Spontaneous Cerebrospinal Fluid Leaks Originating from Multiple Skull Base Defects

ABSTRACT—Spontaneous cerebrospinal fluid (CSF) leaks of temporal bone origin are more prevalent than once believed. Twenty-eight of the 61 cases documented in the world literature have been reported since 1992. All but four of these cases involved unilateral defects. The authors have previously reported experiences with 12 cases, with the vast majority of defects localized to the tegmen tympani. These patients also had demonstrated a single area of bone and dural dehiscence. We report two additional cases of spontaneous CSF leak originating from multiple/distant skull base defects. As in previously reported multisite cases, one of our patients demonstrated an elevated opening pressure on lumbar puncture. Significant time intervals existed between leak site presentations, which emphasizes the importance of careful follow-up for treated patients. Potential etiologies and associated factors are also discussed. This patient subset contributes another dimension to the evolving natural history of spontaneous CSF leakage. (*Skull Base Surgery*, 6(4):227–230, 1996)

Temporal bone CSF leakage originates most commonly from traumatic, infectious, or congenital defects. Spontaneous CSF leakage occurs without apparent cause. Typically, the spontaneous leak patient presents with symptoms and clinical findings consistent with a unilateral middle ear effusion. Myringotomy can then result in profuse watery otorrhea. Because meningitis is not uncommon in this patient group, prompt diagnosis and treatment are essential.

To date, 61 cases of temporal bone spontaneous CSF leakage have been documented. Heightened clinical awareness and recognition of these defects has resulted in 46% (28 patients) of the reported cases appearing in the literature since 1992. All but four of these previously reported cases have involved a unilateral site of leakage. Rotilio, in 1982, described the first multisite case involving a patient with a tegmen tympani leak and a subsequent sphenoid

sinus leak.¹ In 1986, Ferguson reported on a patient who demonstrated bilateral temporal bone leak sites.² Each of these defects were repaired through a transmastoid approach. In 1995, May published a series of spontaneous CSF leaks which discussed two individuals with multisite leaks.³ Table 1 provides additional details of these previously published cases.

The authors have previously reported their experiences with 12 spontaneous CSF leak cases, with the vast majority of defects localized to the tegmen tympani.^{4,5} At the time that these cases were described, all patients demonstrated a single area of bone and dural dehiscence. In this report we present two patients with spontaneous leaks originating from multiple skull base defects. The first case represents a previously unreported patient, while the second case involves a previously reported patient with a unilateral leak who subsequently developed

Table 1. Multisite Spontaneous CSF Leak Cases

<i>Reference</i>	<i>Age</i>	<i>Sex</i>	<i>1st Leak Site</i>	<i>2nd Leak Site</i>	<i>Interval</i>
Rotillio 1982	64	M	L tegmen tympani	sphenoid sinus	2 years
Ferguson 1986	50	F	R tegmen	L Tegmen	2 months
May 1995	64	F	R antrum	R epitympanum	2 years
May 1995	62	F	L mastoid tegmen	L ethmoid sinus	3.5 years
Pappas 1992	57	F	R tegmen tympani	sphenoid sinus	4 years
*	62	M	L sinodural angle	R tegmen tympani	Electively explored

*—previously unreported case; R—right; L—left

a sphenoid sinus leak site. Details of these cases are elaborated in the following sections and are also summarized in Table 1.

CASE REPORTS

Case 1

A 62-year-old male was referred with a history of recurrent meningitis. A left-sided middle ear effusion had been noted and treated with each bout. The patient had undergone a left myringotomy with ventilation tube insertion, which resulted in drainage of clear fluid postoperatively. Analysis of this otorrhea demonstrated B2 transferrin, confirming the presence of CSF.

Clinical examination demonstrated a normal appearing tympanic membrane on the right side and a patent myringotomy tube on the left side. Audiometric studies revealed a conductive hearing loss of mild degree on the left side and moderate degree on the right side.

Computed tomography (CT) demonstrated a bony dehiscence of the mastoid tegmen plate on the left side (Fig. 1). On the right side, marked thinning of the tegmen plate was identified with questionable bony dehiscence (Fig. 2).

The patient underwent surgery for repair of his left-sided defect. A spinal drain was placed following anesthesia induction. CSF diluted fluorescein was injected intrathecally. A transmastoid approach revealed a middle fossa tegmen plate encephalocele involving the sinodural angle. No active leakage or fluorescein was identified. The herniated sac stalk was cauterized and the encephalocele was excised. The defect was repaired with a temporalis fascia graft, and the mastoid cavity was obliterated with abdominal fat. The patient was confined to bed with a spinal drain for 72 hours. No further leakage was encountered.

Because of the patient's history of recurrent meningitis and suspicious CT findings, the contralateral or right mastoid was electively explored 6 months later. As the aditus ad antrum was opened, a bleb of tissue was identified. Dissection was then carried into the root of the zygoma. As this tissue was uncovered, it became evident that this was an encephalocele herniating through the tegmen tympani. There was active leakage of CSF. The necrotic tissue was cauterized and excised. The defect was repaired with temporalis fascia and the zygomatic root was obliterated with a temporalis muscle flap. Again, the patient's postoperative course was uneventful.

Figure 1. Coronal CT view of left temporal bone with bony dehiscence of the mastoid tegmen plate (arrow) and associated soft tissue density.

Figure 2. Coronal CT view of right temporal bone with marked thinning and questionable dehiscence of the tegmen tympani (arrow).

Case 2

A 57-year-old female with a history of recent serous otitis media presented after placement of a right-sided ventilation tube resulted in continuous clear otorrhea. Evaluation of this drainage had demonstrated a significant glucose content. The patient denied any history of head trauma or prior otologic problems.

Physical examination demonstrated a patent right-sided ventilation tube with pulsating clear discharge. Audiometric studies revealed a high-frequency sensorineural hearing loss on the right side and normal hearing on the left side.

A CT scan with intrathecal contrast demonstrated irregularity of the right mastoid tegmen plate with extravasation of contrast into the mastoid and external auditory canal.

The patient underwent a transmastoid exploration during which a tegmen tympani encephalocele was identified. The postauricular incision was extended superiorly, and a middle fossa craniotomy was performed. The defect was isolated extradurally, and the herniated tissue was cauterized and excised. A fascia graft was then laid over the bony defect within the middle fossa.

The patient did well postoperatively until 4 years later when she noted profuse clear rhinorrhea. This was confirmed on physical examination when the patient leaned forward. A CT scan with intrathecal contrast was obtained which demonstrated extravasation of contrast material into the right sphenoid sinus (Fig. 3). There was no evidence of leakage from the previously repaired temporal bone site.

A lumbar puncture demonstrated a marginally elevated opening pressure of 35 cm H₂O. This study was otherwise unremarkable. The patient underwent surgery during which a right frontotemporal approach provided defect exposure. The defect was localized to the roof of

the right side of the sphenoid sinus. Repair was accomplished with muscle and fascia grafts. Her postoperative course was uneventful, and there was no further evidence of CSF leakage.

DISCUSSION

While the etiology of spontaneous CSF leaks remains elusive, defining characteristics continue to be documented. From previously reported cases, we know that the vast majority of patients are adults, with a leakage site occurring distant from the labyrinth.^{3,4} Typically at the time of repair, an encephalocele is found herniating onto

Figure 3. Coronal CT view of the paranasal sinuses with intrathecal contrast demonstrating extravasation of contrast into the right sphenoid sinus (arrow).

the ossicular chain through a tegmen tympani defect. With the exception of four previously reported cases of multisite defects, these dural fistulas have largely been considered isolated occurrences. With the addition of the cases reported in this document, roughly 10% of all spontaneous CSF leak patients eventually demonstrate multisite involvement. These findings not only establish a disease pattern, but may also provide insight into the underlying pathophysiologic process.

The tegmen tympani, or roof of the middle ear, is the temporal bone's anatomic weak point. Bony dehiscence of this area is not uncommon. In 1965, Ahren and Thulin reported a 6% incidence of multiple tegmen tympani defects in an autopsy study.⁶ Yet, the frequency of accompanying dural fistula or encephalocele is rare in the absence of trauma, infection, or neoplastic disease. Even when bone is removed from the tegmen during mastoid surgery, the exposed dura rarely herniates. Additional predisposing or acquired factors are, therefore, implicated.

In 1990, Gacek presented a spontaneous CSF leak case involving the posterior fossa tegmen plate. Histologic examination of the resected encephalocele demonstrated the presence of arachnoid granulations.⁷ In the same study, Gacek reported an 8.5% histopathologic incidence of temporal bone arachnoid granulations (multiple and bilateral). Unfortunately, both the reported case and the temporal bone examinations in the Gacek study were limited to the posterior fossa tegmen plate. Only two other spontaneous cases reported in the literature have involved the posterior fossa plate.^{8,9} To date similar histopathologic evaluation of the middle fossa plate or tegmen tympani has not been performed.

Increased intracranial pressure has also been postulated as a contributing factor.^{1,2} In all previously and currently reported multisite cases, significant time periods existed between the presentation of initial and subsequent leakage sites. In theory, repair of the presenting leak could then lead to further or restored elevated intracranial pressure and subsequent encephalocele formation. Multisite cases reported by Rotilio and May demonstrated an elevated opening pressure at the time of lumbar puncture, with Rotilio's patient ultimately requiring ventricular peritoneal shunting.^{1,10} Our second case suggested a marginally elevated pressure.

Diagnosis of elevated intracranial pressure can be difficult in an actively leaking patient prior to repair. A dural fistula can act as a pressure valve, releasing CSF as the intracranial pressure increases. Imaging studies often fail to demonstrate enlarged ventricular size or dilatation for this reason. Whether increased intracranial pressure is a potential factor in all spontaneous CSF leak cases or limited to the subset of patients with multiple skull base defects is even more uncertain. Postoperative follow-up

lumbar puncture and imaging studies could resolve this issue.

Other proposed etiologic factors include low-grade inflammation and age-related dural changes.² Whether any one or a combination of these theoretical factors is responsible, multisite spontaneous cases are most likely associated with a generalized or nonfocal process. The cranial floor's weakest or naturally dehiscence areas are then, ultimately, the point of dural herniation or fistulization.

Because 46% of all spontaneous CSF leak cases have been reported over the past 4 years, the incidence of multisite leaks will probably continue to rise. Careful follow-up is, therefore, necessary. We recommend post-surgical clinical examination at 6 month intervals for the first 3 years, and then annually. Thus far, the longest time interval reported between leak site presentations is 4 years. Follow-up imaging studies are unnecessary unless clinically indicated. The preoperative or initial axial and coronal CT scan can serve as a baseline study, verifying remaining skull base integrity.

CONCLUSIONS

A significant percentage of spontaneous CSF leak patients eventually demonstrate second skull base leak site. Careful and regular follow-up is thus indicated. Although the etiology remains unclear, the presence of multisite involvement indicates an underlying nonfocal process. We must continue to document such cases to advance our understanding and management of this problem.

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