

Atypical Presentation of Silent Sinus Syndrome: A Case Report and Literature Review

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Abstract

Introduction: Silent sinus syndrome (SSS) is a condition characterized by ophthalmologic features, such as spontaneous enophthalmos and hypoglobus with ipsilateral maxillary sinus atelectasis and an otherwise asymptomatic presentation. SSS has been documented secondary to a number of external causes, including trauma or surgery, but has less commonly been described in the setting of a potential mass in the deep masticator space.

Case Presentation: A 56-year-old woman with a history of chronic headaches with normal prior sinonasal imaging presented with increasing right-sided facial pain and headaches that radiated to her occiput, subjective visual changes, sharp ear pain, and long-standing subjective diminished sense of smell. Physical examination was normal, while nasal endoscopy demonstrated lateral bowing of the medial maxillary wall on the right. Magnetic resonance imaging demonstrated a homogenous 2 × 2 × 2.4 cm T1- and T2-weighted, hyperintense mass lesion in the deep masticator space splaying the right medial and lateral pterygoid muscles concerning for a possible lipomatous lesion. Computed tomography revealed an atelectatic and opacified maxillary sinus with inward bowing of the posterior maxillary wall and increased orbital volume on that side. Endoscopic maxillary antrostomy was performed with biopsy of the retromaxillary space lesion and with near immediate resolution of the patient's symptoms. Histologic examination of the mass demonstrated mature adipose tissue with few aggregates of benign small vessels.

Discussion: This is an unusual presentation of SSS, with an accompanying enlargement of the retromaxillary fat pad. We herein review our clinical experience with SSS and provide a literature review of the presentation, management, and perioperative considerations for SSS.

Keywords

silent sinus syndrome, chronic maxillary atelectasis, chronic sinusitis, paranasal sinuses, maxillary antrostomy

Introduction

Although the constellation of symptoms was described in the literature as early as 1964, the term silent sinus syndrome (SSS) itself was first coined by Soparkar et al. in 1994.^{1,2} The clinical findings of SSS are that of spontaneous enophthalmos and hypoglobus associated with ipsilateral maxillary sinus atelectasis in the absence of other sinus symptoms.¹ The pathophysiology of SSS remains debated but is largely believed to be due to low-grade inflammation of the maxillary sinus resulting in hypoventilation of the sinus walls and, subsequently, the accumulation of secretions within the sinus.^{3,4} Continued resorption of gas from the sinuses then leads to a negative pressure gradient and inward

bowing of the sinus walls, often leading to the diagnostic ophthalmologic symptoms of enophthalmos and hypoglobus.^{3,4}

SSS is similar to and often mistaken for the clinical phenomenon of chronic maxillary atelectasis (CMA), as

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both share the symptomatology of hypoglobus and enophthalmos in the setting of an atelectatic maxillary sinus.⁵⁻⁷ Although debated in the literature, these conditions are believed to lie on a clinical spectrum, in which SSS presents in the absence of symptoms of chronic sinusitis, and CMA may involve symptoms of chronic rhinosinusitis (CRS).^{5,7} Some also use the term CMA to refer to cases of SSS secondary to external causes, such as those with a history of sinusitis, facial surgery, or trauma.^{1,4,5,7} To our knowledge, SSS/CMA is uncommonly described in the setting of an accompanying potential mass in the deep masticator space. In this study, we sought to review the diverse presentations, radiologic findings, and clinical management of SSS and CMA.

Methods

Study Population and Chart Review

We performed a retrospective chart review that was approved by the Johns Hopkins institutional review board (IRB00216775). Patients were identified through a review of the electronic medical record from 2009 to 2019 using the keyword “silent sinus syndrome.” The medical charts were reviewed for each patient, and data regarding patient demographics, medical history, clinical presentation, perioperative management, imaging, and postsurgical outcomes were collected. Clinical presentation and management were noted for each patient. Written informed consent was obtained for the individual case presentation.

Clinical Diagnosis and Data Analysis

Patients with the diagnosis of “silent sinus syndrome” in the medical record were further defined based on study criteria for SSS versus CMA. These definitions were similar to the criteria used by Brandt and Wright and Vander Meer et al. for CMA and SSS, respectively (Table 1).^{5,7}

A history of sinus symptoms or CRS is a common association with CMA, in contrast to SSS.⁷ CRS is defined by as sinonasal inflammation persisting for at least 12 weeks and is characterized by the cardinal

symptoms of nasal obstruction/congestion/blockage, mucopurulent nasal drainage, facial pain/pressure/fullness, and a decreased or loss of sense of smell.⁸ As there is variability in the chart review for documentation of a history of chronic sinusitis as a potential etiology of SSS, we chose to identify such patients using radiologic evidence of chronic paranasal sinus mucosal thickening. Patients who had evidence of mucosal thickening in the maxillary sinus in addition to at least 1 other sinus were considered to have a potential history of CRS and thus CMA and not SSS. Patients with a history of sinonasal symptomatology who did not meet imaging criteria for CRS were considered within the SSS group. Patients who did not meet the criteria for either CMA or SSS were excluded from the study.

Imaging Review

Patients with available computed tomography (CT) imaging were evaluated by an otolaryngologist for evidence of CRS (as defined above), the presence of increased fat in the retromaxillary space, the presence of a distinct retromaxillary or pterygopalatine fossa mass, deviated septum, and hypoglobus. Patients without dedicated sinonasal CT imaging available for review were excluded from the study.

Results

Case Presentation

A 56-year-old woman with a history of chronic headaches presented with increasing right-sided facial pain, subjective visual changes, a long-standing diminished sense of smell, and acute headaches uncontrolled with over the counter pain medications. She had no history of CRS, nasal obstruction or drainage, sinus surgeries, or vision problems.

She had a distant history of facial trauma, having been hit in the face with a baseball in childhood. However, between this incident and the presentation of her headaches, the patient had previously undergone a CT scan of the sinuses that revealed normal maxillary sinus volume bilaterally and was without mucosal thickening in the sinuses. On physical examination, the patient was found to have normal extraocular movements and minimal right-sided enophthalmos. Otherwise, the remainder of a comprehensive physical examination was normal. Nasal endoscopy revealed deviation of the right medial maxillary wall laterally, with otherwise healthy mucosa bilaterally.

Prior to her presentation to the otolaryngology service, that patient underwent a noncontrast-enhanced magnetic resonance imaging (MRI), which on T1-weighted and T2-weighted imaging demonstrated

Table 1. Diagnostic Criteria for SSS and CMA.

CMA	1. Enophthalmos and/or hypoglobus 2. Maxillary sinus opacification noted on CT imaging
SSS	1. Enophthalmos and/or hypoglobus 2. Maxillary sinus atelectasis or opacification on CT imaging 3. Absence of radiologic findings of CRS on CT imaging

Abbreviations: CMA, chronic maxillary atelectasis; CRS, chronic rhinosinusitis; CT, computed tomography; SSS, Silent sinus syndrome

a homogenous, hyperintense retromaxillary and pterygopalatine fossa fat signal concerning for a lipomatous lesion (Figure 1). These findings had not been seen on a prior MRI. On noncontrast-enhanced CT, both coronal and axial images (Figure 2) displayed an opacified, atelectatic right maxillary sinus, expansion of the right orbit, and homogenous fat density in the retromaxillary space and pterygopalatine fossa.

Subsequently, the patient opted to undergo surgical intervention consisting of endoscopic maxillary antrostomy and biopsy of the retromaxillary space tissue, which was uneventful. Biopsy of the tissue revealed mature adipose tissue with few aggregates of benign small vessels. Postoperatively, the patient had near immediate resolution of her symptoms.

Demographics

The retrospective review identified 45 patients with a chart diagnosis of “silent sinus syndrome.” Thirty-five of these patients had imaging available for review. Only 31 of these patients met study criteria for SSS or CMA. Of the 31 patients included in the study, there were 14 females (45.2%) and 17 males (54.8%). The age of diagnosis of SSS/CMA in the chart was available for 29 of the 31 patients, with the average age at diagnosis being 49.8 years (standard deviation = 15.9), the youngest age being 19 years, and the oldest age being 91 years.

Imaging Findings

Twenty-six of the 31 patients (83.9%) did not have findings of CRS on CT imaging, thus meeting the study

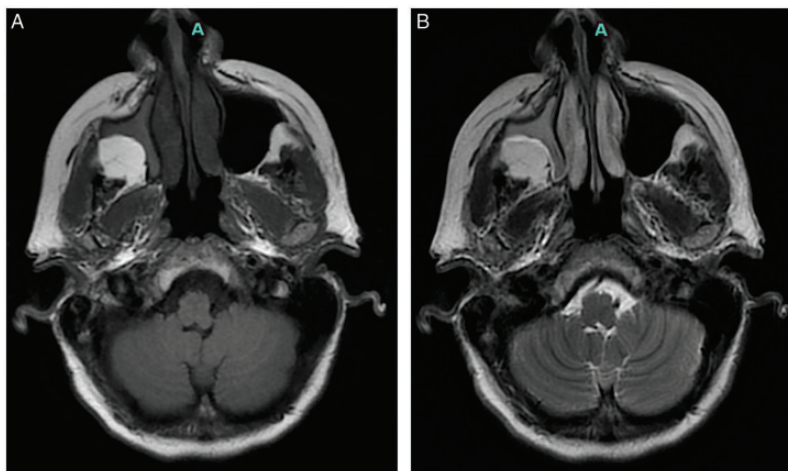


Figure 1. Retromaxillary lipomatous lesion. Noncontrast-enhanced MRI with (A) T1-weighted and (B) T2-weighted axial images demonstrating homogenous, hyperintense retromaxillary and pterygopalatine fossa fat signal.

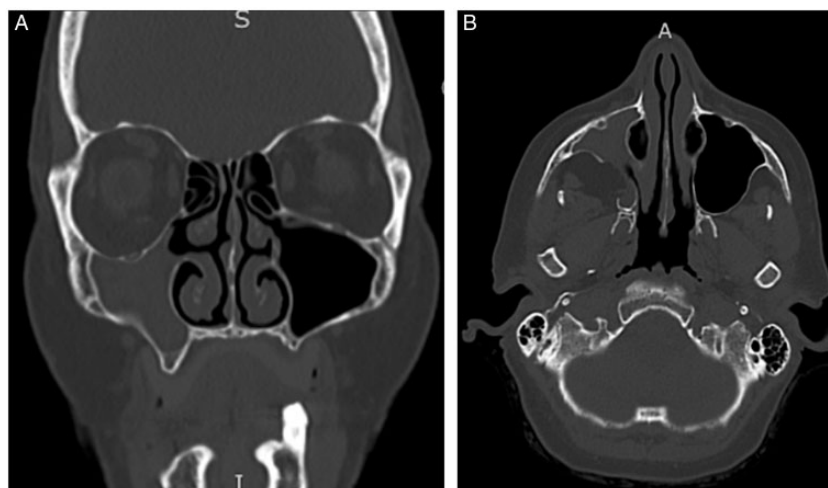


Figure 2. Right silent sinus syndrome. (A) Coronal and (B) axial noncontrast-enhanced CT displaying opacified, atelectatic right maxillary sinus, expansion of the right orbit, and homogenous fat density in the retromaxillary space and pterygopalatine fossa.

criteria for SSS. Within this group of 26 patients, 20 (76.9%) had findings of mucosal thickening in the maxillary sinus alone and 6 (23.1%) did not have evidence of mucosal thickening in any of the sinuses. The remaining 5 patients (16.1%) had radiologic findings of CRS in both the maxillary sinus and at least 1 additional sinus. These 5 patients met the study criteria for CMA.

Laterality of the affected sinus, as well as the presence of a deviated septum, mild increase of fat or liposity in the retromaxillary space, and significant retromaxillary or pterygopalatine fossa fat were determined on CT imaging for patients in the SSS group (Table 2). Of the 3 patients with significant retromaxillary or pterygopalatine fossa fat seen on CT imaging, only 1 patient (the presented case) had relevant pathology for this tissue, which revealed normal retromaxillary space fat.

Presence of Additional Symptoms

Three of the 26 patients (11.5%) had a history of sinus surgery, and 4 unique patients (15.4%) had a noted history of facial trauma, prior to their diagnosis of SSS. Overall, 6 patients (23.1%) reported an episode of acute sinusitis within 1 year of their clinical presentation of SSS. Nine of the 26 patients (34.6%) had a history of smoking.

The presence of specific symptoms, such as facial pain or pressure, headaches, nasal obstruction and/or congestion, nasal drainage, and decreased smell and/or taste, is reported in Table 3. Additional ophthalmologic symptoms are also documented in Table 3.

Clinical and Surgical Management

Sixteen of the 26 patients (61.5%) underwent sinus surgery, which included maxillary antrostomy for SSS. Ten patients (38.5%) did not undergo sinus surgery. Three patients (11.5%) underwent surgical ophthalmologic management: 2 underwent orbital implant insertion and 1 underwent an orbitotomy with orbital volume augmentation. The 2 patients who underwent an orbital

implant insertion also underwent a maxillary antrostomy several months prior to their orbital surgery, while the patient who underwent orbitotomy did not undergo sinus surgery.

Discussion

Although there is variation in the literature regarding the exact definitions and nomenclature of SSS versus CMA, it is acknowledged that both SSS and CMA encompass the clinical presentation of enophthalmos or hypoglobus in association with a contracted ipsilateral maxillary sinus.^{3,5} Additionally, the term CMA has been used to describe such presentations secondary to external etiologies as well as those with symptomatology similar to chronic sinusitis.⁵ The entities of SSS and CMA also stand in contrast to primary maxillary sinus hypoplasia, which may involve a hypoplastic or absent uncinate process, abnormalities of the lateral nasal wall and ostiomeatal complex, and subsequent total opacification of the affected sinus.^{9,10} Although sometimes related to chronic sinusitis, maxillary sinus hypoplasia is often an incidental imaging finding and is distinguished from SSS and CMA by the absence of diagnostic ophthalmologic findings.^{9,10}

Despite these definitions, various diagnostic criteria have been utilized by literature reviews in order to distinguish the diagnosis of SSS and CMA.^{5,7,11} In this study, we used the presence of CRS findings on CT imaging in other sinuses other than the affected maxillary sinus as a proxy to exclude patients with a clinical history of CRS and chose to then describe the clinical symptomatology found in the remaining patient population. We found that even among this patient population, many patients categorized as “silent sinus syndrome” actually presented with a CMA picture and symptoms of CRS. This is consistent with Brandt’s literature review where 85% of patients categorized as SSS also met diagnostic criteria of CMA.⁶ Among our group of 26 SSS patients, we found that 60.9% of patients were categorized as SSS patients reported facial pain or pressure, 13.6% reported decreased smell, 43.5% reported

Table 2. Radiographic Findings for SSS.

CT Imaging Finding	Number of Patients (%)
Left-sided maxillary atelectasis	10/26 (38.5)
Right-sided maxillary atelectasis	16/26 (61.5)
Grossly deviated septum	15/26 (57.7)
Septal deviation in direction of affected sinus	11/15 (73.3)
Increased liposity in retromaxillary space	19/26 (73.1)
Significant retromaxillary/pterygopalatine fossa fat	3/26 (11.5)

Abbreviation: CT, computed tomography.

Table 3. Presence of Additional Symptoms in SSS.

Reported Symptom	Number of Patients (%)
Facial pain or pressure	14/23 (60.9)
Headaches	11/24 (45.8)
Nasal obstruction and/or congestion	10/23 (43.5)
Nasal drainage	7/22 (31.8)
Decreased smell and/or taste	3/22 (13.6)
Diplopia	8/25 (32.0)
Abnormal extraocular movements	0/26 (0)
Changes in visual acuity	0/25 (0)

nasal obstruction or congestion, and 31.8% reported nasal drainage. There is significant overlap between CMA and SSS, and the presence or absence of symptoms cannot be used as the sole distinguishing feature between these disease entities.

The patient of interest described in the case presentation was initially referred to us for a possible mass lesion in the retromaxillary and deep masticator space seen on MRI. Further workup revealed that the patient had SSS and biopsy of the lesion confirmed, histologically benign fatty tissue. We found 2 additional patients in our review with bowing of the posterior maxillary wall and a significant increase in the retromaxillary fat pad. This could potentially be interpreted as a lipomatous mass as it was when referred to us. Additionally, 73.1% of the patients in our review had some degree of increased fat present in the retromaxillary space on imaging when compared to the contralateral, normal, side. This corresponding increase in fat signal is a documented radiologic finding in SSS. For example, Rose et al. noted that 9 of the 13 cases of SSS with an inward/inferiorly caving maxillary roof and concave medial and posterolateral maxillary sinus walls had a concomitant increase in the radiolucency of the pterygopalatine fossa on CT imaging when the posterolateral maxillary wall was involved.¹¹ Kohn et al. evaluated 22 patients with CMA retrospectively and found that all patients had increased infratemporal fossa (ITF) fat.¹² They found an inverse correlation between ITF fat and affected maxillary sinus, $r = -.53$ ($P < .05$).¹² This is likely a compensatory mechanism for loss of volume of the maxilla. Three of our patients had a significant increase of fat in this area on imaging, which may be related to the severity of maxillary sinus atelectasis.

With regard to the demographics of SSS, several literature reviews and case presentations have reported an average age of diagnosis of approximately 40 years, with no bias toward laterality or gender.^{3,11,13} We report similar findings, with an average age of diagnosis of 49.8 years, 45.2% female patients, and 38.5% having left-sided maxillary disease. Deviation of the nasal septum toward the affected maxillary sinus has also been commonly described in SSS, with Rose et al. reporting this in 83% of SSS patients with a septal deviation.¹¹ In this study, we found that 73.3% of patients with a septal deviation had a deviation in the direction of the affected maxillary sinus. It is unclear if a deviated septum has a role in the pathogenesis, is a result of this disease process, or is completely incidental.

In this study, we additionally attempted to capture the ocular symptoms of SSS/CMA. Diplopia is a common symptom reported in SSS, with 28.0% of 84 patients described by Numa et al. presenting with diplopia.³ In this study, we found that 32.0% of patients with available data reported diplopia. With regard to other

ophthalmologic symptoms, no patients in our study exhibited abnormal extraocular movements or a decrease in visual acuity.

A final goal of our study was to describe the clinical management of SSS. Although historically, the surgical management of SSS was a Caldwell-Luc procedure, SSS is now routinely managed with endoscopic sinus surgery.³ The surgery is technically challenging secondary to increased risk of orbital injury. The uncinate is atelectatic and the orbital floor is oftentimes at a lower position within the maxillary sinus. Meticulous surgical technique is critical, and image guidance navigation may help to delineate the lamina papyracea and orbital floor. Of the 2 main methods of uncinectomy, the swing-door technique where the uncinate is taken down in a posterior to anterior fashion is likely safer than the traditional anteroposterior uncinectomy.^{14,15} Before performing the swing-door uncinectomy, a ball-tip seeker, or other blunt instrumentation, should be used to separate the uncinate from the lamina papyracea. In cases with a severely atelectatic maxillary sinus, frontal sinus instruments and angled endoscopes may need to be used to adequately perform the surgery. In the end, a wide ostium will ensure proper aeration of the maxillary sinus and prevent recurrence.

In our study population, 61.5% of patients underwent endoscopic maxillary antrostomy, while 38.5% of patients did not undergo a surgical intervention. Three total patients underwent either orbital implant, following maxillary antrostomy, or orbitotomy with orbital volume augmentation. It is noted that no matter the specific nomenclature assigned to the maxillary atelectasis or the diversity of symptoms associated with the clinical presentation, the standard of care is to remove the maxillary sinus ostium obstruction via uncinectomy and maxillary antrostomy. Moreover, there is increasing support in the literature that orbital reconstruction in patients with SSS/CMA is largely unnecessary as most cases resolve with sinus surgery alone.^{4,16} Significant bony remodeling, with return of the orbital contents to its normal dimensions, can occur after relieving the negative pressure.^{16,17}

Conclusion

Often confused in the literature, SSS and CMA encompass the clinical entity of enophthalmos and/or hypoglobus associated with ipsilateral maxillary atelectasis. In this study, we presented a patient initially thought to have a retromaxillary mass who was found to have more pronounced retromaxillary fat in the setting of SSS. We suspect that this is likely related to a compensatory increase in liposity in the retromaxillary space in SSS patients. Moreover, we reviewed the diversity of symptom presentation and the patient population of

SSS/CMA, including the presence of diplopia, headaches, and predilection of the disease for nonsmokers. Given the range of clinical presentations associated with SSS/CMA, it is imperative that clinicians have a high degree of suspicion for SSS/CMA, and a thorough understanding of the disease process, in patients with spontaneous unilateral maxillary disease and increased liposity of the retromaxillary space such that they may anticipate the need for further otolaryngology and ophthalmologic management.

Declaration of Conflicting Interests

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Ethical Approval

Our institution does not require ethical approval for reporting individual cases or case series.

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Statement of Human and Animal Rights


This article does not contain any studies with human or animal subjects.

Statement of Informed Consent

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

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