

New Considerations in the Cause of Spontaneous Cerebrospinal Fluid Otorrhea

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Objective: To examine the demographic and radiographic features of patients with spontaneous cerebrospinal fluid otorrhea and to determine whether they display similar characteristics to patients with idiopathic intracranial hypertension.

Study Design: Retrospective case review.

Setting: Academic, tertiary referral center.

Patients: All individuals presenting with spontaneous cerebrospinal fluid otorrhea, diagnosed between 2000 and 2009, undergoing primary surgical repair.

Interventions: All patients underwent surgical repair via a transmastoid, middle fossa, or combined transmastoid–middle fossa approach.

Main Outcome Measures: Patient demographics such as age, race, sex, height, weight, and body mass index, the presence of a radiographically empty or partially empty sella, and preoperative radiographic and intraoperative surgical findings of the temporal bone.

Results: Twenty-three patients underwent primary surgical repair for spontaneous cerebrospinal fluid otorrhea. Fifteen patients

underwent preoperative magnetic resonance imaging of the head with 12 (80%) demonstrating the presence of an empty or partially empty sella. Mean body mass index of those patients with an empty or partially empty sella was 38.0 kg/m² compared with 28.5 kg/m² for those without an empty sella.

Conclusion: Patients with spontaneous cerebrospinal fluid otorrhea are often middle-aged and obese, with females being affected nearly twice as often as males. Empty or partially empty sella was observed in 80% of patients with spontaneous cerebrospinal fluid otorrhea as demonstrated by preoperative magnetic resonance imaging. Patients with spontaneous cerebrospinal fluid otorrhea who display these demographic and radiographic features should be further evaluated for the presence of idiopathic intracranial hypertension. **Key Words:** Empty sella—Encephalocele—Idiopathic intracranial hypertension—Spontaneous cerebrospinal fluid otorrhea—Temporal bone.

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Cerebrospinal fluid (CSF) otorrhea is defined by the presence of CSF within the confines of the temporal bone. Trauma is responsible for most instances of CSF otorrhea, although iatrogenic, neoplastic, infectious, and congenital causes are also possible (1). Spontaneous CSF otorrhea refers to those cases in which none of these aforementioned causes can be found. Spontaneous CSF otorrhea typically occurs in 2 distinct populations: young children and middle-aged adults (2). Children presenting with spontaneous CSF otorrhea typically do so after an episode of meningitis. This is often due to congenital abnormalities of the lateral internal auditory canal, which leads to CSF leakage into the middle ear and predisposes the patient to meningitis after an episode of acute otitis media (3). Less common congenital anatomic abnormalities that may predispose to spontaneous CSF otorrhea include a

dehiscent fallopian canal or oval window, an enlarged cochlear aqueduct, or a defect in the Hyrtl fissure secondary to insufficient ossification (3–5). In adults with spontaneous CSF otorrhea, the presenting complaint is usually one of decreased hearing or aural fullness (6,7). In contrast to children, adults often have single or multiple defects of the tegmen mastoideum and/or tegmen tympani with normal inner ear anatomy (3). The diagnosis is often made after myringotomy for a persistent effusion with findings of clear fluid draining through the myringotomy site.

There is still significant uncertainty regarding the pathophysiology of spontaneous CSF otorrhea. Some suggest that small, congenital bony defects of the middle fossa tegmen enlarge over time secondary to constant CSF pressure (2). This enlargement leads to eventual dural herniation and subsequent bony and dural thinning with resultant CSF otorrhea. A second theory proposed by Gacek et al. (1) and Gacek (8) is that abnormally located arachnoid granulations are responsible for spontaneous CSF leaks observed within the temporal bone and paranasal sinuses. Autopsy studies of the temporal bone have

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shown a 21% incidence of arachnoid granulations along the middle fossa tegmen and a 9% incidence within the posterior fossa (8,9). The arachnoid granulations are thought to act as minor CSF reservoirs that, because of their abnormal location outside dural venous sinuses, are unable to properly return CSF to the venous system. Subsequent thinning and eventual erosion of bone is thought to ensue as a consequence of persistent pressure at the bony sites underlying the arachnoid granulations. The fact that the temporal bone is pneumatized beneath the thin plate of bone of the middle fossa tegmen explains why such a process could eventually lead to CSF within the mastoid or middle ear spaces. Although both of these theories might play a role in the pathogenesis of patients with spontaneous CSF otorrhea, recent evidence suggests that additional factors are involved.

Several recent studies have shown that a majority of patients with spontaneous CSF otorrhea are obese, middle-aged females (10–15). Schlosser et al. (16) have noted a similar demographic pattern among patients with spontaneous CSF rhinorrhea as well as a strong clinical and radiographic correlation between these patients and those with idiopathic intracranial hypertension (IIH). IIH previously known as benign intracranial hypertension, has been defined as a syndrome consisting of elevated intracranial pressure, without evidence of a mass lesion or hydrocephalus, with CSF that is normal in composition (17). It is predominantly found in obese females of child-bearing age (17). Studies of patients with spontaneous CSF rhinorrhea fitting this description also demonstrate findings of an empty or partially empty sella turcica in up to 100% (range, 63%–100%) of reported cases (18). Although approximately 5% to 6% of the general population will have evidence of a radiographic empty sella, this finding has been observed to a much greater degree in patients with IIH (approximately 70%) (12,16–18). This raises the question of whether patients with spontaneous CSF otorrhea represent an atypical phenotype of IIH.

The purpose of our study was to examine the demographic features as well as the radiographic and surgical findings of patients presenting with spontaneous CSF otorrhea. On the basis of the current body of evidence, we hypothesize that patients with spontaneous CSF otorrhea are much more likely to have an empty or partially empty sella compared with the reported population norms. In addition, we hypothesize that most patients with this condition will have a body mass index (BMI) >30.0 kg/m² and will be middle-aged, with females being affected more often than males. Examining demographics and radiologic findings of patients with spontaneous CSF otorrhea is critical to better understanding the pathophysiology of this process and has the potential to aid in the diagnosis and appropriate management of this complex condition.

MATERIALS AND METHODS

We performed a retrospective review of all patients with spontaneous CSF otorrhea undergoing primary surgical repair at a single tertiary referral center between January 2000 and May

2009. Patients with CSF otorrhea secondary to trauma, infection, cholesteatoma, and presumed iatrogenic insult, as well as those having undergone previous attempts at repair elsewhere, were excluded from the study population. A total of 23 patients met the inclusion criteria. This study was approved by the institutional review board of the Medical University of South Carolina (no. 19034).

Clinic and hospital charts, including anesthesia records and surgeon-prepared intraoperative reports, were retrospectively examined for all patients. Demographic information, including age, sex, race, height, weight, and BMI were recorded for each patient. In addition, presenting symptoms as well as the status of various medical comorbidities were noted for each patient. All available preoperative computed tomographic (CT) and magnetic resonance imaging (MRI) studies were examined for the presence or absence of empty or partially empty sella, ventricular enlargement, and bony or dural irregularities along the middle and posterior fossae. Computed tomographic scans were also evaluated for the number of distinct bony defects present within the middle and posterior cranial fossae. Surgeon-prepared operative reports were used to determine the surgical approach, repair method used (e.g., fascia, bone), number, side, size and location of the defect(s), and presence or absence of a meningoencephalocele.

Statistical Analysis

All data analyses were performed with SPSS 15.0 and Sigma Stat 3.5 (SPSS, Inc., Chicago, IL, USA). Categorical variables are presented as percentages (%), and continuous variables are presented as mean ± SD. *t* Tests were used to compare BMIs between those patients with empty or partially empty sella and those with a normal-appearing sella. Differences in BMI between male and female patients as well as racial groups were also examined. *p* < 0.05 was considered statistically significant for all statistical tests.

RESULTS

Demographic and Clinical Findings

Twenty-three patients underwent primary surgical repair for spontaneous CSF otorrhea during a 9-year period. Patients' mean age was 60 years (range, 42–76 yr) with 10 men (43.5%) and 13 women (56.5%). Fourteen patients (60.9%) were Caucasian, and 9 (39.1%) were African-American. Thirteen cases (56.5%) occurred on the right side, whereas 10 (43.5%) involved the left. Mean height was 66.3 inches (range, 61–71 inches), whereas mean weight was 103.7 kg (range, 79.0–158.7 kg). Mean BMI for the entire study group was 36.3 ± 6.7 kg/m². (Obesity is defined by a BMI > 30 kg/m²).

The mean BMI for the 10 men was 36.0 ± 8.4 kg/m², whereas the mean BMI of the 13 women was 36.5 ± 5.6 kg/m² (*p* = 0.873; Table 1). Caucasian patients had mean BMI of 35.4 ± 8.2 kg/m², whereas African-American patients had mean BMI of 37.7 ± 3.4 kg/m² (*p* = 0.428; Table 1).

In terms of clinical presentation, 2 patients reported a previous episode of meningitis. None of the patients, however, demonstrated any focal neurologic abnormalities preoperatively. Hearing loss and aural fullness were the most common presenting complaints. Additional patient symptoms included pulsatile tinnitus and headache. None

TABLE 1. Body mass index as a function of preoperative MRI findings of the sella turcica

	No. patients	Age, mean \pm SD, yr	F/M ratio	BMI, mean \pm SD, kg/m ²	<i>p</i> (age; BMI)
Sex					
Female	13	60.8 \pm 8.1	N/A	36.5 \pm 5.6	0.356; 0.873
Male	10	59.4 \pm 10.0		36.0 \pm 8.4	
Race					
Caucasian	14	61.2 \pm 8.0	7:7	35.4 \pm 8.2	0.216; 0.428
African American	9	58.7 \pm 10.2	6:3	37.7 \pm 3.4	
Status of sella					
Normal sella turcica	3 (20% ^a)	61.3 \pm 10.7	1:2	28.5 \pm 1.4	0.198; 0.034
Partially empty or empty sella turcica	12 (80% ^a)	62.7 \pm 9.0	9:3	38.0 \pm 6.7	

^aPercentage of patients with a preoperative MR image.

of the patients presented with CSF rhinorrhea, although several patients did present with CSF otorrhea before myringotomy. One patient also had a sphenoid meningoencephalocele that was repaired transnasally at a later date. Lumbar puncture opening pressures were not performed because patients were not subjected to lumbar drain placement before, during, or after surgery.

Radiographic Findings

Preoperative CT scan of the temporal bones was obtained in 17 of 23 patients, whereas 15 of 23 patients underwent preoperative MRI of the head. Of the 15 patients with a preoperative MR image, 12 (80%) demonstrated evidence of a partially empty or empty sella (Table 1). Picture archiving and communication system technology was used to make sellar measurements from MR images in the sagittal plane in all cases. Partially empty sella was defined by the presence of soft tissue of the pituitary gland occupying less than one half of the vertical height of the sella turcica, whereas empty sella was defined by the

presence of only a thin layer of pituitary tissue at the base of the sella turcica (Fig. 1). Patients demonstrating tissue occupying more than half of the sagittal height of the sella turcica were classified as having a normal sella. The mean BMI of those patients with a partially empty or empty sella turcica was 38.0 \pm 6.7 kg/m², a value that was significantly higher than the BMI of 28.5 \pm 1.4 kg/m² for those with a normal radiographic sella turcica (*p* = 0.034; Table 1). None of the patients with preoperative MR image had evidence of abnormal lateral ventricle size or other intracranial pathologic processes based on the accompanying report provided by a staff neuroradiologist.

Of the 17 patients who underwent high-resolution CT of the temporal bone, 8 (47%) demonstrated evidence of 2 or more bony defects, whereas all (100%) had evidence of one or more bony defects. No extra-axial masses were observed on any of the CT scans. In all cases, the non-contrasted, high-resolution temporal bone scans were not useful in identifying the status of the pituitary gland within the sella turcica.

Surgical Findings

All patients were treated surgically with a middle cranial fossa (MCF), a transmastoid, or a combined approach (Table 2). Details of these operative techniques have been described elsewhere (19). The most common site of a defect was the mastoid tegmen, demonstrating involvement at the time of surgery in 19 (82.6%) of 23 cases. Meningoencephaloceles were found in 15 (65.2%) of the 23 cases, whereas tissue suspected of being an arachnoid granulation was found in 2 cases (8.7%). Approximately 43% of patients (*n* = 10/23) demonstrated 2 or more separate bony defects at the time of surgery. A variety of substances and techniques were used to repair the defects. A layered closure using either temporalis fascia or perichondrium along with bone or cartilaginous support was used in more than 80% of cases. Surgical success was defined by a normal result from the otoscopic examination with tympanogram disclosing no abnormalities and/or negative repeat myringotomy for middle ear fluid at the 6- and 12-month postoperative time intervals. In all 23 patients, the CSF otorrhea was eliminated, although 3 patients required a second operative procedure. Of these 3 patients, 2 underwent a transmastoid approach, whereas the third underwent an MCF approach for the original repair. Two subsequently underwent an MCF approach,

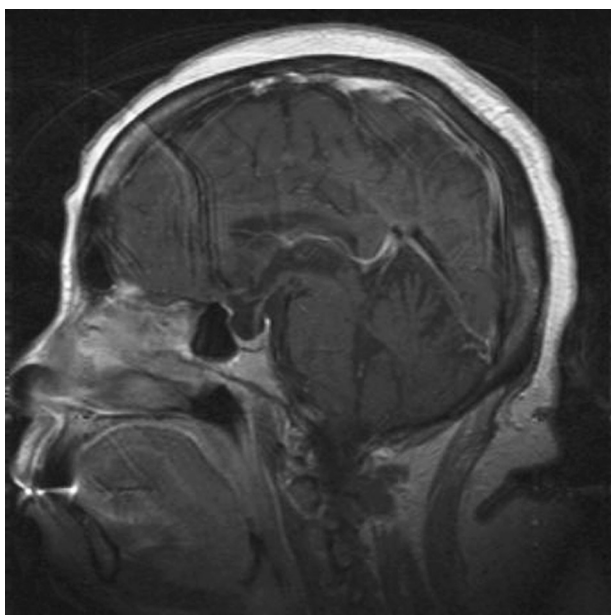


FIG. 1. Sagittal, T1-weighted, postcontrast MR image demonstrating findings of an empty sella turcica.

TABLE 2. Surgical approaches for repair of spontaneous CSF otorrhea

Surgical approach	No. patients	BMI, mean \pm SD, kg/m ²	Mean no. defects at surgery	Percentage with meningoencephalocele at surgery	Site of defect(s)
MCF	7	32.9 \pm 5.3	1.7	85.7	MT, antrum, epitympanum, TT
Transmastoid	14	39.2 \pm 6.6	1.4	57.1	MT, antrum, epitympanum, subarcuate cell tract
Combined MCF and transmastoid	2	30.5 \pm 0.78	1.0	50.0	MT

MT indicates mastoid tegmen; TT, tegmen tympani.

whereas the third underwent blind sac closure of the external auditory canal with plugging of the middle ear and eustachian tube. The mean BMI of these 3 patients was 35.3 kg/m², whereas their mean age was 56.6 years.

DISCUSSION

Spontaneous CSF otorrhea is an uncommon entity that requires a high index of clinical suspicion for proper diagnosis and management. Unlike traumatic, infectious, neoplastic, or iatrogenic CSF otorrhea, spontaneous leaks have been relegated to those cases without an obvious underlying cause. The management of patients with spontaneous CSF otorrhea is typically surgical, with previous studies as well as the current study demonstrating successful outcomes after appropriate repair (6–10). The size and location of the bony defect(s) as well as the presence of an encephalocele often factor into the surgical approach used. In the current study, as in previous studies, both the MCF and transmastoid approaches were used (Table 2) (12,13,15). Findings from the present study revealed encephaloceles in more than 50% of cases and one or more defects of the tegmen tympani, tegmen mastoideum, and/or antrum area (Table 2). These findings are comparable to previous reports (15,20). Although adjunctive treatments such as diuretic agents to reduce CSF pressure or CSF diversion tactics (either via lumbar drain or via ventriculostomy) have been described in cases of spontaneous CSF rhinorrhea, there are few data reported on such usage in cases of spontaneous CSF otorrhea (16,18). Yet, despite this well-established treatment method, the cause of “spontaneous” CSF otorrhea remains uncertain. In fact, it seems more likely that these cases are not truly “spontaneous” but rather result from pathophysiologic processes that have not yet been elucidated.

Various theories for the pathogenesis of spontaneous CSF otorrhea have previously been put forth. One theory suggests that congenital bony dehiscences in the area of the mastoid tegmen may predispose patients to eventual dural herniation and CSF leakage into the pneumatized spaces of the temporal bone (2). However, autopsy studies demonstrate that tegmen defects are present as much as 30% of the time, whereas the incidence of CSF otorrhea remains considerably lower (21). We found a mean age at presentation of 60 years, similar to the findings in a recent review by Nahas et al. (20) where the mean age at presentation was 56.5 years. Although congenital disorders

may present later in life, this demographic information suggests that an additional pathophysiologic process may be involved in these patients.

A second theory for the pathogenesis of spontaneous CSF otorrhea describes abnormally located arachnoid granulations as the cause (1,8). Normal CSF absorption into the venous system is dependent on properly functioning arachnoid villi, the small outpouchings of the subarachnoid space that lie within the dural venous sinuses. According to Gacek et al. (1) and Gacek (8), arachnoid granulations represent abnormally located arachnoid villi tissue, serving as a reservoir of CSF as well as a site of pressure transmission to the underlying bone. Although CSF pulsations likely occur over the entire cranial base, without any seemingly harmful sequelae, it is the constant pressure being applied to the thin cortex of an underlying pneumatized bone (such as the ethmoid, sphenoid, and temporal bones) through the aberrant arachnoid granulation that is thought to be responsible for the development of CSF otorrhea in these cases. Although this theory is certainly plausible, it must be recognized that compared with the studies presented by Gacek et al., the presence of arachnoid granulations at the site of temporal bone defects has not been found to the same degree in the current study or in recent studies (10–13). Admittedly, in the current study, microscopic analysis of tissue at the site of the bony defects was not performed in all cases. However, the relatively high incidence of arachnoid granulations within the posterior and MCF, as demonstrated in a previous autopsy study, suggests that their presence alone is insufficient to lead to clinically apparent CSF otorrhea (8,9).

We propose that “spontaneous” CSF leaks at the cranial base result from 2 pathologic processes, one anatomic and the second physiologic. Physiologically, an elevation of intracranial pressure seems to be present in most patients as evidenced by changes in the sella turcica and/or by direct CSF pressure measurements (22). Central venous obstruction and primary arachnoid villi dysfunction with decreased CSF resorption are 2 possible mechanisms that could cause increased CSF pressure. To date, no definitive pathologic processes of the arachnoid villi have been identified. Sigmoid or transverse sinus thromboses are classic examples of venous obstruction causing elevated central venous pressure and impairing the normal pressure gradient across the arachnoid villi. As in our cases, however, imaging studies in most patients with intracranial hypertension fail to show signs of central venous

thrombosis. A “functional” obstruction caused by hypercompliant and highly compressible venous sinuses has been postulated as a factor in some patients (23). In this scenario, a very mild elevation of intracranial pressure leads to compression of central venous sinuses, initiating a cycle resulting in more severe and sustained intracranial hypertension. Finally, obesity itself can elevate central venous pressure as discussed below. It is likely that many individuals have an elevation of intracranial pressure, yet spontaneous CSF leaks from the cranial base are uncommon. Thus, in addition to this physiologic factor, it is probable that an anatomic predisposition involving thinning of the cranial base is necessary. Such thinning or actual dehiscence may be congenital, secondary to an aberrant arachnoid granulation, or simply the result of chronic IIH. IIH usually develops in the fourth or fifth decade of life, whereas spontaneous CSF otorrhea (and rhinorrhea) tends to occur years later (20,24,25). This observation suggests that the process of a thinned area of the cranial base developing an actual dehiscence with CNS herniation takes years to occur. Once herniation is present, even more time would be required for the attenuated dura to break and leak spinal fluid.

Although the precise physiologic aberrancy in cases of spontaneous CSF otorrhea has not been identified, recent clinical studies have pointed to demographic features of patients with spontaneous CSF otorrhea as potential clues to the cause. More specifically, patients who develop spontaneous CSF otorrhea are typically middle-aged and obese (BMI > 30 kg/m²), with females being affected more often than males (6,11–15). In the current study, these same characteristics were prevalent among our spontaneous CSF otorrhea patients, with a mean age of 60 years, mean BMI of 36.3 kg/m², and a female predominance noted. Recent reports from the Centers for Disease Control and Prevention indicate that the mean BMI among U.S. adults approaches 28 kg/m² (26). Although we recognize that a control population of a similar demographic would provide a more meaningful comparison, there seems to be a noticeable increase in the BMI of patients with spontaneous CSF otorrhea compared with at least the general U.S. population. These clinical characteristics suggest that increasing age, obesity, and female sex all play a role in the development of spontaneous CSF otorrhea. In fact, patients with spontaneous CSF otorrhea display a similar demographic profile to those with spontaneous CSF rhinorrhea, with middle-aged, obese females characterizing most of these cases as well (16,18). Schlosser et al. (16) and Schlosser and Bolger (18) have demonstrated a noticeable association between patients with spontaneous CSF rhinorrhea and those with IIH, formerly known as benign intracranial hypertension. IIH is a well-known clinical entity characterized by elevated intracranial pressure without mass lesion or abnormalities in CSF composition (17). Patients with IIH often have visual disturbances, headaches, pulsatile tinnitus, and possibly endocrine dysfunction (17). Much like patients with CSF otorrhea or CSF rhinorrhea, patients with IIH tend to be middle-aged, obese females. However, this seemingly obvious associa-

tion of IIH with spontaneous CSF rhinorrhea and otorrhea has only recently been evaluated.

The modified Dandy criteria have been used to formally diagnose IIH in clinical practice (24). Strict adherence to the modified Dandy criteria requires measurement of the intracranial pressure, through either lumbar puncture or ventriculostomy, neither of which was performed in the current study. Nevertheless, the patients in the current study did meet several of the criteria, in particular the presence of an empty sella. Prichard et al. (12) examined the similarity between patients with IIH and spontaneous CSF otorrhea and found that 5 of 7 patients with spontaneous CSF otorrhea demonstrated findings of an empty or partially empty sella. Schlosser et al. (16) and Schlosser and Bolger (18) noted in their series that nearly all patients with spontaneous CSF rhinorrhea demonstrated MRI findings of an empty sella. In the present study, 12 (80%) of 15 patients with a preoperative MR image demonstrated an empty or partially empty sella compared with the 6% of the general population that reportedly demonstrates an empty sella (12,16). Studies investigating the MRI incidence of empty sella in subjects with normal sella are extremely limited, however, and the referenced “normative” incidence data is based on autopsy studies (27). We recognize that these methodological differences limit the ability to draw accurate comparisons. Nevertheless, the current study and previous reports do seem to suggest that the presence of an empty or partially empty sella is more common in patients with spontaneous CSF leakage.

Several theories have been put forth regarding the pathophysiology of IIH, with many suggesting that obesity, particularly central obesity, may be responsible (25,28). Central obesity is thought to increase intrathoracic pressure, which, in turn, leads to decreased venous return and ultimately increases cerebral venous and CSF pressure. Others have suggested narrowing of cerebral venous sinuses to be the underlying cause of IIH, whereas some have noted a strong association with polycystic ovarian disease, possibly suggesting a hormonal dysfunction (29,30). Despite continued research, the exact mechanisms involved in the development of IIH remain unknown. We acknowledge that the possibility of a multifactorial cause must be entertained given the nature of this complex condition. However, the striking similarity between patients with CSF otorrhea and those with IIH, as observed in the current study, suggests that the underlying pathophysiology may in fact be similar in both patient populations. As a result, patients with spontaneous CSF otorrhea should be considered for ophthalmologic and neurologic evaluation to identify other, potentially occult, problems associated with IIH. One must keep in mind that an IIH patient who is actively leaking CSF (in the form of either CSF otorrhea or CSF rhinorrhea) may not have the classic signs or symptoms of IIH secondary to the reduction in intracranial fluid pressure as a consequence of the leak. After the repair of the leak, however, the pressure relief valve is removed, and IIH may then become manifest. In addition to ophthalmologic referral to check

visual fields and visual acuity on a regular basis, neurologic and possibly neurosurgical referral is necessary to document elevated intracranial pressure via lumbar puncture. Consideration of a diuretic, such as acetazolamide, is often given in the management of IIH and may be important in patients with IIH having undergone surgical repair of a “spontaneous” CSF leak to try and lessen the degree of intracranial hypertension (31). Complications of IIH are generally related to loss of vision and should be actively identified. Although medical management through the use of steroids may play a role in temporarily halting a rapidly progressing visual loss, some patients may require lumboperitoneal or ventriculoperitoneal shunt procedures or possibly optic nerve sheath fenestration to prevent potentially irreversible visual loss (31).

Limitations of the current study include its retrospective nature, relatively small sample size, and the incompleteness of the radiographic data for the subjects involved. The lack of a meaningful and comparable control group is an additional weakness that might be addressed in future studies through the use of a prospective study process.

CONCLUSION

To date, this study is the largest series to investigate the relationship among spontaneous CSF otorrhea, MRI findings, and IIH. Demographic data from our study revealed a preponderance of obesity and female sex, which is similar to patients with IIH. Empty or partially empty sella was observed in 80% of patients with spontaneous CSF otorrhea in the current study, further linking these patients to those with IIH. We propose that an underlying pathophysiologic process, combined with an underlying anatomic defect (i.e., a congenital bony dehiscence or aberrant arachnoid granulation), is likely responsible for most cases of “spontaneous” CSF otorrhea. Patients with spontaneous CSF otorrhea, especially those who display these demographic and radiographic features, should be evaluated for IIH by the appropriate personnel, such as a neurologist and ophthalmologist. Further studies on the precise physiologic abnormalities underlying both IIH and spontaneous CSF otorrhea will aid in the management of patients with this challenging condition.

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