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Case Report

Spontaneous cerebrospinal fluid leakage through fistulas at the clivus repaired with endoscopic endonasal approach

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Abstract

Background: Causes of cerebrospinal fluid (CSF) leakage are primarily traumatic or iatrogenic in origin. In contrast, spontaneous CSF leakage is somewhat rare, and detection of the fistula can be challenging. Meningitis associated with CSF leakage can be life threatening. It is therefore critical to surgically repair the fistula once the underlying cause has been accurately identified. Spontaneous CSF leakage located at the clivus is an extremely rare condition.

Case Description: We present the case of a 38-year-old male with sudden-onset headache and subsequent disturbances of consciousness. The patient was diagnosed with severe meningitis caused by CSF leakage through fistulas at the clivus, which were clearly identified on dynamic imaging using high-resolution computed tomography (CT) with intrathecal injection of contrast medium. After the meningitis was resolved, successful endoscopic repair of the CSF fistula with autologous materials was performed. There has been no recurrence of meningitis for 5 years.

Conclusion: Spontaneous CSF leakage at the clivus is an extremely rare condition. High-resolution CT cisternogram could accurately detect CSF leakage through the clivus. A transnasal endoscopic approach was a useful and reliable method of repairing the fistula at the clivus.

Key Words: Cerebrospinal fluid, clivus, endoscope, fistula

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INTRODUCTION

The cause of cerebrospinal fluid (CSF) leakage is predominantly traumatic, iatrogenic, or neoplastic, and can occasionally be spontaneous or congenital.[16,19,21] Obvious bony erosion or traumatic destruction may be the underlying cause of CSF leakage, which frequently requires surgical repair of the fistula. Because CSF leakage is a risk factor for meningitis, early repair of a CSF fistula is necessary once detected.[19,21,27]

Recent neuroradiological advancements enable physicians to detect the precise localization of the CSF fistula, even in cases with subtle spontaneous CSF leakage.[3,14] Moreover, satisfactory surgical outcomes following endoscopic repair of CSF leakages have been increasingly reported.[10,13,18] We present here an extremely rare case of CSF leakage through fistulas at the clivus in the context of severe recurrent meningitis. The CSF fistulas were clearly identified on high-resolution dynamic computed tomography (CT) and successfully treated using a transnasal endoscopic approach.

CASE REPORT

A 38-year-old male presented with progressing high fever, headache, and vomiting for 2 days. Owing to gradual deterioration of his level of consciousness into a stupor, he was transported by ambulance to our hospital. Neurological examination revealed severe nuchal stiffness with no focal deficits. He never suffered any rhinorrhea before admission. The result from serological examination showed that the white blood cell count was 21,200 and C-reactive protein level was 24.0 mg/dL, indicating a remarkable infectious state.

A CT study was performed using 64-slice multidetector computed tomography (MDCT; Toshiba Aquilion, Tokyo, Japan). Three-dimensional images with surface rendered and multiplanar reformatted (MPR) images were reconstructed on the workstation. The axial CT scan images revealed multiple intracranial air bubbles with pneumatization of the dorsum sellae [Figure 1a], fluid collection in the sphenoid sinus [Figure 1b], and bone defects at the clivus [Figure 1c]. Hydrocephalus or empty sella, as causes of intracranial hypertension, were not found. The bone CT reconstruction clearly showed multiple bone defects on the upper and middle third of the clivus [Figure 1d].

According to previous medical records, the patient had experienced severe meningitis once 3 years prior. However, he never recognized CSF rhinorrhea and a CT scan was not performed at that time. Therefore, fluid collection in the sphenoid sinus or intracranial air had never been detected. This had been treated successfully with antibiotics, although the cause of the meningitis remained unknown.

The cause of the high-grade fever was investigated with CSF sampling, which revealed evidence of severe meningitis; sugar, 1 mg/dL, protein, 576 mg/dL, and cell count, 27920/mm^3. CSF culture showed presence of *Streptococcus pneumoniae*. The meningitis was treated appropriately with the antibiotic meropenem, which resulted in a good recovery of general symptoms and resolution of high-grade fever.

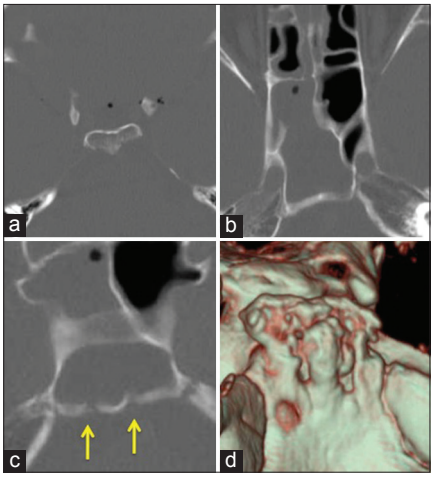

Figure 1: A CT scan of the axial images reveals (a) multiple intracranial air bubbles and pneumatization of the dorsum sellae, (b) fluid accumulation in the sphenoid sinus, and (c) bony defects at the clivus (arrows). (d) Reconstructed three-dimensional CT image clearly demonstrates multiple bony erosive defects at the clivus

The multiple bone defects at the clivus were considered the cause of the CSF leakage. We therefore aimed to determine the dynamics of CSF through the defects. High-resolution CT cisternography was performed with intrathecal infusion of 5 mL contrast medium. The reconstructed MPR images confirmed the passage of contrast medium through the bone defects at the clivus into the sphenoid sinus 1 h after injection of the contrast medium [Figure 2]. Moreover, these MPR images revealed pneumatization of the dorsum sellae. Surgical repair of the CSF fistulas was planned in order to prevent further recurrence of meningitis. Interestingly, he never encountered CSF rhinorrhea during admission, likely because he was maintained in supine position until surgery. The repair of CSF fistula was performed under general anesthesia 5 days after resolution of his fever.

The mucosa over the posterior wall of the sphenoid sinus was completely removed in order to visualize the clival bone accurately. The intraoperative endoscopic observation revealed CSF leakage through bone dehiscences at the clivus detected on the preoperative CT images. The fistulas were successfully repaired in a multilayered fashion with fibrin glue, absorbable polyglycolic acid felt (Neoveil, Gunze, Japan), autologous fat, and fascia grafts from the abdomen followed by a pedicled nasoseptal mucosal flap [Figure 3]. The pedicled flap was placed to wholly cover the fat graft. CSF lumbar drainage was in place for 4 days. The postoperative clinical course was uneventful, and there was never any rhinorrhea. Two weeks after surgical repair, CT cisternography showed no evidence of CSF leakage into the sphenoid sinus. In addition, recurrence of CSF

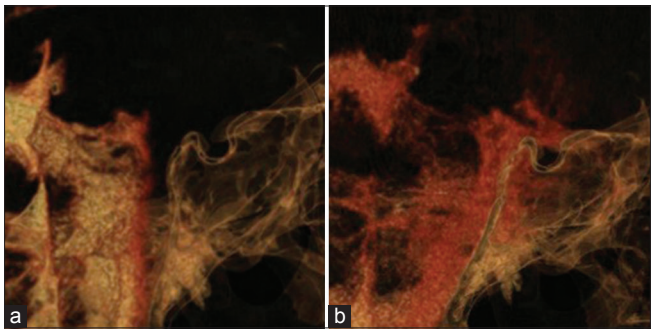

Figure 2: Sagittal reconstruction of MPR images obtained by high-resolution CT with intrathecal injection of contrast medium. (a) Thirty minutes after injection, contrast medium advances into the interpeduncular and prepontine cistern, but not into the sphenoid sinus. (b) One hour after injection, contrast medium is clearly shown entering into the sphenoid sinus through the bone defects at the clivus

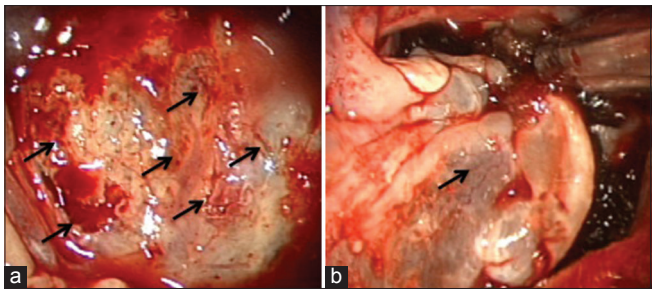

Figure 3: Intraoperative endoscopic views. (a) CSF leakage through the multiple bone defects at the clivus (arrows) was confirmed clearly. (b) Pedicled nasoseptal mucosal flap (arrow) overlayed along the clivus with fibrin glue soaked sheet of polyglycolic acid (Neoveil)

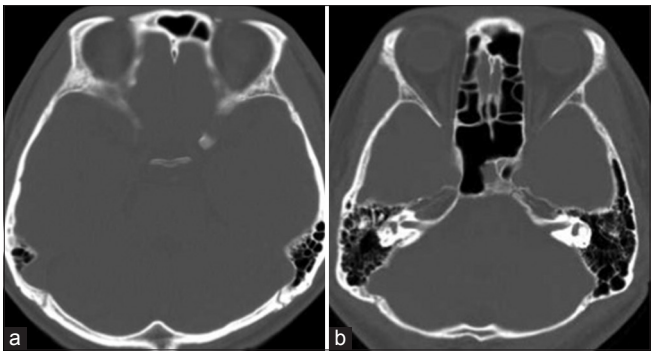

Figure 4: CT scan reveals (a) no intracranial air, and (b) no fluid accumulation in the sphenoid sinus during the follow-up period

accumulation in the sphenoid sinus or intracranial air on CT scan was not found for 5 years after discharge [Figure 4].

DISCUSSION

The sphenoid sinus is a primary location for CSF leakage related to not only head trauma and postoperative causes, but also spontaneous leaks that can be safely and effectively managed using endoscopy.[8,10,11,18,24] For primary spontaneous CSF fistulas, there is marked predominance for the sphenoid sinus (60%),[20] and symptoms are usually characterized by a longer history of CSF rhinorrhea and/or repeated episodes of meningitis, like this presented case.

Although CSF leakage through defects in the middle fossa floor into the sphenoid sinus extending laterally have been frequently reported, fistulas located at the clivus are extremely rare.[2,6,26,27] The presence of congenital dehiscence and the formation of a small meningocele are reported to be the most likely causes for these CSF leakages. Some authors described the mechanisms of CSF leakage at the clivus as follows: CSF pressures and hydrostatic pulsatile forces may induce pit holes at the site of the arachnoid villi with herniation of dura, arachnoid, and/or brain tissue. Subsequently, an encephalocele can develop and lead to a CSF leak into the sinus.[17,28] In other reports, the findings of pits or irregularities along the floor of the middle cranial fossa have been observed in 63% of patients with CSF leakage.[25] These areas can be potential sites for CSF fistulas, although they have not been reported at the clivus. Meanwhile, in our case, the radiographic features of increased intracranial hypertension, such as empty sella and arachnoid pits, could not be found. Additionally, the intraoperative endoscopic finding along the clivus was many dehiscences on the posterior wall of the sphenoid sinus, below the sellar floor, without the formation of any meningocele.

Defects in the sphenoid bone might also arise as the sinuses develop. Bony defects are always located at the upper part of the clivus, above this spheno-occipital synchondrosis, which makes clival dysgenesis as a cause of spontaneous CSF leak less likely.[28] That is why spontaneous CSF rhinorrhea located at the clivus is an extremely rare condition. Early in fetal life, the complete cranial base – from the foramen magnum to nasal septum – is made of cartilage. A plaque constitutes the midline for its entire length with no interruption, and ossification centers develop later in this plaque. The process of ossification is endochondral in nature, without fusion points that could explain a development defect and eventual areas of dehiscence.[1,9] Another possible etiology is excessive pneumatization of the sphenoid bone with further weakening of the bone more posteriorly. During the development of the sphenoid sinus, aerial expansion of the sinus occurs through trabecular bone resorption, leaving a thin bony wall at the clivus. Moreover, the expanding mucosa causes bone absorption and may leave a thin bony wall at some points, which combined with other additional factors may ultimately lead to a continuity defect later in life.[9]

It is considered that spontaneous CSF transclival leakage is caused by a combination of anatomical and functional factors. From the point of anatomical explanation, excessive pneumatization of the sphenoid bone leads to weakening of the bone posteriorly[28] and disruption of the dura and arachnoid. As a functional factor, a pressure gradient produced by continuous CSF pressure may play a crucial role, leading to clival fenestration.[7,28] It is still argued that even physiological elevations and fluctuations in intracranial pressure encourage the development of dura and arachnoid herniations.[5] In our case, the reconstructed MPR images revealed extensive pneumatization of the dorsum sellae and clival recess, suggesting the patient had been at significant risk of CSF leakage.[4,29] However, from the intraoperative findings, there was no evidence of encephalocele and meningocele. In general, the clivus has an anatomical feature not affected severely by CSF pulsatile pressures and turbinate flow.

It might be impossible to draw conclusions from one case, but workup for patients with meningitis of unknown etiology is described below. First, it is important to check a CT scan for points of fluid collection in the sphenoid sinus and intracranial air at the sellar, suprasellar, and parasellar regions. Subsequent thin slice CT bone images of the sphenoid sinus were needed to detect the fistula in the current case. Second, it is important to check the medical history for repetitive meningitis, but it is not a requirement. Third, CT cisternography is useful to evaluate fistulas through which CSF can pass.

Wielgosz has reported the usefulness of high-resolution CT for bone defect localization and management of spontaneous CSF rhinorrhea from leakages in the posterior wall of the sphenoid sinus.[30] Akyuz clearly showed a bone defect of the clivus with MDCT in a patient with CSF rhinorrhea from a transclival meningocele.[20] In our patient, a bone defect in the clivus was also identified using MDCT, a technique characterized by higher spatial resolution, faster speed, greater anatomic coverage, and higher quality MPR and three-dimensional reconstruction images. Moreover, the dynamics of CSF passage through bone defects in the clivus into the sphenoid sinus were clearly identified on the high-resolution CT cisternogram with the intrathecal infusion of a contrast medium. Therefore, high-resolution CT was considered an effective tool for localization of the bone defect.

A satisfactory surgical outcome requires exact preoperative diagnosis, an appropriate operative approach, and a surgeon's skill and experience. Transnasal endoscopic repair has now been established as a reliable method for CSF leakage repair in the ethmoid and sphenoid sinuses.[2,26] Cui *et al*. reported their successful experience of endoscopic repair for recurrent CSF rhinorrhea in a series of 32 patients without further postoperative recurrence.[12,23] Stamm reported transnasal endoscopic-assisted techniques in the clival region could provide clear visualization, and would be a superior alternative to open surgical approaches in most cases.[7]

Recently, materials to repair bone defects have been presented by many surgeons. Many cases with CSF fistulas in the sphenoid sinus are repaired using a procedure of packing with autologous muscle or fat. The reported success rate of endoscopic fistula repair is considerably high, depending mainly on the accurate detection of the fistula. Cassano addressed the effectiveness of the overlay apposition of a lower turbinate mucoperiosteal graft fixed with fibrin glue and oxidized cellulose polymer (Surgicell, Ethicon Inc., USA).[8] Akyuz *et al*. presented a case of transclival meningocele similar to the case reported here, and managed the bone defect, following the removal of the meningocele, using a fascia lata graft, fat tissue, and bio-glue (Bio-glue, Cryo Life Inc., USA).[2,20] In our patient, a pedicled nasoseptal mucosal flap and fascia/fat grafts from the abdomen were used as autologous materials for repair of the bone defect at the clivus. Some authors have indicated pedicled mucosal flaps are suitable for the closure of CSF fistulas because they are highly vascularized enough to regenerate *de novo* mucosa at the bone defect.[22]

How to follow up with patients after surgical repair of CSF fistula is a very important problem that has not been discussed before in the literature. It is difficult to confirm surgical success leading to the reliable resolution. In our case, CSF lumbar drainage was placed for 4 days. CT cisternography was performed again 2 weeks later, which showed no evidence of CSF leakage into the sphenoid sinus. After discharge, CT scan was performed to rule out findings of CSF accumulation in the sphenoid sinus or intracranial air. If CSF rhinorrhea appeared again and CT scan showed fluid accumulation in the sphenoid sinus, I would consider CT-cisternography should be performed immediately. Van Zele *et al*. insisted on the importance of follow up with patients after surgical repair of CSF fistulas and their follow up was longer (10.3 years, 3–15) than other reports (1 month to 2 years).[1,9,15,28] However, they reported 100% success rate of closures during the follow-up. Although the follow-up of our case is 5 years, at least 10 years follow-up seems to be necessary to confirm success of surgery.

In conclusion, spontaneous CSF leakage through fistulas at the clivus was clearly identified in a dynamic study using MDCT with intrathecal injection of contrast medium. The repair was successfully performed with an endoscopic endonasal approach without further recurrence.

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