Evolving Treatment of Frontal Sinus Cholesteatoma: A Case Report

Nizar Tejani, MD; Rijul Kashirsagar, MD; Brian Song, MD; Jonathan Liang, MD

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CLINICAL MEDICINE

ABSTRACT

Introduction: Cholesteatomas are lined by squamous epithelium, contain keratin debris, and can cause bony erosion. Although commonly found in the middle ear space and mastoid, cholesteatomas may develop in adjacent structures including the paranasal sinuses. Frontal sinus cholesteatoma (FSC) is a rare condition with fewer than 30 reported cases. The aims of this study are to describe the clinical presentation, diagnostic imaging, and endoscopic treatment of FSC and to review the literature focusing on the pathogenesis, diagnosis, and historical and contemporary treatments of FSC.

Case Presentation: A 45-year-old man presented with a 1-week history of right eyelid and forehead swelling. Results of computed tomography scans and magnetic resonance images revealed a right frontal sinus lesion of soft-tissue density with bony dehiscence along the superior orbit and posterior table. He underwent right-sided endoscopic sinus surgery at a tertiary care center in January 2017. Intraoperatively, the frontal sinus contained keratin debris suggestive of FSC. This suspicion was confirmed postoperatively by pathologic analysis after subtotal resection.

Discussion: The pathogenesis of frontal sinus cholesteatoma varies based on its type (congenital vs acquired). Clinical diagnosis remains challenging but is aided by nasal endoscopy, computed tomography, and magnetic resonance imaging. Historically, FSC has been managed by total extirpation through open approaches, which can entail substantial morbidity. With sophisticated endoscopic sinus instrumentation and image guidance, FSC can be successfully treated via an endoscopic approach. Serial débridements and washouts in an outpatient setting may adequately manage the residual disease in the postoperative period.

INTRODUCTION

Cholesteatomas, otherwise known as epidermal inclusion cysts or keratomas, are characterized by an accumulation of keratin debris surrounded by squamous epithelial cells. Commonly found in the middle ear and mastoid, cholesteatomas can infrequently develop in the paranasal sinuses and neighboring structures, including the pterygopalatine space and petrous apex of the temporal bone. Although a rare site for cholesteatoma, the frontal sinus is the most common location for paranasal cholesteatomas, followed by the ethmoid and maxillary sinuses.

Frontal sinus cholesteatoma (FSC) is rare, with fewer than 30 reported cases. First described in the English literature by Spencer in 1930, this disease presents as a soft, unilateral mass on the forehead. Patients may complain of frontal headache, swelling, and vision changes. Left untreated, it can lead to severe complications, including bacterial meningocerebritis, loss of vision, disfiguration, malignant degeneration, and death.

The treatment of this disease is surgical resection. Historically, FSC has been treated with open approaches. However, with the advent of endoscopic sinus surgical techniques and stereotactic intraoperative image guidance technology, this disease can be treated with minimally invasive surgical techniques.

This case study describes the presentation, imaging, and endoscopic treatment of FSC, with a review of the literature focusing on pathogenesis, diagnosis, and the historical and contemporary treatment.

CASE PRESENTATION

Presenting Concerns

A 45-year-old man presented with a 1-week history of progressive swelling of the right eyelid and forehead. The patient denied any pain or visual deficit. He stated that he has had an irregular, raised bump on his head since he was a child that had never been medically or surgically treated. In addition, he recalled sustaining repeated head trauma in childhood.

Results of the physical examination revealed a 2 cm × 2 cm area of swelling of the right forehead with a palpable, underlying 1 cm × 1 cm defect of the frontal anterior table. No clinically significant findings were seen on nasal endoscopy.

Preoperative computed tomography (CT) and magnetic resonance imaging (MRI) were performed. Findings on the right frontal sinus revealed an expansile lesion of soft-tissue density with marked bony remodeling and multifocal bone dehiscence along the anterolateral superior orbit (2.3 cm) and posterior table.

Figure 1. Preoperative computed tomography scans (A. axial and B. coronal planes, respectively) demonstrating the expansile lesion (frontal sinus cholesteatoma) extending into the frontal sinus. There is bony remodeling with dehiscence of the posterior table (asterisk) and orbital roof (caret, ^).

Figure 2. Intraoperative image guidance technology, this disease can be treated with minimally invasive surgical techniques.

Keywords: cholesteatoma, endoscopic sinus surgery, frontal sinus cholesteatoma (FSC), rhinology

Author Affiliations

1 Department of Otolaryngology, Head and Neck Surgery, Louisiana State University Health Sciences Center, Shreveport
2 Department of Otolaryngology, Head and Neck Surgery, Kaiser Permanente Oakland Medical Center, CA

Corresponding Author

Nizar Tejani, MD (nizartejani@gmail.com)

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without intradural or intraorbital extension (Figures 1 and 2). There were no overlying inflammatory changes of the scalp.

**Therapeutic Intervention and Treatment**

After a course of preoperative tapered-dose prednisone, the patient was taken to the operating room for right-sided endoscopic maxillary antrostomy, total ethmoidectomy, sphenoidotomy, and frontal sinusotomy. Intraoperatively, the right frontal sinus was noted to have an expansile lesion with white keratin debris (Figure 3). Because of limited access via the endoscopic approach, keratin debris of the superolateral right frontal sinus could not be completely débrided, and a small portion was left in situ. The right frontal sinus was also noted to have underlying polypoid mucosa. There was substantial broad dehiscence of the posterior table, focal (1 cm × 1.5 cm) dehiscence of the lateral anterior table, and focal dehiscence of the right medial orbital roof. The posterior table dehiscence was notable for exposed dura without evidence of a cerebrospinal fluid leak or intradural keratin spread. Cultures yielded light growth of *Moraxella catarrhalis* and normal respiratory flora. The pathologic findings demonstrated abundant keratin debris, consistent with cholesteatoma (Figure 4).

**Follow-up and Outcomes**

Postoperatively, the patient was discharged home the same day and was prescribed an oral course of corticosteroids and ciprofloxacin. He was instructed to use nasal saline irrigation at least once daily. His forehead swelling resolved, and he had no neurologic or ophthalmologic sequelae. He was followed-up for 1 year and underwent serial in-office débridements using rigid endoscopy (Figure 5) with further washout of residual keratin debris. A consulting neurosurgeon recommended conservative management of the posterior table defect with observation because postoperative imaging findings demonstrated near-total removal of FSC (Figures 6 and 7).

One year after his surgery, the patient presented to the office with increasing, intermittent pressure over the right side of the forehead. This was particularly worse after his daily saline irrigation. Results of nasal endoscopy demonstrated a patent frontal sinus outflow tract with no evidence of keratin debris and moderate polypoid swelling. The patient was prescribed budesonide nasal rinses to reduce the inflammation. He was instructed to seek follow-up care if his symptoms worsened or did not improve. His latest follow-up visit was 2.5 years after surgery, and nasal endoscopy findings showed some mild keratin debris in the frontal sinus, which was managed with outpatient monitoring and washout.

**DISCUSSION**

**Pathogenesis**

FSC can be separated into primary (congenital) and secondary (acquired) causes. The pathogenesis of primary cholesteatoma is attributed to aberrant embedding of ectodermal epithelial cells during the fusion of epidermal surfaces. This is thought to occur during development of the face between the third and fifth weeks of embryonic development.\(^1,5\) The pathogenesis of acquired cholesteatomas is less clear; 3 mechanisms have been described: Implantation by trauma or surgery, migration of cells from the nasal vestibule or external auditory canal, and squamous metaplasia of epithelium caused by chronic inflammation.\(^1,5\) A history of trauma or surgery has been reported in only a few cases of FSC.\(^4\) The cause of this patient’s cholesteatoma was not apparent. The presence of his bony forehead prominence since birth suggests a primary cause. Yet, the history of repeated head trauma in childhood and polypoid changes intraoperatively indicate acquired disease caused by either traumatic implantation or chronic inflammation.

**Diagnosis**

The diagnosis of FSC requires clinical suspicion coupled with supportive radiologic and intraoperative findings. A
Patient had a 1-year postoperative visit for endoscopy with débridement. He had noticed mild intermittent pressure in the right frontal area, prompting evaluation in the otolaryngology clinic. A maxillofacial CT scan without contrast enhancement was obtained.

Patient had onset of right eyelid and forehead swelling. He underwent right endoscopic maxillary antrostomy, total ethmoidectomy, sphenoidotomy, and frontal sinusotomy. Pathologic findings showed residual keratin debris (k) with surrounding inflammatory mucosa.

A definitive diagnosis is made pathologically. A review of the literature published in 2007 examined 12 cases of FSC and found common clinical symptoms, including frontal headache, proptosis, orbital and supraorbital edema, blurry vision, and loss of vision. Ophthalmologic symptoms were attributed to mass effect. The duration of symptoms for these cases varied widely from 10 days to 20 years.

The patient described in this report was initially thought to have a frontal sinus mucocele. This preoperative diagnosis is common in reported cases of FSC. Mucoceles demonstrate a similar constellation of symptoms and bony erosion on imaging studies. Furthermore, 65% of mucoceles are found in the frontal sinus.

Other differential diagnoses include chronic rhinosinusitis, fibroma, osteoma, dermoid cyst, and malignancy. In addition to a thorough history and physical examination, nasal endoscopy, CT, and MRI can aid in the initial workup because cholesteatomas characteristically demonstrate bony erosion on imaging. An MRI may be useful in distinguishing sinusitis from a tumor or soft-tissue lesion.

**Management**

FSC has been historically managed by total extirpation via open approaches, including trephination and osteoplastic flaps. Before the availability of advanced imaging techniques such as CT and MRI, the open approach was often necessary to visualize the extent of bony erosion and dural involvement as well as to treat far-reaching lateral disease. However, open approaches can lead to substantial morbidity, including recurrence of disease, mucocele formation, chronic osteitis, frontal neuralgia, violation of the dura, hematomata, and disfigurements caused by loss of bone.

An endoscopic approach to resection of FSC is now widely favored because advancements in endoscopic sinus surgical techniques can provide sufficient exposure for removal of disease. Furthermore, such techniques can facilitate functional mucociliary clearance to enable adequate drainage of the sinus cavity. Although the endoscopic approach also holds risks of bleeding and dural injury, the widespread use of stereotactic image guidance in sinus surgery enhances precision and mitigates risk of injury to critical intracranial and extracranial structures. Additionally, this approach offers superior cosmetic results.

Nevertheless, the open approach may still be necessary in the modern age. Cases have been reported in which severe intracranial complications occurred after endoscopic surgery, thereby requiring subsequent open resection. In one reported case, the authors initially attempted subtotal endoscopic resection. However, after 10 months of serial débridements, the patient returned with worsening symptoms and was taken back to the operating room for open resection.

### Table 1. Timeline of the case

<table>
<thead>
<tr>
<th>Date</th>
<th>Event</th>
</tr>
</thead>
<tbody>
<tr>
<td>12/29/2016</td>
<td>Patient had onset of right eyelid and forehead swelling</td>
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<tr>
<td>1/5/2017</td>
<td>Evaluation in the otolaryngology clinic; maxillofacial CT scan without contrast enhancement obtained</td>
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<tr>
<td>1/12/2017</td>
<td>Brain MRI with and without contrast agent obtained</td>
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<tr>
<td>1/16/2017</td>
<td>Patient underwent right endoscopic maxillary antrostomy, total ethmoidectomy, sphenoidotomy, and frontal sinusotomy. pathologic findings were consistent with cholesteatoma</td>
</tr>
<tr>
<td>1/30/2017</td>
<td>Patient had a 2-week postoperative visit for endoscopy with débridement; no recurrent symptoms, and patient was recovering well</td>
</tr>
<tr>
<td>2/15/2017</td>
<td>CT of head without contrast enhancement obtained. MRI of orbit, face, and neck with and without contrast agent obtained</td>
</tr>
<tr>
<td>2/16/2017</td>
<td>Patient had a 1-month postoperative visit for endoscopy with débridement and frontal sinus washout. There were no recurrent symptoms, and patient was recovering well. Neurosurgery consultation: No dural defects seen on MRI, and no reconstruction needed</td>
</tr>
<tr>
<td>1/12/2017</td>
<td>Patient had a 1-year postoperative visit for endoscopy with débridement: He had noticed mild intermittent pressure in the right frontal area, but endoscopic image shows patent frontal outflow tract. Corticosteroid nasal rinses were started to reduce inflammation</td>
</tr>
<tr>
<td>3/1/2018</td>
<td>Patient had not reported any recurrent symptoms</td>
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</table>

CT = computed tomography; MRI = magnetic resonance imaging.
In our patient, the FSC was managed via a purely endoscopic approach. Although lateral frontal recess access was limited, an endoscopic approach allowed for near-total removal of FSC with minimal morbidity. Postoperative CT of the head, a useful tool in detecting recurrent disease, was ordered. Our patient also underwent postoperative MRI of the brain at the recommendation of the consulting neurosurgeon to assess the need for graft placement over the exposed dura. This imaging study may be useful in patients with dehiscence of the posterior table of the frontal sinus. Although certain authors advocate for a second-look surgery if keratinization of the frontal sinus mucosa is present, this was not deemed necessary for our patient. Residual keratin debris in the lateral recess was managed with postoperative débridement and washout.

Recurrence has been reported in the literature, particularly in cases of partial resection. Rapid recurrence of signs and symptoms may ominously indicate carcinomatous degeneration.

There are no well-defined recommendations for the frequency and length of follow-up of disease in FSC. We recommend regular monitoring after endoscopic subtotal resection. Postoperative follow-up of patients may continue until the patient is devoid of clinically significant symptoms. Recurrence of symptoms warrants in-office rigid and flexible endoscopy for adequate visualization. We used non-contrast-enhanced CT to monitor disease progressive and bony erosion, but MRI may also be considered, because it has been traditionally used to monitor cholesteatoma of the middle ear.

CONCLUSION

FSC is a rare entity that commonly presents as a frontal headache with ocular symptoms. This study illustrates that in patients with FSC, endoscopic resection with serial office débridements can be a reliable and less morbid treatment than traditional open approaches. Long-term follow-up and a larger series of patients will help clarify whether endoscopic resection with serial débridement is an acceptable alternative to open approaches for treatment of FSC.

Disclosure Statement
The author(s) have no conflicts of interest to disclose.

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How to Cite this Article

References