Spontaneous Cerebrospinal Fluid Leak at the Clivus: Report of 2 Cases and Review of the Literature

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Abstract

Background: Spontaneous cerebrospinal fluid leaks comprise 5% to 10% of all CSF rhinorrhea. Generally, CSF rhinorrhea occur at cribriform plate, sella, sphenoid sinus and ethmoid air. Primary CSF rhinorrhea from clival defect is extremely rare. We describe two cases of spontaneous CSF rhinorrhea through the clivus defect and review the literature.

Case Presentation: The first patient was a 36-year-old female admitted to our department because of clear watery discharge from the right nostril of 3 weeks which aggravated in prone position. The second case was a 57-year-old man referred to our department with the complaint of intermittent rhinorrhea starting 6 months before surgery. He had a past history of bacterial meningitis few months before stating the rhinorrhea which was treated in another center. In both cases, testing of the fluid for beta-2 transferrin was positive. Magnetic resonance imaging and computed tomography cistern gram showed CSF leak through clivus into the sphenoid sinus. In both patients defect was repaired with abdominal fat, reinforced by fascia lata and naso septal flap via “two nostrils - four hands” endoscopic transnasal technique.

Conclusion: At times, the exact pathophysiology of CSF clival fistulae debated, however a combination of anatomical and functional factors play a role in the occurrence of this rare phenomenon. To date, only 16 cases are reported, and the current study reported a group of two consecutive cases. To date, endoscopic transnasal approach is the best therapeutic option to repair midline skull base defect such as the current cases.

Keywords: Spontaneous cerebrospinal fluid leak; Rhinorrhea; Clivus; Meningitis; Endoscopic endonasal approach

Introduction

Spontaneous or non-traumatic cerebrospinal fluid leaks comprise 5% to 10% of all Cerebrospinal Fluid (CSF) rhinorrhea [1,2]. Generally, CSF rhinorrhea can occur at the cribriform plate, sella, sphenoid sinus, or ethmoid air cells [3,4]. However, primary CSF rhinorrhea due to clival defect is extremely rare. The current study describes two cases of spontaneous CSF rhinorrhea through the clivus which were repaired with endoscopic endonasal trans-sphenoidal approach.

Such cases are extremely rare and upon literature review, only 16 cases with clival defect are reported thus far. The peculiar aspect of this case report is related to its rarity. Moreover, evidence was collected from the literature regarding potential etiology, symptoms, and treatment options.

Case Series

Case 1

A 36 -year-old female was admitted for three weeks of clear watery discharge from the right nostril, which was aggravated in prone position. The patient denied any recent trauma. A review of systems was negative except for headaches and nasal discharge. The nasal fluid tested positive for beta-2 transferrin, indicating that the fluid was CSF.

Brain MRI revealed that the sphenoid sinus was filled with Cerebrospinal Fluid (CSF) and sagittal T2 weighted MRI revealed a fistula tract from prepontine cistern to sphenoid sinus (Figure 1). There was no evidence of benign intracranial hypertension. Computed tomography cisternography revealed that the contrast material passed from the prepontine cistern into the sphenoid sinus through this bone defect in the clivus.

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Before surgery a lumbar puncture was performed to administer 0.25 mL of 10% fluorescein with 10 mL of Cerebrospinal Fluid (CSF) to help visualize CSF leaks during surgery and to ensure there was no leak after reconstruction of the defect. Opening pressure measured before injection of the fluorescein which was 18 cmH2O.

The patient underwent endoscopic transnasal transsphenoidal surgery. The anterior and middle portions of the clivus were exposed between both carotid arteries.

During surgery, the defect was defined to the left of the midline in the clivus. The basilar artery was seen through the defect in prepontine cistern (Figure 2). The defect was closed with a multilayer reconstruction consisting of fat, fascia lata, and naso septal flap (Figure 3). Patient discharged in third postoperative day. There was no recurrence of CSF leak at 2 years follow-up.

**Case 2**

A 57-year-old man referred for clear watery discharge from the right nostril of no obvious cause. He suffered from intermittent rhinorrhea starting 6 months prior to arrival. He reported recent history of bacterial meningitis one month ago, which was treated successfully at an outside hospital. On admission, he had no focal

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**Figure 1:** (a) Coronal CT cisternography shows sphenoid sinus filled with CSF. (b) Sagittal T2-weighted MRI of the brain showing Cerebrospinal Fluid (CSF) leak into the sphenoid sinus through clival defect.

**Figure 2:** (a) Intraoperative endoscopic view show CSF leakage from clivus defect just inferior to the sella. (b) Basilar artery in prepontine cistern behind the defect.

**Figure 3:** Endoscopic view after repair of the defect with abdominal fat.

**Figure 4:** (a) CT cisternography showing right sphenoid sinus filled with CSF and (b) the entry of cerebrospinal fluid into the sphenoid sinus. (c) Axial T2-weight MRI revealed right sphenoid sinus filled with CSF. (d) Sagittal T2 MRI shows CSF leakage from prepontine cistern to sphenoid sinus.

**Figure 5:** Intraoperative endoscopic view shows CSF leakage from clival defect.
neurological deficits. Nasal fluid tested positive for beta-2 transferrin.

Brain MRI revealed that the right sphenoid sinus was filled with CSF (Figure 4). CT cisternography showed that the contrast material passed from the prepontine cistern into the sphenoid sinus through this bone defect in the clivus.

After intrathecal administration of 0.25 mL of 10% fluorescein with 10 mL of cerebrospinal fluid the patient underwent endoscopic transnasal approach. Opening pressure measured before injection of the fluorescein which was 15 cmH₂O. After stripping the mucosa from posterior wall of sphenoid sinus, CSF leak was observed in the lateral wall of the sphenoid sinus [3], however, primary spontaneous CSF fistulas, a sphenoidal fistula is most common (60%). Defects in the roof of the ethmoid sinus or in the floor of the skull defect, and secondary spontaneous, when a cause can be found [5,6]. O’Connell first subcategorized spontaneous CSF rhinorrhea in two groups in 1964; primary spontaneous, when there is no cause for rhinorrhea. However, for primary traumatic CSF rhinorrhea, a sphenoidal fistula is most common (60%)

In these cases the junction of the floor of the middle cranial fossa to the lateral wall of the sphenoid sinus is the most common site of fistula in the patient with traumatic CSF rhinorrhea. However, for primary spontaneous CSF fistulas, a sphenoidal fistula is most common (60%).

In a study by Hooper [9], on 138 sphenoid bones, the defect of bones connecting the sphenoid to the cranium leading to CSF leak was observed in 5% of cases; all of which were in lateral wall of the sphenoid sinus [10]. To the best of the authors’ knowledge, only 16 cases of spontaneous CSF leaks from clival defect are reported thus far.

The exact pathophysiology of CSF clival fistulae debated. Morley and Wortzman [11] postulated that congenital bone defects in the middle fossa can explain the leaks through the lateral extensions of the sphenoid sinus. However, from anatomic point of view, there is no clear embryological evidence to explain clival defect causing CSF rhinorrhea.

The clivus is a bony structure composed of the fusion of the posterior portion of the sphenoid body (basisphenoid) and the basilar part of the occipital bone (basioccipital) at the Sphenooccipital Synchondrosis (SOS). SOS fusion can occur at any age, but it usually happens before adolescence. SOS may persist into adult life and may be mistaken for a fracture or defect.

However, this synchondrosis is caudal to the future sphenoid sinus; otherwise, clival ossification would be a continuous enchondral ossification without fusion point that could explain bone defect [12,13]. According to Faizuddin Ahmad et al. [7] most authors believe that excessive pneumatization of sphenoid sinus causes a thin bony wall at some points of clivus and sphenoid. These phenomena, combined with other potential factors such as arterial pulsation and continuous CSF pressure wave ultimately lead to the bone defect at the clivus and CSF leak [14,15].

Sixteen cases of spontaneous CSF leak at the clivus were reported from 1995 to 2018 (Table 1). In all patients, the defect was localized in the upper clivus.

In all sixteen cases reported thus far CSF leak from clival defect occurred in adult patients, which may explain the role of CSF pressure pulsation as a predisposing factor. The CSF pressure pulsation reaches its maximum point in adults, approximately three times higher than that of infants.

Eleven of the sixteen patients were female. In 1995, Coitero et al.
Spontaneous CSF rhinorrhea located at the clivus is an extremely rare condition. To date, only 16 cases are reported, and the current study reported a group of two consecutive cases. It seems that a combination of anatomical and functional factors play a role in the occurrence of this rare phenomenon. To date, endoscopic transnasal approach is the best therapeutic option to repair midline skull base defect such as the current cases. Here, the initial outcome was successful, but long-term follow-up is required.

References


