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CLINICAL CASE

Frontal Mucocele with Intracranial Extension Causing Frontal Lobe Syndrome

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ABSTRACT

Purpose. Mucoceles are mucus-containing cysts that form in paranasal sinuses; although mucoceles themselves are benign, this case report highlights the extensive damage they can cause as their expansion may lead to bony erosion and extension of the mucocele into the orbit and cranium; it also presents a rarely reported instance of frontal sinus mucocele leading to frontal lobe syndrome. A thorough discussion and review of mucoceles is included.

Case Report. A 68-year-old white man presented with intermittent diplopia and a pressure sensation in the right eye. He had a history of chronic sinusitis and had had endoscopic sinus surgery 5 years prior. A maxillofacial computed tomography scan revealed a large right frontal sinus mucocele, which had caused erosion along the medial wall of the right orbit and the outer and inner tables of the right frontal sinus. The mucocele had protruded both into the right orbit and intracranially, causing mass effect on the frontal lobe, which led to frontal lobe syndrome. The patient was successfully treated with endoscopic right ethmoidectomy, radial frontal sinusotomy, marsupialization of the mucocele, and transcutaneous irrigation.

Conclusions. Paranasal sinus mucoceles may expand and lead to bony erosion and can become very invasive in surrounding structures such as the orbit and cranium. This case not only exhibits a very rare presentation of frontal sinus mucocele with intracranial extension and frontal lobe mass effect causing a frontal lobe syndrome but also demonstrates many of the ocular and visual complications commonly associated with paranasal sinus mucoceles. Early identification and surgical intervention is vital for preventing and reducing morbidity associated with invasive mucoceles, and the patient must be followed regularly to monitor for recurrence.

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Key Words: mucocele, frontal sinus, sinusitis, diplopia, orbital extension, intracranial extension, frontal lobe syndrome

Mucoceles are mucus-containing cystic cavities lined with pseudo-stratified columnar epithelium that develop in paranasal sinuses.[1] Sinus ostia obstruction leads to the entrapment of mucus-secreting cells, which encase and expand as the cells continue to expel mucus.[2] The frontal sinus is most frequently involved.[1,3] As the mucocele enlarges to become more space occupying and compressive, it may lead to demineralization and erosion of the sinus walls, which allows the mucocele to protrude into adjacent structures. Because of the proximity of the paranasal sinuses to the orbits, both the mucocele itself and its surgical eradication present potential ocular complications.

This case highlights an extensive mucocele, which not only exhibited many of the more common clinical features associated with mucoceles progressing into the orbit but also involved less-commonly encountered intracranial extension and very rarely associated mass effect on the frontal lobe causing frontal lobe syndrome; this case therefore underlines the extensive involvement potential of mucoceles and appropriateness of swift intervention. It additionally provides a comprehensive, yet concise, discussion, which serves as a relevant information reference for clinicians.

CASE REPORT

A 68-year-old white man presented for an eye examination with a chief complaint of diplopia. He reported that the onset of diplopia had been 1 month prior; it was intermittent and presented about every other day with a duration of a few hours at a time. The diplopia had both a horizontal and vertical component, and it happened more with extended reading or computer use. He denied headaches but described a pressure-type sensation in the right eye, emphasizing that it ''feels different'' than the left eye, and said it had felt that way since the onset of the diplopia. The patient had noticeable ptosis on the right, but he reported that it had been like that for years with no noted change in lid position.

The patient's last eye examination had been 9 months prior, and his ocular history was significant for a retinal detachment in the right

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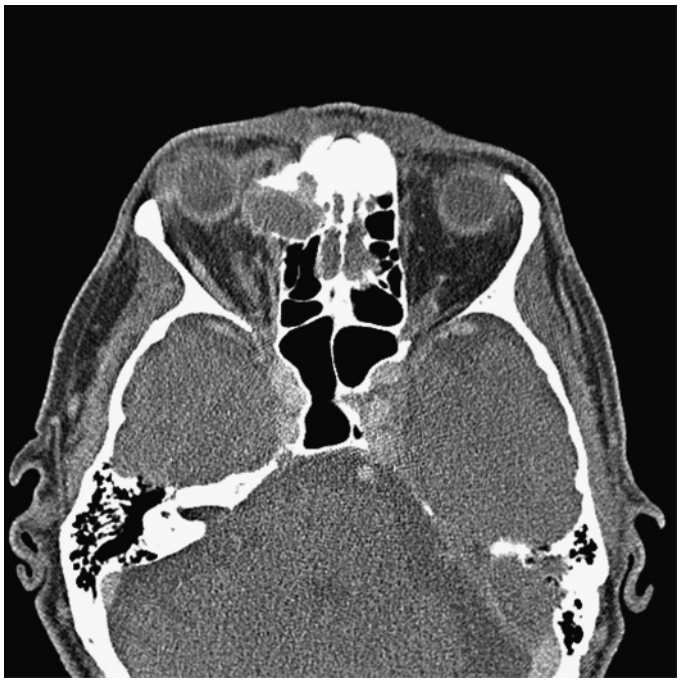

FIGURE 1.
Axial CT showing the mucocele invading the right orbit.

eye (OD) about 40 years prior, which had been repaired with a scleral buckle, and a low-risk choroidal nevus in the left eye (OS).

His medical history included asthma, attention-deficit/hyperactivity disorder, chronic sinusitis (status—post right functional endoscopic sinus surgery, bilateral maxillary antrostomy, uncinectomy, and partial ethmoidectomy in 2009), tinnitus, hearing loss, well-controlled non–insulin-dependent diabetes, genital herpes, lower back pain, and low low-density lipoprotein cholesterol level. His medications included the following: albuterol 90 μg inhaler (2 puffs 4 times per day as needed), budesonide 80/formoter 4.5 μg inhaler (Symbicort, AtraZeneca) (2 puffs bid), metformin 850 mg tid, and 81 mg aspirin daily.

Upon examination, the patient's visual acuity was 20/25+ OD, 20/20 OS with minimal refractive change. His pupils were round and reactive to light in both eyes without afferent pupillary defect. His extraocular muscle motilities were full and smooth OS, but OD was restricted in left gaze (medially) and superior left gaze (superonasally). The patient subjectively had diplopia intermittently in right and primary gazes and constantly in left and superior left gaze. The patient's phoric posture was measured at 4 prism diopters right hypotropia in primary gaze but was worse on right head tilt. He had visible proptosis, and Hertel exophthalmometry was measured at 19 mm OD and 16 mm OS. Additionally, mild resistance to retropulsion was appreciated in the right eye.

Slit lamp and dilated fundus examination was remarkable for ptosis OD with a marginal reflex distance of −0.5 mm OD, 2.5 mm OS, mild nuclear sclerotic cataract in both eyes (OU), asteroid hyalosis OD, and small tilted discs with 0.2 OD and 0.3 OS cup-to-disc ratios. He had chorioretinal scarring and scleral buckling inferiorly OD and a small, stable flat choroidal nevus in the temporal macula OS.

Based on the clinical examination, primary differentials for the diplopia at that point were a space-occupying orbital lesion or less likely extraocular muscle infiltration or entrapment. A maxillofacial computed tomography (CT) scan without contrast was ordered initially, and a 4 prism diopter base-up Fresnel prism was placed over OD to alleviate the diplopia.

The CT scan indicated a significant expansile lesion of the right frontal sinus with heterogenous attenuation of its intraluminal contents. There was marked osseous thinning with erosion noted along the medial wall of the right orbit, and significant protrusion of the lesion into the superior aspect of the orbit, which abutted the right superior rectus and produced mass effect on the right superior oblique (Fig. 1). There was notable demineralization of the inner table of the right frontal sinus and protrusion of the lesion about 1 cm toward the frontal lobe with mild mass effect on the frontal lobe (Fig. 2). There was, additionally, focal expansion and demineralization along the outer table of the right frontal sinus. There were additional radiologic findings consistent with his previous bilateral maxillary antrostomies, uncinectomies, and partial ethmoidectomies. The radiologist felt that the findings in the right frontal sinus were likely long-standing and favored mucocele formation versus chronic sinusitis or less likely sinus neoplasm.

The patient was immediately referred and seen the following day by the hospital's otolaryngology clinic. He had been seen in that clinic 5.5 years prior. At that visit, the patient reported that he performed sinus rinses twice per day but "always" has green mucous discharge from the right nostril with occasional exacerbations. Rigid nasal endoscopy was performed, and the right side was noted to have a widely patent maxillary antrostomy; the middle meatus and frontal recess were completely occluded with watery polypoid disease but no pus or purulence to culture. Given the examination and CT results, the frontal sinus lesion was thought to be an expansile mucocele versus mucopyocele. He was prescribed an oral prednisone burst followed by taper. A magnetic resonance image with contrast and a stealth CT for intraoperative guidance for another anticipated sinus surgery were obtained (Figs. 1 to 4). The patient was then referred to our affiliate hospital's

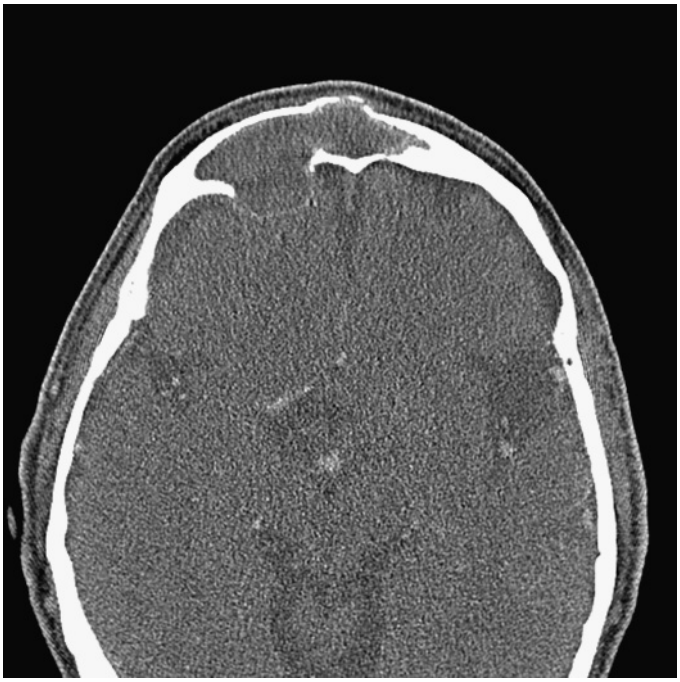

FIGURE 2.
Axial CT showing the right frontal sinus mucocele's erosion through the inner table and mass effect on the right frontal lobe.

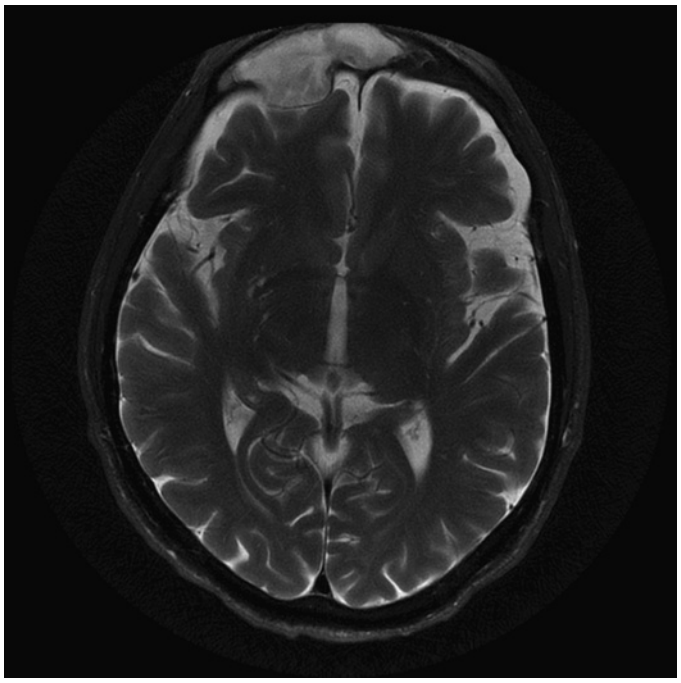

FIGURE 3.

Axial magnetic resonance imaging showing the mucocele's protrusion posteriorly with mass effect on the right frontal lobe.

Otolaryngology Skull Base Clinic, where it was recommended that he undergo a stealth-guided modified endoscopic Lothrop procedure to drain the mucocele, and this was scheduled for about 3 weeks thereafter.

The magnetic resonance image with contrast demonstrated the right frontal sinus mass and illustrated extension into the right ethmoid sinus, right superior orbit, and onto the right frontal lobe. There was no significant associated brain edema noted.

The patient underwent stealth-guided endoscopic right ethmoidectomy, radial frontal sinusotomy, marsupialization of the mucocele, and transcutaneous irrigation. Contents of the mucocele were cultured and were negative for fungal and acid fast elements, but it grew *Prevotella intermedia* and pan-sensitive *Staphylococcus aureus*. His follow-up care was remarkable only for persistent edema at the frontal recess, but after a steroid taper, his postoperative care was uneventful.

The patient again presented for an eye examination about 2 months after his sinus surgery. A retrospective interview was conducted with the patient regarding symptoms before surgery related to the mass effect on his frontal lobe. An extensive questionnaire was all reported as negative by the patient, except for mild changes in spatial orientation. Interestingly, with the same questionnaire as was asked of the patient, in independent interviews with two of his daughters, changes in the patient before the mucocele discovery had been evident to them. They reported a considerable change in his memory, some falls and running into things, worsening difficulty with personal financial planning and money management, and also that he had casually disclosed some potentially inappropriate, or at least awkward, personal information to them. Without elicitation, they also reported that his right ptosis had been worsening before the frontal sinus surgery.

At that examination, he reported that the diplopia resolved within about 1 week of his sinus surgery, and he removed the Fresnel prism at that time with no noted diplopia afterward. He felt his right globe was moving back upward into its natural position and his ptosis seemed less prominent to him. He had only a slight restriction of motility in superonasal gaze OD, and he had subjective diplopia only in extreme left and superior left gazes. His phoric posture was slightly exophoric in primary gaze with no vertical deviation. The right eyelid ptosis had improved by 1 mm, as the marginal reflex distance OD was 0.5 mm compared with the previously recorded −0.5 mm. He also had a 1-mm reduction in proptosis OD as measured by Hertel exophthalmometry. All other findings were stable and an annual examination was recommended, or sooner as needed.

DISCUSSION

Mucoceles are mucus-containing, epithelial lined cystic cavities that develop in paranasal sinuses as a result of chronic obstruction of the outlet ostia that connects the sinus to the nasal cavity.[1] Ostia obstruction may occur as a result of anomalous anatomy, inflammation, trauma, polyps, masses such as bony tumors, allergy, or past surgical procedures.[1,3–8] The ostia obstruction leads to entrapment of the mucus-secreting epithelial cells in the sinus; the cells continue to secrete their mucoid substance, resulting in the expansive nature of mucoceles.[2] In addition to ostia obstruction, inflammatory cytokine and prostanoid production may also play a role in mucocele formation.[9] If the mucocele's retained mucoid secretion becomes infected pus, it is referred to as a pyocele or mucopyocele.[3–5]

Mucoceles may form in any of the paranasal sinuses, but they most frequently involve the frontal sinus (60 to 89%) or ethmoid sinus (8 to 30%), infrequently involve the maxillary sinus (<5%), and seldom involve the sphenoid sinuses.[1,3,5,7,10,11] The frontal

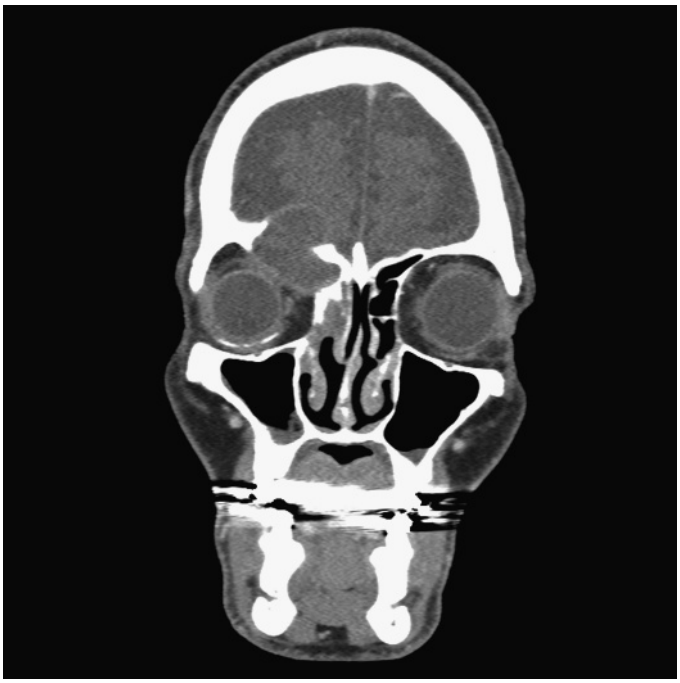

FIGURE 4.

Coronal CT with contrast showing the right frontal sinus mucocele's extension; note the downward displacement of the right globe.

and ethmoid sinuses are particularly pertinent to eye care, as the superior orbital wall includes the floor of the frontal sinus, and the medial orbital wall includes the lateral aspect of the ethmoid sinus.[3] Mucoceles have no sex or age predilection but are most frequently diagnosed in the fourth through seventh decades of life.[2,3,12]

The mucocele itself is benign, although the slowly expanding mucocele may lead to resultant erosion and reabsorption of the bony sinus walls; such erosion can lead to mucocele protrusion into the orbits, nasopharynx, or cranium, although intracranial extension is less common.[1,2,5,13] The posterior wall of the frontal sinus is notably thin and, thus, susceptible to erosion, which can lead to exposure of the cranial contents to the mucocele.[3] Ruptured intracranial mucoceles can lead to serious complications such as meningitis, brain abscesses, seizures, meningoencephalitis, or cerebrospinal fluid fistula.[1,5,12,13]

Patients with mucoceles may present with a spectrum of symptoms, which will vary based on the mucocele's location and extent. Symptoms may be rhinologic or neurologic, including nasal discharge, nasal obstruction or postnasal drip, headaches, dizziness, seizures, maxillofacial pressure, subcutaneous forehead or periorbital tenderness or swelling, facial asymmetry, or even cranial nerve damage.[3–5,7,11,14–16] Most frequently, the symptoms may be ophthalmologic.[11] If the mucocele has invaded the orbit, patients may have diplopia, globe displacement, extraocular motility restrictions, proptosis, mechanical ptosis, or a retrobulbar pain or pressure sensation, as was the case with this patient. The direction of the globe displacement and proptosis is an important indication of the site of the offending lesion. The mucocele essentially acts as a space-occupying lesion that increases the orbital content volume and can even cause increased intraocular pressure, decreased visual acuity, visual field defects, or compressive optic neuropathologic vision loss.[2,3,5,11,14,17] Enophthalmos, though rare with mucoceles, is possible, as sinus destruction could potentially increase the orbital space and allow inward globe displacement.[14]

With posterior frontal sinus erosion into the cranium, there is potential for problems associated with the mucocele's mass effect on the frontal lobe, such as a frontal lobe syndrome. Frontal lobe syndrome implies a collection of signs and symptoms attributed to damage to the frontal lobe's prefrontal area; there are multiple frontal lobe syndromes, whose features largely depend on the portion of the frontal lobe affected, although there is often overlap of syndromes.[18] Associations include memory dysfunction and personality, emotional, and social behavioral changes such as apathy, boastfulness, impulsiveness, recklessness, poor forethought or planning, lack of insight, inattention, indifference, poor judgment, or disinhibition.[8,18,19]

Frontal lobe syndrome attributed to mass effect from a frontal sinus mucocele with intracranial extension is exceedingly rare.[12] One case was reported by Ada et al.[20] of a giant, bilateral frontal sinus mucopyocele with major intracranial extension mentioning memory impairment and personality changes in the patient. Another from Visocchi et al.[12] reported a giant frontal mucocele complicated by subdural empyema causing personality changes in a patient. Only one other relevant case was found, reported by Sarsilmaz et al.,[8] where a giant frontal mucocele with intracranial extension and frontal lobe mass effect caused memory impairment, perception difficulties, and personality and behavioral alterations in the patient.

In this case, the frontal lobe mass effect was causing mental status (memory), social behavior (disinhibition), and planning and goal-directed behavior (financial planning and management) changes associated with frontal lobe syndrome. The patient's spatial orientation difficulties could be attributable to his globe displacement rather than, or in addition to, the mass effect. This case nicely displays the potential helpfulness of a third party in history taking but more importantly illustrates how the insidious nature of mucocele growth causing frontal lobe mass effect can cause subtle and guileful changes in mental status and behavior, often unrecognizable by the patient themselves. Therefore, frontal lobe syndrome is likely more frequently present than is clinically reported.

It is recommended, as used in this case, that otolaryngology is consulted for a physical examination and confirmatory radiologic imaging when mucocele is suspected. Computed tomography imaging is useful for gauging the size and extent of mucocele involvement, including the presence and amount of sinus wall bone expansion and erosion. Bone erosion along the sinus walls tends to be smooth and uniform given the mucocele's slow expansion. The criteria used for diagnosing mucoceles based on CT are as follows: a mass with isodense homogenous contents, well-defined margins, and surrounding patchy osteolysis.[3,8] The contrast enhancement of the lesion on T1-weighted magnetic resonance imaging can vary based on the proportion of mucus, water, and protein content of the mucocele, although they tend to be fairly bright; they appear iso-hyperintense on T2-weighted imaging.[3,5,8,11] This is helpful for differentiating mucoceles from neoplastic activity, where both T1- and T2-weighted images tend to be iso-intense relative to the brain.[3,8,21]

Surgery is the required treatment for complete mucocele eradication and reestablishment of normal sinus drainage. Complete removal of the mucocele is paramount. This procedure may be achieved with transnasal endoscopic surgery, which is now the preferred approach over the more invasive external craniotomy with craniofacial surgery, but a combination craniofacial and endoscopic procedure may be required.[1–3,22] A transcaruncular approach may also be performed for more direct access to frontoethmoid mucoceles or a transorbital neuroendoscopic procedure for more complex mucoceles.[1,2,23] Reconstruction of bony defects from thinning or erosion may be required. Such procedures are usually performed by otolaryngologists, although collaboration with ophthalmologists and neurosurgeons may be prudent, depending on the extent of the mucocele's involvement.

There are several potential orbital complications of sinus surgery owing to the close proximity of the paranasal sinus to the orbits. This is particularly true for surgeries involving the ethmoid sinuses because of their position relative to the orbit and the extreme thinness of the lamina papyracea (orbital lamina of the ethmoid bone), which serves as the bony boundary that separates the ethmoid sinus from the medial orbit. Inadvertent direct trauma to orbital contents during surgery is possible; the medial rectus is near the lamina papyracea and is susceptible to direct damage or entrapment. Intraorbital hemorrhage and direct optic nerve damage are also possible, among a plethora of other complications that can result if direct or inadvertent orbital entry occurs during sinus surgery.[24] Postoperative complications may also occur, including intraorbital hematoma, abscesses, or mucocele recurrence.[22]

Mucoceles may initially form several years or even decades after previous sinus surgery or trauma and then have a high rate of recurrence, particularly in patients whose mucocele location is frontoethmoidal and those who have had multiple sinus surgeries or chronic sinusitis.[15,22] The reported recurrence rate is generally around 10%, although a large single-center series published by Devars du Mayne et al. reported 23.5% recurrence rate with an average onset of recurrence being at 4 years, but as long as 41 years; therefore, that study advised CT imaging every 2 years for at least 4 to 5 years after surgery, but more frequently in patients whose risk of recurrence is elevated because of persistent inflammatory sinus diseases.[3,11,22]

CONCLUSIONS

This case not only exhibits a very rare presentation of frontal sinus mucocele with intracranial extension and frontal lobe mass effect causing a frontal lobe syndrome but also demonstrates many of the ocular and visual complications commonly associated with paranasal sinus mucoceles. It emphasizes the importance of keeping sinus lesions in one's list of differentials when patients present with suspicious symptoms; it also underscores the clinical relevance of this differential, as proptosis (83%) and diplopia (45%) are among the most common complaints associated with mucoceles.[3] Early identification and surgical intervention with complete mucocele evacuation is vital for preventing and reducing morbidity associated with invasive mucoceles.

Given potential postoperative complications after mucocele eradication and the frequency of mucocele recurrence, patients should be monitored regularly and long term, both clinically and radiologically.

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