

A Rare Case of Idiopathic Spontaneous Cerebrospinal Fluid Leak Due to a Clival Defect - A Case Report

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Abstract

Cerebrospinal fluid leak is the most common skull base lesion which otolaryngologists do encounter. It may occur due to defect anywhere along the skull base. Primary spontaneous CSF leak has been a subject of interest owing to its increasing incidence and the associated morbidity and recurrence rates. Although the most common sites of CSF leak are cribriform plate followed by ethmoidal roof, a primary spontaneous CSF leak originating from a defect at the clival region is extremely rare. Not many cases of clival defect were documented in the literature till date. Also, in certain instances, the presentation of the patient may be one of the complications of CSF Rhinorrhea like meningitis, without any prior symptoms or signs suggestive of the condition. So, the probability of csf leak should never be ignored whenever evaluating a case of meningitis.

We report a rare case where a patient presented with meningitis and was treated by a neuro physician initially, which on further investigation revealed a CSF leak originating from a defect at the clivus. The patient was successfully managed by transnasal endoscopic repair with no recurrence or complications after one year of follow up period.

Keywords: Cerebrospinal Fluid; Rhinorrhea; Diagnosis

Introduction

Cerebrospinal fluid rhinorrhea is a result of direct communication between the subarachnoid space and the nasal cavity. CSF rhinorrhea are broadly categorized into traumatic and non-traumatic. Majority of cases are due to trauma. Spontaneous non traumatic cerebrospinal fluid leaks are uncommon and account for only about 5-10% of all csf rhinorrhea [1].

The clivus is a bony structure located in the deepest region of the skull base and spontaneous csf leak arising from a defect at the clival region are extremely rare with very few cases reported in the literature till date.

The csf leak route can act as a direct pathway for the retrograde spread of bacterial pathogens into the cranial cavity and can lead to life threatening sequelae like meningitis. The diagnosis of csf rhinorrhea can be missed when the patients does not have any prior history of signs or symptoms of csf rhinorrhea and present directly with serious complications like meningitis. Therefore, it is essential to have a high index of suspicion in evaluating a case of meningitis to rule out the possible etiology of csf leak. This is a case report of a patient who has presented with clinical features of meningitis which on scrupulous investigation revealed a cerebrospinal fluid leak arising from an uncommon location.

Case Report

A 55- year old female presented to the neurology OPD in a tertiary care hospital with signs and symptoms of meningitis and was treated by a neuro physician. Computed tomography and magnetic resonance imaging of brain were done which revealed a collection in the sphenoid sinus (Figure 1,2 and 3). Meningitis was thought to be secondary to sphenoid sinusitis and the patient was referred to ENT OPD for further evaluation after two weeks of medical management for meningitis. Thorough ENT evaluation was done and a suspicion of csf leak was made. The patient is advised for a Magnetic resonance cisternography, which revealed a defect at the clivus and collection in the sphenoid sinus. Surgical repair of the clival defect was planned and the patient and the attenders were counselled regarding the procedure and an informed consent was taken. Transnasal endoscopic csf leak repair was performed. Intraoperatively, upon identification and widening of sphenoid ostium, fluid collection was noticed, which on draining revealed the csf leak from a bony defect at the clivus (Figure 4). Muscle graft was harvested from the anterolateral aspect of the thigh along with fascia lata. The mucosa was stripped off from the posterior wall of sphenoid sinus and the defect area was delineated.

The muscle graft was snugly fit using bath plug technique into the clival defect taking care not to injure the basilar artery during the process. The closure of defect was confirmed using valsalva maneuver. The graft was further reinforced with fascia lata, fat and surgical. Anterior nasal packing was done which was removed after 48 hours.

Immediate postoperative period was uneventful and the patient was discharged on postoperative day 4. The patient was on regular followup for one year without any recurrence or complications.

Figure 1: Sagittal view of the CT PNS showing a defect at the clival region (yellow arrow) and isodense opacification in the sphenoid sinus (red arrow).

Figure 2: Sagittal section of the T2 MRI brain showing hyperintensity involving the sphenoid sinus.

Figure 3: Axial T2 MRI brain showing hyperintensity involving the sphenoid sinus.

Figure 4: Intraoperative image showing the clival defect.

Discussion

Cerebrospinal fluid rhinorrhea most commonly occurs due to nonsurgical trauma (80%). Iatrogenic causes account for about 16% followed by non-traumatic or spontaneous causes 4% [2].

Patients with cerebrospinal fluid leak usually present with a history of unilateral watery nasal discharge, headache and repeated episodes of meningitis or even brain abscess if timely intervention is not done.

Etiological factors that predispose to spontaneous csf leak are idiopathic, conditions with elevated intracranial pressure like benign intracranial hypertension, hydrocephalus, intracranial space occupying lesions or skull base neoplasms like nasopharyngeal carcinoma, sino-nasal tumors or skull bases erosive conditions like osteomyelitis and mucoceles.

O'connell in 1964, was the first to subcategorize spontaneous csf rhinorrhea into

- Primary spontaneous csf rhinorrhea -when there is no cause of skull defect
- Secondary spontaneous csf rhinorrhea- when a cause can be found [3].

According to a systematic review done by Psaltis, *et al.* [4], spontaneous csf leak account for 40% in which cribriform plate (52.7%) and sphenoid sinus (30.2%) were the common sites. Among the patients with localisation at the sphenoid sinus, primary spontaneous csf leaks arising from a clival defect are extremely rare.

Several theories have been mentioned in the literature, but the exact pathophysiology of spontaneous cerebrospinal fluid leak is not completely understood.

The clivus is a bony structure formed by the fusion of posterior portion of the sphenoid bone (basisphenoid) and basilar part of occipital bone(basiocciput) by sphenoid occipital synchondrosis. This fusion usually occurs before adolescence and is caudal to future sphenoid sinus. Failure of this synchondrosis, may result in continuous enchondral ossification of the clivus without a fusion point that could explain the defective development and eventual areas of dehiscence. Excessive pneumatization of the sphenoid

bone resulting in a thin bony wall at the clivus may also be another possible etiology [5]. Along with these etiological factors, other functional factors like increased intracranial pressure, continuous pressure effect from csf and arterial pulsations may give rise to a bony erosion and may lead to csf leak [6].

Thibaut van zele, *et al.* reviewed 6 cases of transclival spontaneous csf leak, and divided clivus into upper, middle and lower parts, with most common location of bony defect being the upper central part of the clivus [7].

It is imperative to identify the exact location of the csf leak for successful surgical repair. High resolution Computed tomography scan stands as a first line radiological imaging modality in diagnosing and identifying the site of leak in CSF rhinorrhea. MR cisternography is reserved for patients when the high resolution CT imaging shows nonspecific findings [8]. Radionuclide CT cisternography is considered as a third line imaging modality in patients with clinical suspicion of CSF rhinorrhea with negative findings on high resolution computed tomography and MR cisternography [9].

External and endoscopic surgical approaches have been indicated for the csf leak repair. More recently, endoscopic transnasal approach has replaced the older techniques.

The Basilar artery runs just posterior to the clivus. Due to the close proximity to the defect area, the muscle graft should be gently pushed into the clival defect to avoid injury to the basilar artery.

Exact localization of site of defect, delineating the defect area intraoperatively are important for achieving proper closure. Transnasal endoscopic approach offers excellent exposure and visualization of the surgical field with successful results.

Conclusion

Spontaneous cerebrospinal fluid leak due to a clival defect is an extremely rare condition. Prompt suspicion of the possibility of csf leak and thorough evaluation is necessary for accurate diagnosis. Transnasal endoscopic repair is a standard treatment option with successful outcome.

Conflicts of interest

There are no conflicts of interest.

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