

# Resolution of Chronic Aspiration Pneumonitis Following Endoscopic Endonasal Repair of Spontaneous Cerebrospinal Fluid Fistula of the Skull Base

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## Abstract

Spontaneous cerebrospinal fluid (CSF) rhinorrhea due to a skull base defect requires prompt diagnosis and treatment. Multiple surgical options are available for repairing the fistula, including the two-layer “fascial apposition” method and use of a pedicled nasal-septal flap. A 44-year-old obese woman presented with 4 months of progressive cough, exertional dyspnea, hoarseness, and intermittent fluid drainage from the right nostril. Chest computed tomography (CT) imaging and bronchoscopy showed chronic pneumonitis, which was confirmed by pulmonary wedge resection. CT and magnetic resonance imaging of the skull base, as well as laboratory analysis of the nasal fluid for  $\beta$ 2-transferrin, confirmed a skull base defect causing CSF rhinorrhea. During surgery, insertion of a lumbar drain with the intrathecal fluorescein administration was performed, followed by endoscopic endonasal repair using an autologous fascial apposition graft and pedicled nasal-septal flap. Both the CSF leak and the pulmonary complications resolved following the operation with no symptoms at 11-month follow-up. This is the first reported case of spontaneous CSF rhinorrhea complicated by chronic aspiration and pneumonitis. Increased diagnostic complexity due to chronic pulmonary complications resulted in unnecessary interventions and treatment delays. Prompt recognition of spontaneous CSF leaks is essential to prevent potentially harmful complications.

## Keywords

- ▶ cerebrospinal fluid
- ▶ pneumonitis
- ▶ endoscopic
- ▶ endonasal
- ▶ aspiration

## Introduction

Spontaneous cerebrospinal fluid (CSF) fistula of the skull base is an uncommon condition associated with multiple etiologies and can have a variable presentation. Of note, obese, middle-aged, and multiparous women most commonly develop this condition.<sup>1,2</sup> Intractable, watery discharge from the nose (rhinorrhea) that tests positive for  $\beta$ 2-transferrin is pathognomonic.<sup>1</sup> However, the quality and quantity of

discharge may result in misdiagnosis as allergic rhinitis or sinusitis and delay treatment, particularly if the condition presents with atypical complications. We present the first known case of spontaneous CSF rhinorrhea complicated by chronic pneumonitis secondary to CSF aspiration, which was successfully resolved by endoscopic endonasal repair, and discuss the relevant literature associated with the complications of spontaneous CSF rhinorrhea.

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## Case Report

### History and Presentation

A 44-year-old woman presented with 4 months of progressive cough, exertional dyspnea, hoarseness, and intermittent fluid drainage from the right nostril. She was initially evaluated by a pulmonologist, who noted that she was otherwise healthy besides being obese (body mass index: 36.5 kg/m<sup>2</sup>) and having diminished lung sounds bilaterally at the bases. Computed tomography (CT) imaging of the chest (►Fig. 1) showed patchy opacities in both lower lung lobes. Bronchoscopy with lavage was performed with nondiagnostic results. Multiple courses of oral antibiotics were unsuccessful in improving her symptoms. She was also prescribed codeine, formoterol/budesonide, prednisone, albuterol, montelukast, and fluticasone in various unsuccessful attempts to alleviate her pulmonary symptoms.

Because of the persistent pneumonitis, she was referred to a thoracic surgeon for further evaluation. Due to the uncertain, persistent, and the progressive nature of her lung findings, a thoracoscopic wedge resection of the superior right upper lobe was recommended and performed. The specimen showed acute bronchopneumonia, a 1.5-mm chemodectoma, and acute on chronic inflammation, but no further etiology was determined.

She was then referred to an otolaryngologist for evaluation of the persistent nasal drainage and postnasal drip, which was a major stimulant of her cough and severe enough to

force her to sleep upright. The nasal fluid was more pronounced during a forward tilt test, and was collected and tested positive for  $\beta$ 2-transferrin. This was an unexpected finding, as a spontaneous CSF fistula with chronic aspiration has not been previously recognized as a cause of lung disease.

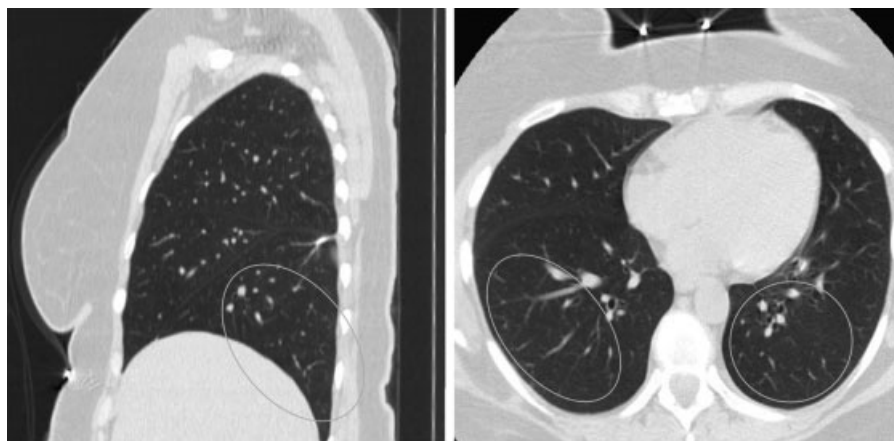
The patient was referred to our otolaryngology and neurosurgery services for evaluation of her spontaneous CSF rhinorrhea. She denied any trauma or previous nasal surgery. She was afebrile with no meningitic or focal neurological signs. She had clear and persistent fluid rhinorrhea from the right nostril when leaning forward. Endoscopic evaluation of her nasal cavity in the clinic setting did not identify a site of CSF leakage. CT imaging of the paranasal sinuses (►Fig. 1) showed bony dehiscence of the skull base and a meningocele extending into the right ethmoid sinus. Although the cause of her spontaneous CSF leak and skull base defect was unknown, it was likely related to her body habitus and idiopathic intracranial hypertension. The patient was informed of her treatment options and elected for urgent surgical repair.

### Operation and Postoperative Course

The CSF leak was repaired via a right endoscopic endonasal approach. Following administration of intrathecal fluorescein through a lumbar drain, a small bony defect and brisk CSF fistula were identified in the region of the right fovea ethmoidalis. The bony and dural defects were repaired using a two-layer reconstruction with autologous fascia lata (fascial apposition method) followed by a pedicled nasal-septal flap.



**Fig. 1** Preoperative CT image of the paranasal sinuses (A) showing meningocele with fluid collection in the right ethmoid sinus (arrows) and of the chest (B) showing lung opacities (circles). CT, computed tomography.



**Fig. 2** Postoperative CT imaging of the chest showing resolution of the lung opacities (circles). CT, computed tomography.

The patient's postoperative course was unremarkable with no further CSF rhinorrhea. The lumbar drain was removed on postoperative day 3. Following surgery, her pneumonitis resolved and she remained free of any pulmonary symptoms at 11-month follow-up. CT imaging of the chest (► **Fig. 2**) showed resolution of the bilateral opacities and pneumonitis.

## Discussion

Diagnosis and treatment of spontaneous CSF rhinorrhea are straightforward, but can be delayed significantly if it presents with atypical complications. Upon review of the literature, the most common and clinically relevant complications are meningitis and pneumocephalus.<sup>3,4</sup> Meningitis is of primary concern for all CSF leaks regardless of origin. Nasopharyngeal CSF leaks are especially prominent sources of meningitis, as pneumococci are responsible for greater than 80% of CSF leak-related meningitis.<sup>5</sup> Other complications, such as intermittent herniation, empty sella syndrome, and spontaneous intracranial hypotension are much rarer.<sup>6–8</sup> Our report of chronic pneumonitis due to CSF aspiration is the first reported instance of this significant and clinically relevant complication.

While our case did not present this issue, difficulties with identifying the skull base defect on standard neuroimaging studies can also result in treatment delay.<sup>9</sup> CT imaging is standard and magnetic resonance imaging can be used if CT results are nondiagnostic.<sup>10</sup> However, it remains that some scans will not clearly identify the responsible skull base defect.<sup>11</sup> Regardless of imaging,  $\beta$ 2-transferrin positive nasal discharge is sufficient cause for endoscopic endonasal exploration and repair<sup>1</sup>; use of intrathecal fluorescein dye will help identify the leak under direct visualization.<sup>10</sup>

Once the location of the CSF fistula is accurately identified, multiple surgical repair options exist. If a meningoencephalocele is identified, it should be reduced and repaired accordingly. Autologous fat and fascial grafting is commonly used to fill in any dead space and create a dural reconstruction, respectively. We prefer to utilize a two-layer “fascial apposition” method, in which the first fascial layer is inserted as a dural inlay and the second is placed in apposition as a dural

overlay and bony inlay.<sup>12</sup> We prefer not to use a rigid buttress unless the defect is exceptionally large, but routinely prepare and rotate a pedicled nasal-septal flap for broad coverage of the defect as described by Hadad et al,<sup>13</sup> and popularized by Kassam et al.<sup>14</sup> Lumbar drainage may be utilized conservatively to temporarily reduce CSF pressure for 2 to 3 days following the operation along with facilitating intraoperative intrathecal fluorescein administration.

In this case, the increased diagnostic complexity related to the pronounced pulmonary complications resulted in unnecessary interventions and treatment delays. This is a meaningful example of why prompt recognition of spontaneous CSF leaks by practitioners of all types is essential to prevent potentially harmful complications including meningitis, pneumocephalus, and even lung disease caused by chronic CSF aspiration.

## Note

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this article. The contents of this article have not been published or presented previously.

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