

TITLE: Mucocels of the Paranasal Sinuses

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Mucocels of the paranasal sinuses are cystic lesions lined by respiratory epithelium and are believed to be caused by obstruction of the sinus ostium or obstruction of a mucus secreting gland, leading to the accumulation of secretions into an expansile mass. Though they are benign, their expansion can cause destruction of surrounding structures. For this reason it is crucial to diagnose and treat these in a timely fashion. The obstruction that begins the process can be caused by congenital anomalies, inflammation, trauma, surgery, or a tumor. The contents are most commonly sterile. An infected mucocel is called a mucopyocel.

Mucocels are rare sinus pathologies; however, they are the most common expansile lesion of the paranasal sinuses. They are slow growing and can take up to 10-15 years to produce symptoms that prompt the patient to seek medical attention. They are most commonly found in the frontal and ethmoid sinuses. There is an increased incidence of maxillary sinus mucocels in Japan compared to the United States and developed European countries. This is thought to be due to the increased number of radical surgeries for sinusitis, like Caldwell Luc procedures, done in Japan in the past. A study in 1979 found 132 cases of maxillary sinus mucocels over a period of 9 years at Tokyo Medical University Hospital. Ninety-nine percent of them had a history of maxillary sinus surgery. The mucosal disruption caused by the surgery sets the stage for mucocel formation. Some suggest that compartmentalization of the antrum after the operation may leave islands of mucosa without drainage and lead to eventually mucocel formation.

In 1978 Natvig and Larsen looked at 112 patients with mucocels and found that the vast majority of them (77%) had frontal sinus mucocels. Fourteen percent were in the frontal/anterior ethmoid sinuses, 5% in anterior ethmoids, 1% in posterior ethmoids, and 3% in maxillary sinuses. A similar distribution is observed in the United States. The study looked at patients that presented to the National Hospital in Norway over a 27 year period (1947 to 1974). Twenty percent of these patients had a history of previous sinus surgery or trauma. Forty-eight percent had chronic sinus disease. One hundred eight of these patients were treated with the

Lynch Howarth operation. Eleven cases of mucocele relapse were discovered on re-evaluation in 1976.

The anatomy of the sinuses is important to understand in order to appreciate how mucoceles form, the symptoms they cause, and how to treat them. The maxillary sinus is drained through its primary ostium which is located in the roof of the sinus. Mucus from the maxillary sinus drains through the primary ostium then the infundibulum until finally it reaches the middle meatus. The frontal recess is the passage for mucus drainage from the frontal sinus. This hour glass shaped passage is the narrowest channel through which the frontal sinus drains. Obstruction here prevents clearance of mucus from the frontal sinus. The Aggar Nasi cell is an ethmoturbinal remnant that represents the most anterior ethmoid cell. It usually borders the frontal sinus primary ostium or floor. Therefore, its size can impact the opening of the frontal recess. It is also worth noting the close proximity of the orbit. Because of the close proximity of the eye it is not uncommon for patients to present initially with only ophthalmic symptoms, like ptosis, proptosis, or impaired eye mobility.

The frontal sinus is funnel-shaped and can vary in size and shape from person to person. There is generally a central septum separating the left from right. The floor slopes inferiorly to the midline where the primary ostium is located on the floor. The frontal recess is an hourglass-like narrowing between frontal sinus and anterior middle meatus. Obstruction here results in a loss of ventilation and mucus clearance from the frontal sinus.

The sphenoid sinus is the most posterior sinus. It has a close relationship to the cavernous sinus. The cavernous sinus houses the internal carotid, nerves carrying sensory information from the face (ophthalmic and maxillary nerves), and nerves carrying motor innervation to the muscles of the eye (oculomotor, trochlear, and abducent nerves). The close proximity puts these structures at risk when a mucocele develops in the sphenoid sinus.

The paranasal sinuses have a mucosal lining made up of ciliated epithelium. A mucus blanket is found on the surface. The cilia beat in a coordinated manner to propel the mucus in a specific pattern of flow. This process is known as mucociliary clearance. The pattern of flow is different for each sinus and persists regardless of creation of new openings in the sinus. In the maxillary sinus, the flow of mucus starts in the floor of the antrum. Flow of mucus is then directed up against gravity towards the primary ostium. In the frontal sinus, the mucus flows up along the medial wall then laterally across the roof. Finally, it moves medially along the floor where some of it exits through the primary ostium located medially in the floor. This mucus will travel down the frontal recess and into the middle meatus. Mucus that does not exit is re-circulated. Mucus flow from the sphenoid and posterior ethmoid sinuses is propelled into the sphenoethmoidal recess and then into the superior meatus. The primary ostium of the sphenoid is located medially in the anterosuperior portion of the anterior wall of the sinus.

Macroscopically, mucoceles appear as thick walled gray colored cysts. Under the microscope one can appreciate pseudostratified columnar epithelium with decreased number of ciliated cells. Reactive changes like fibrosis, granulation tissue, hemorrhage, and even squamous metaplasia can be seen. Sterile mucus and cholesterol crystals along with hypertrophic goblet cells can be seen as well.

Mucoceles are created when there is obstruction of mucus outflow. This can occur secondary to blockage or disruption of a sinus ostium by a variety of causes. Inflammation of the mucosa can obstruct. Trauma or surgery can destroy the outflow tracts of the sinuses and additionally lead to mucosal disruption and scar formation. Masses like polyps and tumors can prevent sinus clearance of mucus. Reports have even shown that radiotherapy can cause significant scarring that can lead to mucocele formation. Accumulation of mucus occurs as the outflow tract is blocked and mucus secretion continues. As the mucocele grows and expands it pushes on the walls of the sinus. The increased pressure causes bone devascularization and chemical mediators lead to osteolysis. Studies have found that cytokines are one of the principal causes of osteolysis of the bone surrounding mucoceles. Inflammatory cytokines such as IL-1, IL-6, TNF alpha, and PGE2 have been found in mucoceles and are thought to be involved in signaling osteoblasts to increase bone resorption. As we enter a new age in medicine, immunotherapy and modulation of these cytokines may be an alternative to invasive procedures for management of mucoceles.

Mucoceles can present in a variety of way depending on their location. Headache is common among all locations but may be absent in some patients. Patients commonly experience facial pressure and swelling. Dental pain can occur in cases in which the maxillary sinus is involved. Ophthalmic manifestations are very common particularly in frontal sinus mucoceles. In fact, patients commonly present to the ophthalmologist first for evaluation of proptosis, periorbital pain, impaired ocular mobility, or blurry vision. Additionally, patients can experience neurologic manifestations such as confusion, meningitis, or CSF leak. The presenting features can vary widely depending on the size and location of the mucocele.

The ophthalmic manifestations can be explained in a few different ways. First, more anterior located mucoceles, like those in the maxillary, frontal and anterior ethmoids, are more likely to produce proptosis, periorbital pain and decreased ocular mobility. In these cases it is the pressure on the globe by the expanding mucocele that pushes it outwards. Additionally, expansion on to the extraocular muscles restricts their movement and function leading to an inability or difficulty moving the eyes. The more posteriorly located mucoceles more commonly present with blurred vision and decreased ocular mobility. This can be explained by expansion of the sinus wall on the optic nerve leading to compromise of its blood supply and optic atrophy. Optic neuritis can also develop from the direct spread of the mucocele's inflammatory contents. Involvement of the abducent or oculomotor nerve may also explain findings of impaired ocular mobility. Periorbital pain is attributed to the inflammation and mucosal stretching caused by the expanding mucocele. The nerve impulses are carried by the trigeminal nerve. Vision loss can be a permanent complication of mucoceles. When associated with sudden onset of vision loss, there is suspected spread of infection or inflammation to the optic nerve. This situation has a poor prognosis for recovery and many of these patients will have permanent vision loss even after the mucocele is removed. On the other hand, when the vision loss is gradual it is attributed to ischemia and there is a better prognosis for recovery of vision after surgery.

Computed tomography (CT) without contrast is the imaging modality of choice for the diagnosis of mucoceles. There are several radiologic features that characterize sinus mucoceles; however, none are unique to mucoceles. Complete opacification of the sinus is very typical. Muroid, low density material is appreciated. Bones of the sinus are generally remodeled and expansion is seen. Sinus walls bow radially outwards. Areas of thickened and thinned bone can

be seen as well as erosions. Herniation into adjacent structures is common. Mucoceles expand towards the path of least resistance which is commonly into the orbit or cranial cavity. Compartmentalization can be appreciated in some mucoceles with a thin bony septum separating the mucocele from the rest of the sinus cavity.

Magnetic resonance imaging (MRI) is not the best imaging modality for diagnosing mucoceles. The variation in protein and water concentration of mucoceles causes varying signal intensity. However, it does allow for differentiation of mucoceles from solid components of tumors. This is particularly true with contrast enhancement. In general, mucoceles appear with a thin peripheral enhancement and tumors have diffuse enhancement.

Surgical treatment is a must as mucoceles will continue to grow and expand and cause irreversible damage. Surgeries can be categorized as external or endoscopic. Some techniques utilize a combination. The external approach traditionally involves complete extirpation of the mucocele and obliteration of the sinus. The endoscopic approach is more conservative and involves marsupialization of the mucocele and preservation of adequate sinus drainage. Regardless of the technique used, long term follow up is imperative as recurrences often occur many years later.

External approaches allow for better visualization and access of the sinus. As the orbit, meninges, or brain become involved, the more an external approach is indicated. This is a more radical approach and is traditionally used for fronto-ethmoidal mucoceles. The classic Riedel procedure entails removal of the anterior and inferior walls of the frontal sinus. Soft tissue from the forehead is collapsed down onto the posterior table of the sinus. This technique leaves the patient with a significant cosmetic deformity. The Lynch Howarth procedure approaches the frontal sinus through the orbit. A curved incision is made from the inferomedial eyebrow along the upper third of the nose. The medial wall of the orbit is perforated and the floor of the frontal sinus is removed. It can also cause cosmetic deformity. In the past, obliteration of the sinus with fat was popular. However, fat in the sinus makes monitoring for recurrent disease very difficult.

Conservative endoscopic approaches with marsupialization accomplish establishment of sinus drainage and not removal of mucosa. This approach has increased safety and efficacy with recurrence rates near 0%. This is the treatment of choice for mucoceles.

Recurrence can still occur regardless of the technique used for removal. Therefore, adequate surveillance is essential. Risk factors for recurrence include surgery during acute infection, presence of multiple mucoceles, and significant extension outside the sinus wall. Periodic nasal endoscopy in the office is recommended to assess patency of the ostium. Recurrences are few if adequate drainage is established. It can take many years for mucoceles to recur.

Case Report

A 50-year-old woman presented to the otolaryngology service with a nine year history of left facial pain and pressure after a Le Forte I osteotomy done for cosmetic reasons several years previously. A CT scan showed an expansile unilocular homogeneous lesion with thin sclerotic margins associated with the left posterior most tooth apex. It also showed mucosal thickening in the mid left maxillary mucosa. The CT was read as a dental cyst and the patient was referred to oral maxillofacial surgery. A curettage and lavage of the left maxillary sinus and I&D of the left maxillary sinus abscess was performed. After surgery, the pain and pressure were resolved but returned two weeks later. The CT was reviewed in conjunction with an assessment of the surgery notes and it was determined that the lesion, previously thought of as a dental cyst, was actually a maxillary mucocele and was abutting the floor of the maxillary sinus around her teeth. The Otolaryngology Department performed endoscopic sinus surgery and antral puncture with marsupialization of the maxillary mucocele. One month after surgery, the patient had no more complaints of facial pain or pressure.

In conclusion, mucoceles are late complications of sinus ostium obstruction or mucous gland obstruction. These expansile lesions are capable of bony destruction and compromise of surrounding structures. Endoscopic sinus surgery is the first choice for treatment, but sometimes external approaches may be necessary

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