Idiopathic Intracranial Hypertension Presenting as Cerebrospinal Fluid Otorrhea

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Abstract

Educational Objective: At the conclusion of this presentation, the participants should be able to 1) recognize spontaneous cerebrospinal fluid (CSF) otorrhea as a possible presenting symptom of idiopathic intracranial hypertension; and 2) identify patients presenting with CSF otorrhea who warrant further evaluation for idiopathic intracranial hypertension and potential medical management.

Objectives:
1) Recognize spontaneous cerebrospinal fluid (CSF) otorrhea as a possible presentation of idiopathic intracranial hypertension; and 2) identify patients presenting with CSF otorrhea who warrant further evaluation for idiopathic intracranial hypertension and potential medical management.

Study Design: Case report and systematic review of the literature.

Methods: Case report of intracranial hypertension presenting as unilateral hearing loss and CSF otorrhea with associated review of the literature.

Results: A 27 year old obese female presented to a tertiary neurootology clinic with unilateral right sided hearing loss and pulsatile tinnitus. Associated symptoms included debilitating headaches and blurred vision. On exam she had a thin, clear fluid filling the right middle ear associated with a 25 dB conductive hearing loss. CT scan showed right sided focal dehiscence of the tegmen tympani and opacification of the middle ear and mastoid. Further testing revealed bilateral papillae dema with lumbar puncture opening pressure of 570 mmH2O (normal 60-250 mmH2O). Idiopathic intracranial hypertension was diagnosed and medical management with a loop diuretic and carbonic anhydrase inhibitor was initiated. Within 3 months of medical management she had symptom improvement and full resolution of CSF otorrhea. Review of the literature found 67-80% of patients presenting with spontaneous CSF otorrhea may have idiopathic intracranial hypertension. Prior reports focus on surgical intervention, but medical management of patients with CSF otorrhea secondary to idiopathic intracranial hypertension has been described.

Conclusions: Idiopathic intracranial hypertension can be associated with spontaneous CSF otorrhea. Further evaluation of these patients is warranted, and in a select subgroup a trial of medical therapy prior to surgical intervention may be reasonable.

Introduction

Idiopathic intracranial hypertension is identified in 67 – 80% of patients presenting with spontaneous CSF otorrhea. 1-3 Although the traditional treatment for spontaneous CSF otorrhea has been surgical intervention, in the rhinology literature there have been reports of successful resolution of CSF rhinorrhea associated with idiopathic intracranial hypertension after medical management along.3-5

Case Report

A 27 year old female presented to a neurology clinic for evaluation of new onset unilateral hearing loss of less than one month duration. Associated symptoms of several months duration included pulsatile tinnitus, constant debilitating headache, blurred vision, and recent significant weight gain. She had no history of acute illness, otologic disease, or prior otologic surgery. She had been seen at an outside emergency department and evaluated for meningitis with reportedly normal findings on lumbar puncture. Examination showed an obese female with a BMI of 41. The right middle ear was opacified with a clear, thin fluid. Nasopharyngoscopy was normal. Audiogram showed a right-sided mixed hearing loss, most severe in the middle frequencies to a maximum of 55dB. The left side showed a mild sensorineural hearing loss, worst in the middle frequencies. Ophthalmologic exam showed papillae dema (2+) bilaterally.

Radiographic examination with a MRI showed flattening of the optic discs (Figure 1) and opacification of the right mastoid with CSF isointense fluid (Figure 2). A fine cut CT scan of the temporal bone revealed an irregular pitting of the skull base with two areas of focal dehiscence in the tegmen tympani (Figure 3). No other intracranial processes or masses were identified.

Due to concerns for possible intracranial hypertension a repeat lumbar puncture was performed and opening pressure was markedly elevated at 570 mm of water (normal <250 mm). No other abnormalities were identified on laboratory assessment of the CSF fluid.

Thus, the patient was diagnosed with idiopathic intracranial hypertension with associated CSF otorrhea. Medical management was initiated for the underlying idiopathic intracranial hypertension including a loop diuretic, carbonic anhydrase inhibitor, and weight loss plan.

Within 1 month of starting medical management the patient experienced significant improvement in symptoms of headache and visual loss. Within 3 months there was near complete resolution of her otologic symptoms with improved hearing and decreased tinnitus. A myringotomy performed at 3 months showed resolution of fluid within the right middle ear space. At one year the patient has had no recurrence of her otologic symptoms and continues with medical management of her idiopathic intracranial hypertension.

Discussion

Several theories regarding the etiology of spontaneous CSF otorrhea have been postulated including (1) progressive enlargement of congenital defects of the middle fossa tegmen (2) abnormally located arachnoid granulations within the temporal bone. Idiopathic intracranial hypertension (previously termed pseudotumor cerebri or benign intracranial hypertension) has recently been postulated to play a role in spontaneous CSF otorrhea; whereby, elevated intracranial pressure may act upon congenital defects of the skull base with resultant CSF leakage.

Coexisting idiopathic intracranial hypertension has been identified in 67-80% of patients presenting with spontaneous CSF otorrhea.4-5 Patients presenting with spontaneous CSF otorrhea warrant workup for idiopathic intracranial hypertension (Table 1). Diagnostic investigation includes ophthalmologic and neurologic evaluation with possible lumbar puncture. Initial evaluation for intracranial hypertension may be normal in patients where the CSF otorrhea acts as a pressure relief valve for the CSF system. As a result, intracranial hypertension may not become symptomatic until after surgical repair of spontaneous CSF otorrhea. Thus, these patients warrant continued follow-up and possibly repeat testing for idiopathic intracranial hypertension following surgical repair of their CSF otorrhea.

The traditional treatment for spontaneous CSF otorrhea has been surgical repair by a transmastoid, middle fossa, or otologic surgery. She had been seen at an outside emergency department and evaluated for meningitis with reportedly normal findings on lumbar puncture. Further intervention, in the rhinology literature there have been reports of successful resolution of CSF rhinorrhea associated with idiopathic intracranial hypertension after medical management along.3-5

Table 1

Modified Dandy Criteria for Diagnosis of Idiopathic Intracranial Hypertension*

1. Signs and symptoms of increased intracranial pressure (headaches, nausea, vomiting, transient obscurations of vision, papilledema).
2. No localizing neurologic signs otherwise, with the single exception being unilateral or bilateral VI nerve paresis.
3. CSF can show increased pressure, but no cytologic or chemical abnormalities otherwise.
4. Normal to small symmetric ventricles must be demonstrated (originally required ventriculography, but now demonstrated by CT).

Conclusions

• Spontaneous CSF otorrhea is often associated with idiopathic intracranial hypertension.

• Evaluation for idiopathic intracranial hypertension is recommended in patients presenting with spontaneous CSF leaks.

• Treatment of idiopathic intracranial hypertension may be appropriate prior to surgical repair of spontaneous CSF otorrhea.

• Further research is needed to determine if treatment of coexistent idiopathic intracranial hypertension has a role in definitive therapy for CSF otorrhea.

References