A dynamic pressure study of spontaneous CSF rhinorrhea in the empty sella syndrome

Case report

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The authors report a case of cerebrospinal fluid (CSF) rhinorrhea in a patient with the empty sella syndrome in whom continuous intracranial pressure (ICP) monitoring revealed intermittent peaks of raised ICP. Following a shunt procedure, ICP reverted to normal and the CSF rhinorrhea ceased for 10 weeks. The sella turcica was packed with muscle after the CSF rhinorrhea recurred.

KEY WORDS • empty sella syndrome • intracranial pressure • lumboperitoneal shunt • cerebrospinal fluid rhinorrhea

SPONTANEOUS cerebrospinal fluid (CSF) rhinorrhea is a recognized sequela of the primary empty sella syndrome. The mechanisms of spontaneous CSF rhinorrhea are varied, and reported cases include patients in whom intracranial pressure (ICP) was either normal or elevated. Raised ICP is probably related to the pathogenesis of the primary empty sella syndrome (ESS). We report a case of primary ESS with spontaneous CSF rhinorrhea, where the use of continuous ICP monitoring revealed a high baseline pressure with intermittent peaks. A lumboperitoneal shunt resulted in the establishment of normal pressure and temporary resolution of the rhinorrhea. Continuous ICP studies on patients with spontaneous CSF rhinorrhea have not been previously reported.

Case Report

This 67-year-old obese woman was referred for investigation of the cause of 2 months of constant headache. Neurological and general examination was normal. Plain skull film revealed an enlarged, globular sella. A computerized tomography (CT) brain scan showed normal-sized ventricles, but also an enlarged sella with density consistent with that of CSF. Lumbar puncture revealed mildly elevated CSF pressure of 230 mm H₂O with normal fluid analysis. A pneumoencephalogram showed air entering the anterior and midportions of the sella, and the pituitary gland flattened against the dorsum and posterior portion of the floor of the sella (Fig. 1). This confirmed the diagnosis of primary ESS. Pituitary function tests revealed some mild anterior pituitary dysfunction. This was not clinically significant. During follow-up examination 3 months after presentation, she reported the onset of an intermittent, clear nasal discharge in addition to her persistent headache.

Lumbar puncture was performed with intrathecal injection of radioactive contrast material (technetium diethylene triamine pentaacetic acid). Serial studies over the subsequent 30 hours demonstrated CSF rhinorrhea, and this was supported by radioactive counts of nasal swabs.

Continuous ICP monitoring with the use of a hollow dural screw attached to a transducer revealed a generally raised baseline pressure with intermittent high peaks (Fig. 2 left). On the basis of the elevated ICP and its presumed causal relationship with the ESS and CSF rhinorrhea, it was decided to perform a lumboperitoneal shunt as an initial procedure, rather than a direct intracranial attack on the fistula itself. A lumboperitoneal shunt was performed at the L-4 level using a medium-pressure lower-end valve. The closing pressure of the valve was 90 mm of saline.

No CSF rhinorrhea was observed in the postoperative period. Intracranial pressure was again measured by the above technique with demonstration...
Fic. 1. Pneumoencephalogram showing air entering the sella.

of normal baseline pressure and elimination of peaks (Fig. 2 right). The patient was discharged from hospital on penicillin V, 250 mg daily, and was initially free of CSF rhinorrhea.

After 10 weeks there was a recurrence of CSF rhinorrhea. She was readmitted to hospital and the sella was explored through a right subfrontal craniotomy. At surgery, an empty sella was found. A fold of arachnoid was found to be extending down into and occupying the sella. A hole was present at the junction of the floor with the anterior wall of the sella. This was packed with crushed muscle and covered with fascia lata. She has subsequently been followed for 8 months and has had no further CSF rhinorrhea.

Discussion

Nontraumatic (spontaneous) CSF rhinorrhea occurring in the empty sella syndrome was first documented by Ommaya, et al., in 1968. Subsequent reports of the association were reviewed by Jordan, et al., in 1977, when he found that 24 cases of spontaneous CSF rhinorrhea occurred in 247 cases (9.7%) of the ESS. Ommaya, et al., classified the mechanisms of nontraumatic CSF rhinorrhea into high-pressure and normal-pressure leaks. His two patients with the ESS apparently had normal ICP.

Kaufman reported two cases of spontaneous rhinorrhea with the ESS and demonstrated persistent craniopharyngeal canals at tomography in both cases. He postulated that the combination of the two anatomical conditions explained the spontaneous fistula. Other authors have speculated on the relevance of raised ICP in the pathogenesis of spontaneous CSF rhinorrhea associated with the ESS. It seems likely that the primary ESS is caused by a combination of a congenitally deficient diaphragma sellae and chronically elevated CSF pressure.

Foley and Posner found that 65% of 43 patients with documented raised ICP measurement in the ESS had elevated CSF pressure also. They surmised that the headache, occasional visual and endocrine abnormalities, and sellar remodeling were all sequelae of pulsatile intrasellar CSF at raised ICP. Bone erosion of the sella floor with subsequent fistula and CSF rhinorrhea is probably another sequela to this combination of diaphragmatic incompetence and elevated ICP.

The ICP monitoring performed in our patient is of interest in that the initial lumbar puncture manometry had revealed only a marginally elevated CSF pressure of 230 mm H2O. However, continuous intracranial monitoring revealed peaks of considerably raised ICP (Fig. 2 left). It is possible that some cases of ESS and CSF rhinorrhea with apparently normal pressure might have had intermittent elevations that could only
be detected with continuous monitoring. The importance of continuous versus static measurement of the ICP has been shown in "normal-pressure hydrocephalus" where intermittent high-pressure peaks have been demonstrated. Most of the previously reported patients have been treated either expectantly with spontaneous cessation of CSF leakage, or by surgical obliteration of the abnormal fistula, usually by packing of the sella turcica with muscle. Greenblatt and Wilson reported two cases of spontaneous CSF rhinorrhea treated with lumbar-peritoneal shunting, where the site of the leak could not be found. The restoration of normal ICP in our patient by means of lumbar-peritoneal shunting resulted in the temporary cessation of the CSF rhinorrhea. However, recurrence of the CSF rhinorrhea necessitated surgical exploration of the sella turcica and obliteration of the hole.

The probable pathogenesis of a CSF fistula through the sella turcica in the ESS could be explained by raised ICP being transmitted through an incompetent diaphragm sellae and causing a remodeling of the floor of the sella. This case demonstrates that, in spite of reducing the ICP to normal, CSF rhinorrhea may still occur through a patent fistula. The effects of long-standing raised ICP, albeit intermittent, resulted in a structural abnormality that was not reversed when the ICP was reduced.

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References


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