

Primary spontaneous cerebrospinal fluid leaks located at the clivus

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ABSTRACT

Transclival meningoceles and primary spontaneous cerebrospinal fluid (CSF) leaks at the clivus are extremely rare lesions and only few of them have been reported in the literature. We report here six cases of transclival primary spontaneous CSF leaks through the clivus. A retrospective case study was performed. We reviewed six cases involving sinonasal CSF leaks located at the clivus treated between 1997 and 2009. Presenting symptoms, duration of symptoms, defect size, site of defect, surgical approach and technique of defect closure, intraoperative complications, postoperative complications, and recurrences are discussed. All CSF leaks were located in the upper central part of the clivus. Two of six patients showed signs of increased intracranial pressure (ICP) including arachnoid pits and/or empty sella. For three patients a purely transnasal approach was used with multilayer reconstruction using a nonvascularized graft, and three patients underwent a transnasal transseptal approach with a multilayer reconstruction, with nasoseptal flap. No recurrences of CSF leaks at clivus or other sites were observed to date with a mean follow-up of 10.3 years (range, 3–15 years). Spontaneous CSF rhinorrhea located at the clivus is an extremely rare condition. To date, only eight cases have been described. Here, we report the largest group of six consecutive cases. Irrespective of the used reconstruction technique in all cases a 100% closure rate was achieved. However, identification of increased ICP is an essential aspect and this condition should be treated either medically or surgically.

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PPrimary spontaneous cerebrospinal fluid (CSF) leaks represent a specific subgroup of all CSF fistulas. A recent meta-analysis shows that ~50.2% of all cases of CSF rhinorrhea are caused by head trauma and 49.2% had a nontraumatic origin. Of these nontraumatic CSF fistulas, spontaneous fistulas are the most commonly reported with 41% of fistulas.¹ Other types of CSF fistulas are related to iatrogenic trauma after surgery (30%), tumors (5%), or congenital malformation of the skull base (3%).¹ The exact cause of CSF leakage in cases of primary spontaneous CSF rhinorrhea remains a dilemma; however, it represents a distinct clinic entity that is likely a variant of idiopathic intracranial hypertension.^{2,3} Most of these patients show clinical signs and radiographic features of increased intracranial pressure (ICP) such as empty sella syndrome (80%),^{4,5} arachnoid pits (63%), and a thinned and broadly attenuated skull base. In 31% of these patients multiple skull base defects on computed tomography (CT) scan are seen.⁶ Patients with spontaneous CSF

leaks are generally obese middle-aged women.⁷ CSF pressure monitoring using lumbar drain pressures shows in most cases an average pressure of 25–27 cmH₂O that is well above the normal range of 10–15 cmH₂O pressure.^{8,9} Although primary spontaneous CSF fistulas can have relatively small bony defects, these defects are frequently associated with the formation of large encephaloceles ranging from 50 to 100%. A second characteristic of primary spontaneous CSF leaks is the high recurrence rate after closure. The overall success rate for a primary endoscopic repair of CSF leaks has achieved of 90%, increasing to 96.6% with the second attempt.¹

Most patients with primary spontaneous CSF fistula present with a longer history of CSF rhinorrhea and isolated or repeated episodes of meningitis. The main complications of persistent CSF leakage are meningitis or brain abscess, which are potentially fatal.

Primary spontaneous CSF fistulas can occur along the skull base at the cribriform plate, ethmoid sinuses, and frontal and sphenoidal sinuses. The most common site of CSF fistulas is found in the roof of the ethmoid sinus or in the floor of the anterior cranial fossa, which communicates with the ethmoid and frontal sinuses. However, for primary spontaneous CSF fistulas, there is marked predominance of the sphenoidal fistulas (60%).¹⁰ Transclival meningoceles and primary spontaneous CSF leaks are extremely rare lesions and only a few of them have been reported in the literature.

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Here, we report six cases of transclival primary spontaneous CSF leaks through the clivus.

METHODS

After obtaining Institutional Review Board approval, we retrospectively reviewed six cases involving sinonasal CSF leaks located at the clivus treated at our institution between 1997 and 2009. Skull base malignancies causing a large defect after resection were excluded. The diagnosis of a sinonasal CSF leak was made, based on symptoms, laboratory evaluation of the nasal discharge, and fine-cut high-resolution CT and magnetic resonance imaging. Retrospectively presenting symptoms, duration of symptoms, defect size, site of defect, surgical approach and technique of defect closure, intraoperative complications, postoperative complications, and recurrences were reviewed.

Surgical Technique

Fistula repair was performed with an endonasal endoscopic approach under general anesthesia. In three patients a purely transnasal approach was used and three patients underwent a transnasal transseptal approach as described previously.¹¹ Briefly, the surgery begins with a classic anterior incision for septoplasty. A mucoperichondrial/mucoperiosteal dissection is made at both sides. The posterior part of the nasal septum is removed and the sphenoid rostrum and anterior wall of the sphenoid sinus are exposed. The next step is the creation of a nasoseptal flap at one side of the nasal septal mucosa. With this technique surgical instruments enter through one nostril and the endoscope through the septum while the other side of the septal mucosa is left intact and preserved.

Intraoperative identification of skull base defects and CSF leaks included endoscopy with intrathecal fluorescein injection. For the classic transnasal approach the middle and superior turbinates were displaced laterally at both sides. After identification of the natural sphenoid ostium it was enlarged laterally and inferiorly with a Kerrison forceps. The bony defect was identified and the edges were carefully drilled to remove overlying mucosa. For all patients a multilayer reconstruction consisting of fat, fascia lata, and flap was used. In three patients a free-grafting flap (turbinate mucosa) was placed into the defect as an onlay mucosal graft and a nasoseptal vascularized flap was used for the other three patients. To stabilize the graft or pedicled flap, Gelfoam (Pharmacia and Upjohn Company, Kalamazoo, MI) were applied over the graft or the pedicled flap and Rapidrhino sponges (ArthroCare ENT, Austin, TX) were packed into the nasal cavity and kept in place for 3 days. No lumbar drains were routinely placed pre-, peri-, and postoper-

atively except if signs of intracranial hypertension were observed.

RESULTS

Between 1997 and 2009, six patients suffering from a CSF leak at the clival region were operated on at the São Paulo Skull Base Center (Table 1). All patients were diagnosed with primary spontaneous CSF fistulas as described previously. The mean age was 52.6 years with a range from 37 to 78 years and five patients were women. All patients presented with a chief complaint of watery rhinorrhea. The average duration of CSF rhinorrhea was 2.5 months. Four of six patients experienced headaches in the months before the diagnosis was made. In all patients a CT scan was performed showing a defect at the clivus (Fig. 1). All CSF leaks were located in the upper central part of the clivus. The average size of the skull base defects was 2.05 mm, ranging from 1.5 to 3.0 mm in diameter. Two of six patients showed signs of increased ICP including arachnoid pits and/or empty sella. All patients underwent an endonasal endoscopic approach under general anesthesia. In three patients a meningoencephalocele was found perioperatively. The two patients with presenting signs and symptoms of intracranial hypertension also received simultaneously a lumboperitoneal shunt. In this group of spontaneous CSF fistula at the clivus, the success rate was 100%. Complications were not noted, with the exception of one episode of postoperative meningitis in one patient. No recurrences of CSF leaks at the clivus or other sites were observed, to date, with a mean follow-up of 10.3 years (range, 3–15 years).

DISCUSSION

In this study, we present a group of six patients with spontaneous CSF leaks originating from the clivus. Spontaneous or idiopathic CSF leaks make up only 3–4% of CSF leak etiologies in most series.^{1,12} Spontaneous CSF leaks arising from the prepontine cistern through the clivus are thus extremely rare. To date, eight cases have been described and we present the largest group of six consecutive cases.^{13–17}

The exact etiology of spontaneous clival CSF leaks remains elusive. For a spontaneous CSF leakage to occur, there has to be an osseous defect, disruption of the dura and arachnoid, and a pressure gradient.¹⁸ It is likely that a combination of these factors, both anatomic and functional, also play a role in spontaneous transclival CSF leaks. In contrast to the congenital bone defects in the lateral extensions of the sphenoid sinus,¹⁷ the particular location of the defects in the clivus are difficult to explain by embryological dysgenesis. The clivus is composed of the posterior portion of the sphenoid body (basisphenoid) and the basilar part of the

Table 1 Description of patients, symptoms, and types of repair

Gender	Age (yr)	Size of the Defect (mm)	Location of the clivus	Duration of CSF Rhinorrhea (mo)	Rhinoliquorrhea	Meningitis	Pneumoencephalocoele	Headache	Meningoencephalocoele	Signs of Intracranial Hypertension	Grafting Material	Lumboperitoneal Shunt	Recurrence
F	37	2.5	Upper central	2	Yes	Postoperative	No	Yes	Yes	Yes	Non vascularized flap	Yes	No
F	61	3.0	Upper central	4	Yes	No	No	Yes	Yes	No	Non vascularized flap	No	No
F	78	2.0	Upper central	3	Yes	No	No	No	Yes	No	Non vascularized flap	No	No
F	42	1.5	Upper central	2	Yes	No	No	Yes	No	No	Nasoseptal flap	No	No
F	48	1.8	Upper central	3	Yes	No	No	No	No	No	Nasoseptal flap	No	No
M	50	1.5	Upper central	1	Yes	-No	No	Yes	No	Yes	Nasoseptal flap	Yes	No

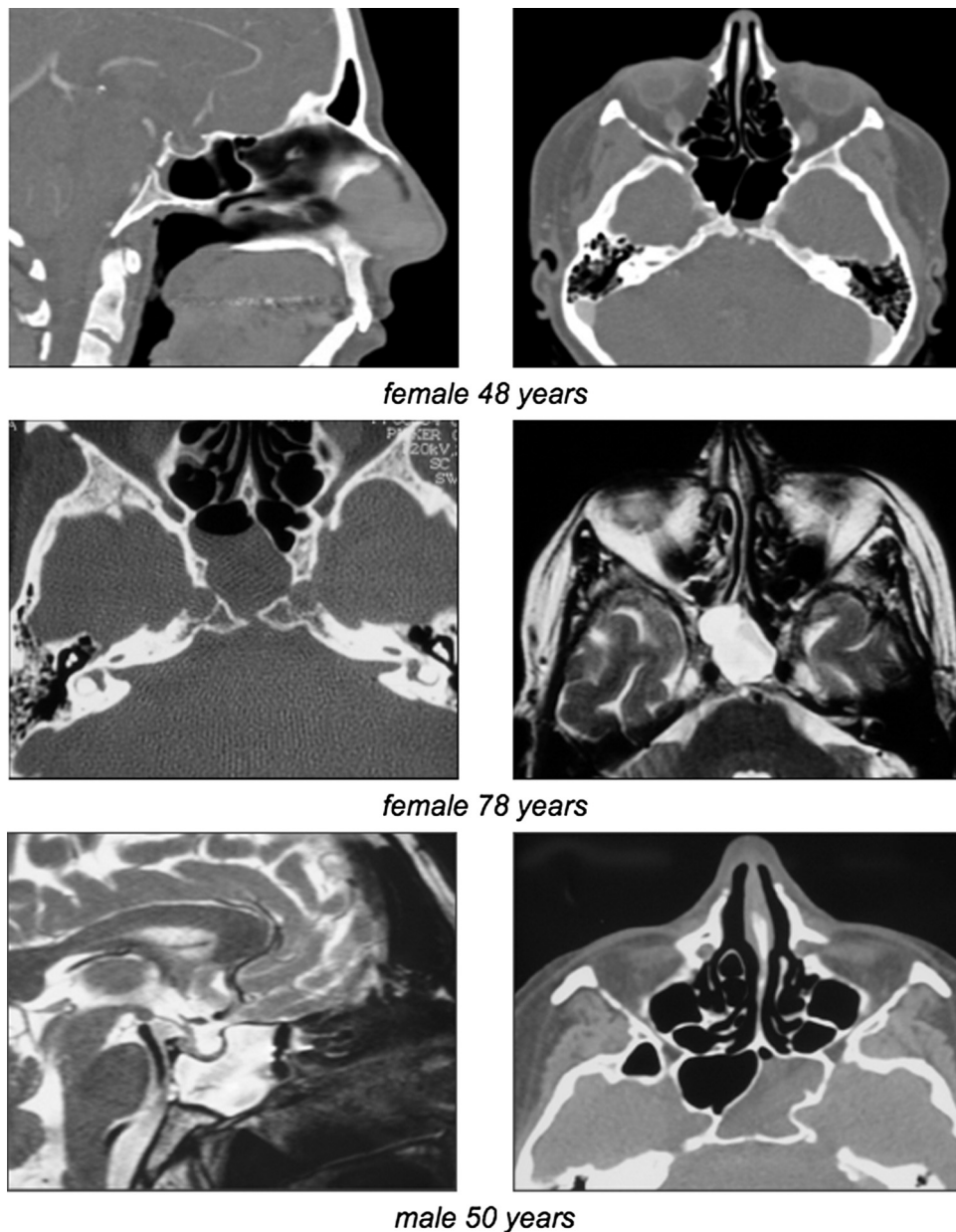
Signs of intracranial hypertension include empty sella, arachnoid pits. CSF = cerebrospinal fluid.

occipital bone (basiocciput) and is further subdivided into upper, middle, and lower thirds.¹⁹ This sphenoccipital synchondrosis may persist into adult life and may be mistaken for a fracture or defect. In our group of translclival CSF leaks the bony defect was always located at the upper part of the clivus above this sphenoccipital synchondrosis, which makes the clival dysgenesis as a cause of spontaneous CSF leak less likely. To date, only one case has been described in which a deficiency in bone development associated with Marfan's syndrome gave rise to clival fenestration and translclival CSF rhinorrhea.^{13-17,20}

The most likely anatomic explanation for translclival CSF leaks 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 some points of the clivus and sphenoid.^{18,21} Additional functional factors such as pulsating effects of arteries¹⁵ or continuous pressure pulses of CSF may ultimately lead to a continuity defect and CSF leak.¹⁷ This is illustrated in two independent cases of clival CSF leaks in saxophone players.^{13,15} In these patients, repetitive increases of ICP through successive Valsalva maneuvers while playing the saxophone, which acted on a congenital thin area, have resulted in a clival bone dehiscence and consequent meningocele with CSF fistula.

In line with the increased ICP hypothesis, two of our patients presented with radiological signs of increased ICP (empty sella and/or arachnoid pits). However, in the remaining four patients in our groups and the previously reported eight cases of spontaneous clival CSF leaks in the literature, no empty sella syndrome or signs and symptoms of increased ICP were observed.¹³⁻¹⁷ Accurately diagnosing patients with spontaneous CSF leaks is critical for the successful repair of these patients because multiple studies have identified increased ICP as a negative risk factor for successful repair.⁷ One limitation of this study is that no other techniques such as ICP monitoring were performed to determine whether these patients had increased ICP.

The association between spontaneous CSF fistula through other areas of the sphenoid and ethmoid sinuses and empty sella syndrome is well described, especially in obese, middle-aged women. In these patients a chronic increased ICP due to increased abdominal and thoracic pressures leads to cranial venous congestion and permanent benign intracranial hypertension.¹⁶ Besides intracranial hypertension, elevated body mass index and female gender are major factors related to the failure of CSF leak repair. Similar to other observations, we noted a predominance of middle-aged women in our group of spontaneous CSF leaks. An overall failure rate of endoscopic spontaneous CSF leak closure of 12.5%²² up to 28.6% has been docu-



female 48 years

female 78 years

male 50 years

Figure 1. Computed tomography (CT) and magnetic resonance imaging scan of the three clival leaks.

mented.²³ Therefore, in case of suspected intracranial hypertension, the placement of lumboperitoneal or ventriculoperitoneal shunts or adjunctive medical treatment with acetazolamide seems advisable and logical to prevent recurrence. In our patient group of spontaneous clival CSF leaks the two patients with empty sella and arachnoid pits on CT and magnetic resonance imaging also underwent a simultaneous procedure with placement of a lumboperitoneal shunt.

Various endoscopic approaches to the sphenoid have been described. Defects in the central sphenoid can be approached through the endonasal endoscopic approach by performing a wide bilateral sphenoidotomy. A traditional midline transseptal approach or the newer transnasal transseptal approach may also pro-

vide excellent access to sellar leaks.²⁴ In our patient groups fistula repair was performed with an endonasal endoscopic approach. In three patients a purely transnasal approach was used and three patients underwent a transnasal transseptal approach. All defects were closed with a combination of fat, fascia, and a free or nasoseptal flap. A 100% success rate of closures was achieved with a mean follow-up period of 10.3 years.

The high success rate of all of these spontaneous clival leaks is in contrast with other spontaneous CSF leaks along the skull base where recurrence rates of 12.5%²² up to 28.6% have been documented.^{23,25}

However, our results and techniques are in line with the other eight cases described, although the follow-up in our patient group is considerably longer. Various

grafting material such as abdominal fat, fascia lata, muscle, and bone have been used in mostly a multi-layer fashion. However, irrespective of the grafting materials used a 100% closure rate of these clival CSF leaks was achieved.^{15,17} In two patients a lumbar peritoneal shunt was placed postoperatively. Although advised by some authors²⁶ and treatment algorithms we do not use routinely prophylactic perioperative lumbar drain in patients submitted for a CSF leakage repair. The lumbar drain may cause a CSF hypotension that predisposes to pneumocephalus and to plug dislocation. Even in spontaneous CSF fistulas, when the etiology is a longstanding but low level of increase in CSF pressure, we can avoid perioperative lumbar drainage. On the other hand, we do use it in the treatment of postoperative fistulas or if signs of intracranial hypertension are present.

CONCLUSION

Spontaneous CSF rhinorrhea located at the clivus is an extremely rare condition. To date, only eight cases have been described. Here, we report the largest group of six consecutive cases. In analogy with other locations of spontaneous CSF rhinorrhea we show here for the first time that radiological signs of increased ICP are also present in clival CSF leaks. Etiologic factors of this condition may include innate skull base malformations, overpneumatized sphenoid sinus, and additional functional factors such as pulsating effects of arteries or increased ICP. To date, the endoscopic endonasal approach is the best option. Irrespective of the used reconstruction technique in all cases, a 100% closure rate can be achieved in this subgroup of spontaneous CSF leaks. However, because spontaneous idiopathic nasal CSF leaks have the highest rate of recurrence after operative repair a long-term follow-up and identification of increased ICP is mandatory and this condition should be treated either medically or surgically, especially in obese middle-aged women.

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