Spontaneous Cerebrospinal Fluid Rhinorrhea as a Presenting Symptom of Aqueductal Stenosis
—Case Report—

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Abstract

A 30-year-old male patient presented with chronic spontaneous cerebrospinal fluid (CSF) rhinorrhea. He had sustained a mild head injury in childhood. Magnetic resonance imaging of the brain showed aqueductal stenosis associated with moderate supratentorial hydrocephalus, and erosion of the cribriform plate. Following insertion of ventriculoperitoneal shunt, the CSF rhinorrhea completely ceased and no direct repair of the CSF fistula was necessary. Long-standing spontaneous CSF rhinorrhea indicates the possibility of concurrent intracranial pathology, such as aqueductal stenosis.

Key words: aqueductal stenosis, cerebrospinal fluid, hydrocephalus, rhinorrhea

Introduction

Aqueductal stenosis usually manifests in infancy or early adulthood with features suggestive of raised intracranial pressure syndrome. Aqueductal stenosis in infancy usually manifests as failure to thrive and/or bulging fontanelle, whereas in childhood or adulthood manifests as typical raised intracranial pressure syndrome. Cerebrospinal fluid (CSF) rhinorrhea is rare. Spontaneous CSF rhinorrhea is uncommon as a primary presenting feature of aqueductal stenosis, with only nine reported cases.3,10,11,13,14,19,28–30 The treatment of these cases has varied widely. We report an adult case of aqueductal stenosis manifesting as spontaneous CSF rhinorrhea probably originating from dehiscence of the cribriform plate following head trauma in childhood.

Case Report

A 30-year-old male laborer presented with profuse watery discharge from the right nostril persisting for 9 months. The leakage was initially intermittent and was accentuated by coughing and bending forwards. The discharge was more profuse in the morning when arising from bed. The discharge became almost continuous throughout the day and night for 15 days, associated with moderate holocranial headaches. There was no fever, malaise, or history suggestive of tuberculosis. He suffered no vomiting, visual obscurations, convulsions, or unconsciousness. The patient reported blunt trauma over the forehead with a cricket ball, but without neurological sequelae. On examination, he was conscious and oriented. His higher mental functions were normal. Fundus examination revealed bilateral papilledema. There were no motor, sensory, or cerebellar signs, and no signs of meningitis.

Biochemical analysis of the nasal discharge was consistent with CSF. Computed tomography (CT) and magnetic resonance (MR) imaging of the brain showed evidence of aqueductal stenosis with enlarged lateral and third ventricles (Figs. 1 and 2). The fourth ventricle was disproportionately small. The cribriform plate was eroded and was probably the site of the CSF leakage. In addition, ‘empty sella’ was present with herniation of the dilated infrachiasmatic pouch of the third ventricle into the sella. Contrast-enhanced and diffusion-weighted MR imaging showed no evidence of a tumor. A moderate-pressure ventriculoperitoneal shunt system was inserted. The CSF rhinorrhea ceased completely after the surgery. The patient was asymptomatic and free of CSF leakage after one year.
Discussion

CSF rhinorrhea was proposed in 1853 to be an excremental liquid expressed from different parts of the brain into the ventricles and then excreted through the ethmoid sinuses and hypophysis into the nose as mucus. Spontaneous CSF rhinorrhea was first reported in 1826 in a boy with progressively enlarging head. Autopsy revealed a large internal hydrocephalus with a fistula between the cranial cavity and the nose. CSF rhinorrhea commonly occurs following head trauma or as a result of intracranial surgery. Less common causes include infection of the paranasal sinuses along with osteomyelitis of the adjacent bone, and congenital anomalies of the brain and its coverings such as meningoceles or meningoencephaloceles. Destructive lesions along the skull base can also produce CSF rhinorrhea. Pituitary tumors cause erosion of the sella turcica floor and are frequently associated with spontaneous CSF rhinorrhea. Several benign and malignant intracranial tumors have also been reported to cause CSF rhinorrhea.

Spontaneous CSF rhinorrhea is leakage of fluid from the subarachnoid space to the frontal, sphenoidal, or ethmoidal sinuses. “Indirect or hypertensive spontaneous rhinorrhea” suggests that CSF fistula is possible only if hydrocephalus coexists with a congenital abnormality of the cranial base. Spontaneous CSF rhinorrhea associated with aqueductal stenosis can occur in various ways. The cribriform plate is an anatomically fragile portion of the anterior cranial fossa base, vulnerable to the normal rhythmic variations in the CSF pressure. This relatively ‘weak site’ can allow transmission of the CSF from the intracranial region to the nasal cavity, presumably related to constitutional or developmental factors. Anatomical studies of embryological variations in the cribriform plate and the traversing olfactory fibers have identified incompletely occluded holes around the olfactory fibers in the cribriform plate, persistent cranial pharyngeal canal, fistula in the retained embryonic lumen of the olfactory bulb, and meningeal dysplasia in the region of the olfactory nerve. Any cause of suddenly increased intracranial pressure, such as sneezing or coughing, could force open a previously occult anatomical defect and allow CSF leakage.

Progressive symptomatic hydrocephalus in young adults usually occurs following obstruction of the ventricular system by a space-occupying lesion. In our case, chronic dilation of the lateral and third ventricles was unusually secondary to aqueductal stenosis. Contrast-enhanced and diffusion-weighted MR imaging did not reveal any tumor in the supracerebellar cistern. Long-standing persistent or intermittent elevated intracranial pressure secondary to the aqueductal stenosis could have primarily resulted in the thinning and erosion of the floor of the anterior cranial fossa. The associated presence of ‘empty sella’ and herniation of a large CSF-containing arachnoid pouch into the sella in our case suggested the unusual nature of dissipation of the raised intracranial pressure.

Spontaneous CSF rhinorrhea may act as a natural vent or protective mechanism to prevent dangerous elevation of intracranial pressure, but may also delay the diagnosis. Occasionally, the symptoms of...
raised intracranial pressure and the CSF rhinorrhea may develop simultaneously. Metrizamide CT and MR cisternography can precisely delineate the site of the CSF leakage. The communication between the subarachnoid spaces and the paranasal sinuses can be depicted vividly and is immensely helpful in planning for surgery.

Direct communication between the lateral ventricle and the frontal sinus may cause CSF rhinorrhea in aqueductal stenosis. A 10-year-old girl suddenly developed massive CSF rhinorrhea following severe edema of the left side of her face. The spontaneous CSF leakage was unusual and occurred through the middle fossa and the abnormally enlarged sphenoid sinus. Exploration found an irregular bony defect in the anterolateral floor of the middle fossa. The dura was also perforated and the brain tissue including the temporal horn protruded through the bony defect into the sphenoid sinus. Three cases of aqueductal stenosis have been reported manifesting as a fistulous communication between the protruding frontal lobe and the defect in the cribriform plate. A case of aqueductal stenosis presented with spontaneous CSF rhinorrhea associated with a empty sella syndrome. Three cases of spontaneous CSF rhinorrhea associated with aqueductal stenosis were managed by insertion of a ventriculoperitoneal shunt and resulted in intraventricular tension pneumocephalus. The effect of chronically raised intraventricular pressure was evident in the frontal porencephalic cyst leading to a fistulous communication with a frontal air sinus and required surgical exploration. The long-standing hydrocephalus and the intermittent valve occlusion appear to have contributed to the disruption of the base of the skull and CSF leakage. The chain of events that led to spontaneous CSF rhinorrhea in these cases was due to a fistulous communication between the brain parenchyma and the ventricular system with the paranasal sinuses, which subsequently required surgical exploration.

Chronic persistent hydrocephalus may induce secondary dilation and obstruction of the subarachnoid spaces. Formation of a subarachnoid-nasal CSF fistula resulting in cranial decompression was reported in a patient with raised intracranial pressure. A fistulous tract or multiple fistulae connecting the ventricular system with the paranasal sinuses may be present with the potential hazard of meningitis. The possibility of latero-sellar fistula of the temporal fossa adjacent to empty sella turcica or passing through a lamina cribrosa associated with primary empty sella also needs to be ascertained. There may be separate defects within the anterior and middle cranial fossae. Head trauma frequently results in a dehiscence of the anterior cranial base in the region of the cribriform plate, which is a relatively common site of CSF fistula. The previous head trauma in our patient could have been a predisposing factor for spontaneous CSF rhinorrhea although this was not confirmed. However, there was no fistulous communication demonstrable between the brain and the paranasal sinuses. This probably was responsible for the immediate and complete cessation of spontaneous CSF rhinorrhea following the ventriculoperitoneal shunt insertion and so surgical exploration could be avoided.

The management of spontaneous high-pressure CSF rhinorrhea is directed towards the primary pathology. Performance of a CSF diversion procedure only may be sufficient. CSF fistula secondary to aqueductal stenosis may be less likely to heal spontaneously for unknown reasons. A supplementary procedure to seal off the dural opening may be required even if the CSF rhinorrhea has stopped, since there may be an atretic, nonfunctioning tract that still serves as a potential pathway for the passage of infection into the intracranial cavity. Surgical repair of the CSF fistula is mandatory if there is persistent CSF rhinorrhea following the CSF diversion procedure, and is also warranted in patients who develop tension pneumocephalus after ventriculoperitoneal shunting due to the negative sump effect which may cause malfunction of the shunt. Multi-layered reconstruction of the anterior cranial fossa using frontal muscle flap, pericranial and temporalis muscle-fascia based flap, or a free vascularized omental flap is necessary. Endoscopic repair of the CSF fistula has been successful with a low complication rate. In our patient, the insertion of ventriculoperitoneal shunt stopped the CSF rhinorrhea completely and permanently and hence no direct repair of the fistula was necessary.

Spontaneous CSF rhinorrhea is rare as a primary presenting symptom of aqueductal stenosis. Long-standing spontaneous CSF rhinorrhea indicates the possibility of a concurrent intracranial pathology. The diagnosis of aqueductal stenosis, although uncommon, should be considered in such a situation.

References

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