Empty sella syndrome as complication of benign intracranial hypertension

Leon A. Weisberg, M.D., Edgar M. Housepian, M.D., and David P. Saur, M.D.

Department of Neurology and Neurosurgery, Neurological Institute of the Columbia Presbyterian Medical Center, New York, New York

An empty sella was demonstrated on air study in five patients with the benign intracranial hypertension (BIH) syndrome. All patients had a protracted course and very high cerebrospinal fluid pressure; two required a shunt procedure. No patient had any endocrine symptoms or visual field defects but an air study was done to exclude a mass lesion in the sella region. Among the last 50 patients seen with the BIH syndrome, there were five cases of an associated empty sella (10%). In these cases, the empty sella is a probable consequence of the long-standing intracranial hypertension associated with a congenital deficiency of the diaphragma sellae.

Key Words • intracranial hypertension syndrome • empty sella • CSF pressure

The benign intracranial hypertension syndrome (BIH) is characterized by headache and papilledema, and usually occurs in obese young women whose cerebrospinal fluid (CSF) is normal in composition except for elevated pressure, and in whom an air study demonstrates a normal ventricular system. The course is usually self-limited but complications include visual sequelae and recurrences. This report describes the development of the empty sella syndrome in patients with BIH and the causal relationship between the two conditions in these cases.

Case Reports

Case 1

A 38-year old man had symptoms of headache, visual blurring, and bilateral papilledema; otherwise his neurological evaluation, including arteriography and air study, were normal. The symptoms and papilledema resolved after repeated lumbar punctures. Ten years later he developed spontaneous, nontraumatic rhinorrhea. A skull x-ray film showed an enlarged sella turcica with destruction of the sella floor. Air study showed intrasellar air with the pituitary gland flattened posteriorly and inferiorly, compatible with a diagnosis of an empty sella syndrome (Fig. 1).

To eliminate the CSF leak a right frontal craniotomy was done, at which time two small encephaloceles in the roof of the right orbit were found and repaired. Extensive arachnoid adhesions in the sella region were lysed and an empty sella was found; it was packed with muscle and covered with pericranium. One week after operation the rhinorrhea had stopped, but the patient now complained of headache. Papilledema was...
L. A. Weisberg, E. M. Housepian and D. P. Saur

found, as well as CSF pressure greater than 500 mm H₂O with a protein content of 18 mg %, Treatment with repeated lumbar punctures and steroids failed to lower CSF pressure, and a shunt procedure was done.

After this procedure the patient became symptom-free, the papilledema subsided, and CSF pressure returned to normal. There has been no recurrence in 3 years of follow-up, and skull films show no change in the size of the enlarged sella.

Case 2

A 38-year-old obese woman had had headaches for 6 months, but no visual symptoms. During an evaluation for infertility, bilateral papilledema was noted. Menstrual history was normal and the patient had no other endocrine symptoms. A skull film showed an enlarged sella with a double floor; CSF pressure was 500 mm H₂O with normal protein. Air study showed an empty sella. Treatment with steroids for 7 days was effective. A follow-up skull film 1 year later showed no change in sella size or shape.

Case 3

A 45-year-old obese woman had had headache and visual blurring for 15 months. On examination she was found to have papilledema. A skull film showed an enlarged sella with erosion of the dorsum. Initial CSF pressure was 400 mm H₂O, and the range on three other lumbar taps was 360 to 420 mm H₂O; the CSF protein was normal. Air study showed an empty sella. Medical therapy was not effective in lowering the CSF pressure and a shunt procedure was done. Follow-up skull films showed remineralization of the dorsum but no change in sella size.

Case 4

A 24-year-old obese woman had symptoms of headache for 4 years, but no visual symptoms. On admission she was noted to have papilledema without other neurological signs. Skull x-ray showed an enlarged sella and erosion of the lamina dura. Air study showed an empty sella (Fig. 2). CSF pressure was initially elevated to 400 mm H₂O, and in five subsequent lumbar punctures CSF pressure ranged from 380 to 440 mm H₂O. Treatment with steroids and repeated lumbar punctures was ineffective, and the patient is still symptomatic with headaches and elevated CSF pressure.

Case 5

A 30-year-old obese woman complained of headache and visual blurring of 10 months' duration. She was found to have bilateral papilledema. A skull x-ray film showed an enlarged sella. Initial CSF opening pressure was 450 mm H₂O which fell to 400 mm H₂O after 5 minutes; the CSF protein was normal. Air study showed an empty sella. Treatment with steroids was effective and the patient was symptom-free in 7 days.

Discussion

Kaufman 6 described three patients with an elevated CSF pressure who were found to have an empty sella on air study. In their review of the empty sella syndrome Neelon, et al., 7 documented elevated CSF pressure in one patient, and reported three others with a prior diagnosis of BIH. Foley and Posner 1 reviewed 116 cases of the empty sella syndrome and found that the CSF pressure was elevated in 16 of 48 cases in which it had been recorded. These authors suggest that the elevated CSF pressure in the presence of a congenitally deficient diaphragma sellae leads to the transmission of CSF pulsation waves and herniation of the subarachnoid cisterns into the sella region. This causes remodeling and enlargement of the sella with subsequent displacement of the pituitary gland posteriorly in the sella floor.

In most cases of BIH, duration of symptoms is less than 3 months, but in 20% the duration was more protracted. 8 Because of the short course, radiographic evidence of elevated CSF pressure in the sella turcica is usually absent, and abnormalities are more frequently seen when the intracranial hypertension has been present for longer than 6 months. 9 Most cases of BIH spontaneously remit or respond to treatment with repeated lumbar punctures or steroids, but occasionally intracranial hypertension is resistant to this therapy. It is in cases with a protracted duration of symptoms or resistance to medical treatment that the empty sella may develop.

All of these five patients with BIH and an associated empty sella had a prolonged period of significantly elevated CSF pressure. A
Empty sella syndrome in intracranial hypertension

causal association between the two conditions is most dramatically seen in the first case. This patient developed BIH, treated effectively with repeated lumbar punctures. The patient developed an external communication to the sella and olfactory region, by means of which the CSF pressure reduced. Several years later spontaneous rhinorrhea developed. The rhinorrhea was treated surgically, and after the decompressive CSF pathway was eliminated, the CSF pressure increased and headache and papilledema recurred. This was refractory to repeated lumbar taps and steroids and required a shunt procedure. In the other cases it is suggested that the protracted intracranial hypertension caused a remodeling and enlargement of the sella with subsequent compression of the pituitary gland.

No endocrine symptoms or visual field defects were noted in these five patients. One patient was evaluated for infertility but all of the female patients had a normal menstrual history, with hypopituitarism unlikely. Endocrine studies, including urinary ketosteroids and ketogenic steroids, diurnal plasma cortisol, luteinizing and follicle stimulating hormone, prolactin, growth hormone, thyroxine, IV ACTH test, and urine specific gravity, were normal in all patients. Growth hormone and cortisol response to insulin-induced hypoglycemia and vasopressin stimulation was done in three patients, and all showed an adequate response. In the empty sella syndrome the pituitary gland is flattened against the posterior wall of an enlarged sella, but endocrine function is usually preserved, as was the case in the patients reported in this study. In these patients with headache, papilledema, and a large sella, despite the paucity of endocrine symptoms or visual field defects, an air study should be done to exclude a mass lesion.

Busch estimated that 22% of the normal population have a diaphragma sellae that is reduced to a thin rim of tissue with a significant opening to allow the subarachnoid space to extend into the sella. If this deficient diaphragm plus intracranial hypertension were all that was required to produce an empty sella, this association should occur in 22% of cases of BIH. A review of the last 50 cases of BIH seen at The Neurological Institute since the first case was noted showed only five cases (10%) of an empty sella. Therefore, other factors must be implicated which may include the protracted course and the severity of the pressure elevation, as well as obesity and arterial hypertension.

References


Address reprint requests to: Leon A. Weisberg, M.D., Department of Neurology, 1430 Tulane Avenue, New Orleans, Louisiana 70112.