Spontaneous cerebrospinal fluid rhinorrhea through clival defect

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ABSTRACT

Spontaneous cerebrospinal fluid fistula secondary to clival defect is a very rare condition. Only 2 cases are reported previously in the literature. We report a case of clival defect in a young male patient presenting with spontaneous cerebrospinal fluid rhinorrhea which failed detection by conventional CT and MRI. The patient was referred to our center after failed anterior cranial fossa repair. Thorough radiological investigations successfully detected the defect in the posterior wall of the sphenoid sinus 5 mm distal to the posterior limit of the floor of the sella just on the midline. The fistula was managed successfully through an endonasal transsphenoidal approach. We present the case reviewing the literature concerning the pathophysiology of spontaneous cerebrospinal fluid fistula through the sphenoid sinus.

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Spontaneous CSF fistula constitutes a diagnostic and therapeutic challenge to both radiologists, who should determine the location of the defect, and to neurosurgeons, who must successfully manage the condition to avoid the serious complications of meningitis. Our aim is to emphasize that CSF rhinorrhea should be approached meticulously in cooperation with radiologists to localize the defect precisely before surgery, keeping in mind that rare causes such as a clival defect, can be the source of CSF fistula.

Case Report. Our patient is a 36-year-old male, who was referred from Yemen with 4 months history of excessive rhinorrhea that failed to stop after anterior cranial fossa repair of a presumed cribiform plate defect. The patient presented in Yemen with spontaneous sudden CSF rhinorrhea not preceded by trauma or upper respiratory tract infection. He did not show any sign of meningitis. Conventional brain CT and MRI revealed air fluid level in the sphenoid sinus along with large bifrontal aeroceles communicating with the lateral ventricles. No boney defects could be demonstrated at that time and he was referred to our centre after failed surgery. Examination of the patient revealed excessive CSF rhinorrhea with postural change accompanied by severe headache. No signs of meningitis were detected. Reviewing the CT and MRI we noticed huge bifrontal aeroceles (Figure 1), and small aeroceles posterior to the upper third of the clivus with no apparent boney defect (Figure 2). The CT cisternography with bone window could demonstrate the contrast material leaking into the sphenoid sinus through a defect in the upper third of the clivus (Figure 3). Three dimensional CT reconstructions could clearly show a midline clival defect 5 mm below the cellar floor (Figure 4). Endonasal transsphenoidal approach was performed, and we visualized the CSF leak coming from a defect in the posterior wall of the sphenoid sinus through a dural hole, which also showed the pulsation of the basilar artery. We packed the sphenoid sinus with gel foam and fat from the anterior abdominal wall, bilateral frontal external ventricular drains were inserted for 10 days under...
antibiotic cover to divert the CSF and release the aeroceles. The CSF fistula stopped and a brain CT after one month of surgery showed complete packing of the sphenoid sinus, the frontal aeroceles decreased dramatically, and the prepontine aerocele disappeared.

Discussion. The underlying defect responsible for CSF leak, despite the cause, is due to disruption in the arachnoids and dura mater coupled with a boney defect which can be traumatic or spontaneous, and a CSF pressure gradient acting continuously or intermittently on the disrupted tissues. Most CSF rhinorrhea is traumatic in 90% of cases. It is reported in 2% of head injuries. Non-traumatic CSF rhinorrhea occurs less frequently and is of 2 variants, the first is spontaneous CSF rhinorrhea with normal CSF pressure in 3% of cases and usually due to congenital boney defect. The second is associated with continuous or intermittent increase in CSF pressure in 7% of cases as in primary empty sella syndrome. Davis and Kaye documented a high base line CSF pressure with intermittent peaks in patients with empty sella syndrome.

Early in fetal life the base of the cranium is made of cartilages which constitute a continuous plaque with no interruption in the midline from the foramen magnum to the nasal septum. Ossification centers appear at 4 points: Basioccipit, post sphenoid, pre-sphenoid, and mesethmoid. The pre and post-sphenoid centers are already fused at birth forming the body of sphenoid which is separated
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Spontaneous CSF rhinorrhea usually does not respond to conservative measures. Surgical management of this type of fistula includes both endonasal transsphenoidal approach which is safe and effective, and transcranial extradural or intradural approaches that are recommended by some authors for the lateral extension of the sphenoid sinus to the middle cranial fossa. It is obvious that the endonasal transsphenoidal approach is the preferred procedure for CSF rhinorrhea with clival defect due to the easy accessibility of the defect.

References


from the basioccipital by the sphenoo-occipital synchondrosis. Aside from this synchondrosis which is caudal to the future sphenoid sinus, the ossification is endochondral in nature without fusion points that could explain a development defect and eventual area of dehiscence. According to Hooper, who studied 138 adult sphenoid bones and found 27 defects that could cause CSF leak, non of the defects was situated in the midline of the sphenoid sinus and they were actually located in a very weak boney area that appeared to have been eroded gradually by the pulsation effect of the internal carotid artery or at the superior opening of the transient lateral craniopharyngeal canal. After birth, during the development of the sphenoid sinus, the expanding mucosa causes boney absorption that leads to the formation of the sinus cavity. Lateral extension of the sinus occurs in 27% of adults and can lead to a thin bone at some points passing the lateral limits of the cavernous sinus which, combined with other additional factors, such as the pulsation of the carotid artery can lead to a continuity defect. The internal carotid artery has been found in some cases to erode the lateral margins of the dorsum sella and posterior clinoid.

Coiteiro et al. reported the first 2 cases of rhinorrhea associated with clival defect postulating the pulsation of the basilar artery as a cause in the first case and brisk increase in CSF pressure through repeated Valsalva maneuvers in a saxophone player in the second case. The defects in the 2 cases were in the upper third of the clivus opening into the posterior wall of the sphenoid sinus a few millimeters from the midline as documented by 3-D CT. Our case would be the third reported case in the literature regarding clival defect leading to spontaneous CSF rhinorrhea. The defect was documented 5 mm inferior to the cellar floor.