Multiple Spontaneous Skull Base Cerebrospinal Fluid Leaks: A Single Institution Experience Considering Risk Factors, Recurrence, and Surgical Considerations

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Abstract

Spontaneous cerebrospinal fluid (CSF) leakage from a skull base defect has been considered uncommon, but recent studies have suggested an increasing identification of these leaks and increasing rates of surgical intervention over the past twenty years. These patients may present with otorrhea or rhinorrhea secondary to a connection between the subarachnoid space and the tympanomastoid cavity of the lateral skull base or the sinonasal corridor via the anterior skull base. Spontaneous CSF leakage falls under the guise of nontraumatic etiologies for CSF leak. It is believed that patients with spontaneous CSF leaks without any other apparent etiology may have elevated intracranial pressure. This suggests a possible association with idiopathic intracranial hypertension (IIH). These patients may be obese, suffer from obstructive sleep apnea, and are often female. There have been several prior case reports that have discussed patients with spontaneous CSF leaks to identify a link with specific risk factors and risk of recurrence. A few studies have suggested an association with IIH, but there remains a population of patients with spontaneous CSF leaks without IIH and identifying their risk factors is important. Currently, we have cared for seven patients with multiple spontaneous CSF leaks with associated skull base defects. Our aim is to delineate our institutional experience caring for patients with multiple CSF leaks with associated skull base defects, possible associated risk factors, timeline for surgical intervention, and recurrence.

Introduction

Cerebrospinal fluid leakage is a rare phenomenon and patients with a CSF leak may present with a variety of symptoms. These symptoms include postural headaches, mastoid fluid collection, otorrhea, rhinorrhea, combined oto-rhinorrhea, tinnitus, or rarely meningitis. There are a variety of etiologies for CSF leaks, and this includes both traumatic and atraumatic. The atraumatic or spontaneous CSF leaks often occur at the anterior or lateral skull base. When this occurs, there is a communication with the sinonasal corridor of the anterior skull base leading to rhinorrhea or a communication with the tympanomastoid cavity of the lateral skull base that may present with otorrhea. The risk factors associated with a an atraumatic or spontaneous CSF leak include obesity, idiopathic intracranial hypertension, and obstructive sleep apnea. A definitive association between these risk factors and development of a spontaneous CSF leak has not been proven, but there are papers that suggest a correlation. Given the rarity of one CSF leak, identifying the underlying etiology for multiple CSF leaks would potentially suggest an underlying process predisposing these patients to CSF leakage. Ultimately, identification of the site(s) of CSF leak requires significant scrutiny of the history, clinical presentation, and imaging. Once identified, the necessary surgical approach must be identified and then a multidisciplinary approach is often undertaken for the surgical intervention. The surgical interventions include endonasal endoscopic and open cranial approaches. Patients who undergo surgical repair of their CSF leak are then followed as these patients may be at risk for recurrent CSF leak given a possible underlying process leading to their presentations.

Methods and Materials

This is a retrospective chart review of a single surgeon case series of patients treated at one institution with surgical intervention.

Results

Seven patients were identified at our institution who presented with multiple skull base defects leading to CSF leak. The sites included anterior defects of the cribriform plate, lateral sphenoid, and posterior table of the frontal sinus. Lateral skull base defects can occur along the petrous temporal bone and included tegmen tympani, tegmen mastoideum, and semicircular canal dehiscence.

Six of the patients were female and the average age of patients was 56.5 (36-71). The rate of recurrence was 42.8% (3/7). The average BMI was 39.8 (25-66), rate of IIH was was 42.8% (3/7), and the rate of OSA was 57.1% (4/7). Meningitis occurred in 28.6% (2/7) patients.

Discussion

Spontaneous CSF leaks are a rare clinical finding, but rarer is the finding of multiple spontaneous CSF leaks in the same patient. In our experience, nearly all the patients were obese and female, with these individuals also having obesity associated phenomena of IIH and OSA. Through prior case series and one review, literature has found correlations between intracranial hypertension and CSF leaks. One theory suggests that the CSF leak is a way for the body to compensate for the elevated intracranial pressure. OSA has been evaluated, and findings suggest that longer periods of apnea have been associated with higher elevations in ICP. These sustained periods of ICP elevation may start the cycle that leads to an IIH phenotype in these patients. The role of IIH in CSF leaks has been further studied over the past couple decades with the increasing prevalence and identification of spontaneous CSF leaks and skull base defects. While the role of IIH has been consistently implicated, skull base repair without significant recurrence rate in the absence of IIH treatment would suggest additional factors play a role. In the review study, 20% of their patients underwent management of their IIH. In this case series, our study included patients with recurrences that required further intervention, but these were not exclusive to the patients with IIH. The heterogeneity of presentation, diagnosis, and clinical findings in these patients combined with low numbers makes it difficult to define the causative conditions that lead to their clinical presentation. Given this, further work will need to endeavor to define the patient population that necessitates IIH intervention after skull base repair to prevent recurrence.

Conclusions

The increasing identification of spontaneous skull base CSF leaks has led to an increased prevalence of surgery to repair these leaks. This has led to the observation that recurrent spontaneous CSF leaks may have modifiable patient factors that contribute. Further research regarding the role of obesity, OSA, and IIH is warranted.

Table 1. Recurrent CSF Leak Patient Information

Age	Site of Leak 1	Site of Leak 2	Recurrence	Meningitis	IIH	BMI	OSA	Gender	Imaging
55	Rt MCF	Rt MCF	Y	Y	Y	59	Y	F	Right tegmen defect and posterior mastoidectomy/petrous ridge
61	Rt SSCD	Lt SSCD	N	N	N	25	Y	F	Right tegmen tympani likely dehiscent and right and left semicircular canals are dehiscent
36	Lt MCF	Lt MCF	Y	N	N	32	N	F	Tegmen dehiscence
66	Lt Cribriform plate	Lateral Sphenoid defect	N	N	Y	39	N	F	Left cribriform plate/Ethmoid roof
48	Rt MCF	Sphenoid	N	N	Y	66	Y	F	Right frontal sinus inner table defect with encephalocele, Left nasoethmoid defect with meningocele, Bilateral sphenoid wing with connection to sphenoid sinus, Bilateral tegmen tympani and mastoideum defects,
59	Lt Post Table Frontal Sinus Encephalocele	Lt MCF	N	N	N	27	N	F	Left frontoethmoidal encephalocele; Left tegmen mastoideum
71	Lt MCF	Rt MCF	Y	Y	N	31	Y	M	Right tegmen defect and bilateral semicircular canal dehiscence

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