CASE REPORT

Clival defect causing primary spontaneous rhinorrhea

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ABSTRACT
Primary cerebrospinal fluid (CSF) rhinorrhea due to a defect in clivus is an extremely rare entity, till date only four such cases are reported in the world literature. We present a case of 55-year-old female who presented with primary spontaneous CSF rhinorrhea, for which endonasal surgery was performed about 9 years back. Patient developed recurrent CSF leak with a history of meningitis. Repeat imaging showed clival defect causing CSF leak. Transsphenoidal repair was performed under neuronavigation. In this paper, we discuss the possible mechanism of fistula formation and literature is reviewed.

Key words: Cerebrospinal fluid, clivus, rhinorrhea, spontaneous

Introduction
The incidence of spontaneous cerebrospinal fluid (CSF) rhinorrhea, among the cases of CSF rhinorrhea is only 3-4%. Even rarer are cases in which cause of leak could not be ascertained. We describe a case of 55-year-old woman in whom the exact cause of the CSF leak could not be localized initially, it was believed to be from the sphenoid sinus at first surgery, endonasal repair of the CSF leak failed to benefit the patient. Such cases are extremely rare and to the best of our knowledge only four cases are described in the world literature who were shown to be having clival defect. In this report, we discuss the possible etiology and management of such a rare condition.

Case Report
This patient presented to us about 10 years back with a history of primary spontaneous CSF rhinorrhea, for which she was investigated by computed tomography (CT) cisternography and magnetic resonance (MR) cisternography. The exact site of the leak could not be identified and it was believed to be from the sphenoid sinus; hence, endonasal repair of the sphenoid sinus leak was tried using fat graft, fascia lata and glue. She remained asymptomatic for 3 years, when she had recurrent leak with a history of meningitis, she was treated conservatively at a peripheral center and improved. Patient again presented to us with a short history of 20 days of recurrent CSF leak leading to meningitis and altered sensorium. She was initially treated with intravenous antibiotics and lumbar CSF drainage. Once meningitis settled, her CT cisternography and MR cisternography were done, which showed the clival defect and possible site of leak [Figure 1a]. CT cisternography showed that the contrast material passed from the preoptine cistern into the sphenoid sinus through this bone defect in the clivus [Figure 1b]. Patient underwent sublabial rhino septal transsphenoidal repair of the leak. Previous fat globules and fascia grafts from the sphenoid sinus were removed. Neuronavigation was used to localize the exact site of leak [Figure 1c], arachnoid was seen bulging out of the defect and CSF was seen draining from the site. Fat was used to plug the leak site and reinforced with fascia and tissue glue. A lumbar subarachnoid drain was left in place for 4 days. There was no recurrence of CSF leak at 6 months of follow-up.

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Figure 1: (a) Computed tomography (CT) film showing midline clival defect in axial section. (b) CT cisternography film showing cerebrospinal fluid leaking into the sphenoid sinus from the prepontine cistern, through the defect site. (c) Sagittal image of CT cisternography showing the leakage of contrast from the pontine cistern and filling up the sphenoid sinus cavity

Discussion

Head trauma is the most common cause of CSF rhinorrhea (approximately 80%) while surgery in the paranasal cavities and skull base is the cause in another 16%. Only 3-4% cases of CSF leak are spontaneous.\(^1\) Cribriform plate, sella, sphenoid sinus and ethmoid air cells are the common sites of CSF leaks.\(^2\) Ommaya\(^3\) and Ommaya et al.\(^4\) thought that all cases of CSF rhinorrhea should have a cause. He subcategorized them into “high pressure”, usually due to the tumors and hydrocephalus and “low pressure” caused by congenital abnormalities, osteomyelitis, focal atrophy in the cribiform plate and sella. These atrophic areas can become filled with a pouch of CSF, which can enlarge with normal CSF pressure waves, erode and eventually rupture out of the bone causing CSF leak. Spontaneous CSF rhinorrhea has been subdivided into primary and secondary by O’Connell, depending whether the cause is unknown or could not be identified.\(^5\)

Sponatenous CSF leak from prepontine cistern through a clival defect is a very rare entity and to the best of our knowledge only four cases have been described in the literature.\(^6,7\) Ahmad et al. had described two cases of spontaneous CSF leak through the clivus.\(^7\) One patient had intermittent CSFrhinorrhea, magnetic resonance imaging (MRI) and CT showed a fistulous tract through the mid clivus into the sphenoid sinus. Transnasal transphenoidal repair of the defect was carried out in this patient. The second patient had presented with a similar history. His MRI revealed a defect in the clivus, through which CSF filled sac was ballooning into the sphenoid sinus and was the cause of the CSF leak, sublabial rhinoseptal transsphenoidal repair was performed. These two patients had an uneventful recovery and had no recurrence. Coiteiro et al. had reported two cases of CSF fistula through the clivus and one of their patients had presented with Neisseria meningitis and a short history of CSF leak while second patient had presented with a history of CSF leak for 3 months. Both were treated with transsphenoidal route and had a good outcome.\(^6\)

At times it is difficult to identify the exact cause of CSF leak in spontaneous CSF rhinorrhea. It is believed that all CSF leaks have a common pathophysiological mechanism, which involves disruption of arachnoid, dura and bone and a CSF pressure gradient that is either continuous or intermittently greater than the healing tensile strength of the disrupted tissues.\(^6\) Our patient had a bony defect in the clivus through, which it is likely that the arachnoid and dura might have bulged and with continuous CSF pressure waves must have weakened them to rupture and cause CSF leak. As we know, the maximum CSF pressure is attained in an adult along with higher pressure waves.\(^4,5\) This can also explain the occurrence of leak during adulthood, in all our reviewed patients of clival defect. Other reasons for the defect can be excessive pneumatization of the sphenoid bone and expansion of the sphenoid mucosa, which can cause thinned out bony walls and this combined with other factors can lead to a defect in the clivus.\(^7\)

Hooper had examined 138 sphenoid bones, 5% of which had a defect connecting the sphenoid to the cranium.\(^9\) The most common site of CSF rhinorrhea through the sphenoid sinus is the junction of the anterior portion of the lateral wall of the sphenoid sinus and the floor of the middle fossa.\(^10\) Persistent craniopharyngeal canal can also lead to a leak.\(^9\) The body of sphenoid at birth is formed by fusion of pre- and post-sphenoid centers of ossification. The process of ossification is enchondral in nature and without fusion points that could explain a development defect and ultimate area of dehiscence.\(^1,2\)

Our case also highlights the fact that it is extremely important to identify the site of CSF leak pre-operatively in cases of primary CSF rhinorrhea because a missed site can lead to improper treatment and recurrence of the leak. Even though, our case was a recurrent one, but neuronavigation helped us in localizing the exact site of the defect and its use in such cases cannot be overlooked. If possible, it is imperative to plug the site of the defect using fat globule and re-enforce the repair site with fascia fat and glue.

Conclusion

Sponatenous CSF leak from prepontine cistern through a clival defect is a very rare entity. The most common site of CSF rhinorrhea through the sphenoid sinus is the junction of the anterior portion of the lateral wall of the sphenoid sinus and the floor of the middle fossa, it is imperative to plug the site of the defect using fat globule and re-enforce the repair site with fascia fat and glue.
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Conflicts of interest
There are no conflicts of interest.

References