual loss or occasionally at reducing intractable headaches, when more conservative measures failed. Surgery is also indicated for patients in whom visual field studies are unreliable, or for those non-compliant with medication or regular follow-up [1].

In many centers, optic nerve sheath fenestration is the preferred method for patients with progressive visual loss in whom medical therapy is ineffective [1]. Lumboperitoneal shunt is the preferred surgical procedure for patients with intractable headache and for those with progressive visual loss unresponsive to optic nerve sheath fenestration [2,3].

Sub-temporal decompression is a surgical technique that has been used in recent years as a procedure of last resort to protect or to retrieve vision, and/or to manage intractable headache when more common surgical procedures have failed [1,4]. According to this rationale, sub-temporal decompression was performed in our patient after repeat failures of ventriculo- and lumboperitoneal shunts. Following the procedure a clinical and radiographic transtentorial herniation was observed.

This seemingly "paradoxical" transtentorial herniation should be induced from the physical changes resulting from sub-temporal decompression in the hydrodynamics of CSF. The tentorium divides the intracranial space into the supra- and infra-tentorial compartments, which in normal conditions freely communicate. The hydrostatic pressure of cerebrospinal

fluid allows brain buoyancy within these compartments. Craniectomy exposes the supratentorial compartment to atmospheric pressure. Paradoxical transtentorial herniation is possible in pseudotumor cerebri patients following sub-temporal decompression due to a negative gradient between the atmospheric pressure and the intracranial pressure, which is enhanced by the negative pressure created by lumboperitoneal drainage of CSF by a patent lumboperitoneal shunt.

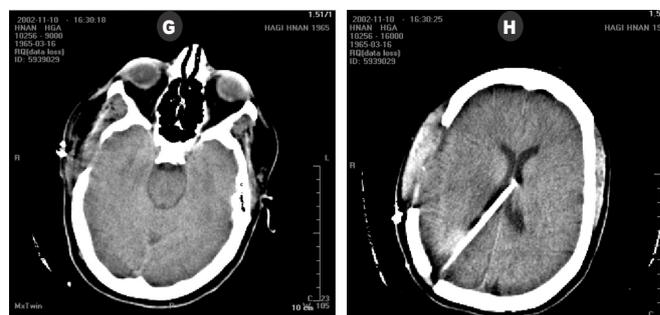
Clinical and radiographic signs of paradoxical transtentorial herniation have been reported shortly after either diagnostic or therapeutic lumbar puncture [5] in patients with craniectomy, but none of them had pseudotumor cerebri. On the other hand, patients with this disorder [4] who were treated by sub-temporal decompression followed by lumbar CSF diversion did not suffer transtentorial herniation.

Ligation of the lumboperitoneal shunt catheter, as performed in our case, is the obvious therapeutic measure to reverse transtentorial herniation. Its removal is probably less advisable, since it could lead to a protracted CSF leak in an already compromised patient.

Clinicians have to take into account that a pseudotumor cerebri patient with this shunt may experience life-threatening paradoxical transtentorial herniation following sub-temporal decompression. Therefore, it is suggested that a simultaneous ligation of the lumboperitoneal shunt catheter be done



[E,F] Axial CT of head 48 hours after sub-temporal decompression shows right-to-left transtentorial herniation and midline shift.



[G,H] Axial CT of head following the ligation of the distal catheter of the lumboperitoneal shunt demonstrates complete resolution of the transtentorial herniation and midline shift.

at the time of surgery for sub-temporal decompression.

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