Fronto-Orbital Mucoceles: Our Experience

Claudio Rinna, MD, Andrea Cassoni, MD,
Claudio Ungari, MD, Massimiliano Tedaldi, MD
Tito Matteo Marianetti, MD

Abstract: Frontal sinus mucoceles are rare benign neoplasms that can result in bony erosion extending from the borders of the sinus into the orbital cavity.

The authors report the fronto-orbital mucoceles they have observed in the last 8 years.

The authors used an “open surgery” approach in 12 fronto-orbital mucoceles, characterized by an osteoplastic frontal flap, through a coronal incision or Lynch incision. In this way, they were able to expose the frontal sinus and to remove completely the mucocele from the periorbita. Only one recurrence is reported 2 years after surgery. Excellent aesthetic results have been reached in all cases.

In the literature, many different positions are reported for the treatment of mucoceles. The endoscopic surgery of the paranasal sinuses has become the procedure of choice for mucoceles of maxillary, ethmoidal, and sphenoidal sinuses. However, few authors have recommended the endoscopic approach for fronto mucoceles. The indications and limitations of endoscopic and “open” surgery are critically discussed in the light of the authors’ personal experience and current literature.

Key Words: Fronto-orbital, mucoceles, open surgery

Mucoceles are benign cystic-like lesions with inflammatory features that develop into one or more paranasal sinuses. Mucoceles present a progressive feature, occupying all the sinus and eroding its walls. This possibility seems to be linked to the pressure in the sinus and to the production of inflammatory substances from the hyperplastic mucosa. Lund demonstrated that in the membrane that lines the mucocele, the prostaglandin E2 (PGE2) levels are higher than normal. This prostaglandin, along with other inflammatory agents, would be able to activate the osteoclasts with the consequent bony resorption.

Frontal mucoceles develop in response to an obstructed drain through the fronto-nasal duct, the cause of which could be a particular density of mucous (for instance in patients with cystic fibrosis) or a mechanical obstruction of the channel. Mucoceles can be correlated to past traumas, nasal polyposis, past sinusitis, concurrent osteoma, a postoperative reaction of cicatrization, or a particular anatomical structure of the fronto-nasal channel. It must be emphasized that a malignant tumor can cause the obstruction of the duct with secondary mucocele. The differential diagnosis must be done with osteoma, which is not unusual in the frontal sinus and sometimes associated with the mucoceles, fibrocystic dysplasia, inborn defects such as brain prolapse, and primitive and secondary frontal sinus tumors.

The frontal mucoceles, which represent about 60% of the paranasal sinus mucoceles, can remain symptomless for a long time. Eroding the bony walls, they can extend downward to the socket, the ethmoidal sinus, and the nasal cavities; forward to the ventral wall of the frontal sinus and the upper lid; and upward inside the anterior cranial fossa. Although the incidence of the frontal mucoceles has not been carefully established, they are considered responsible for 2.7% of the nonendocrine proptosis.

The orbital cavity invasion never has an infiltrating feature, but it is always characterized by a compression of the endo-orbital structures, with exophthalmus caused by the pressure increase inside the socket. Proptosis, together with the presence of a painless frontal mass of stretched-elastic consistency,
could lead to a clinical suspect of mucocele. Other signs and symptoms are frontal headache, diplopia (by compression of the superior rectus muscle or the superior oblique muscle), ptosis and palpebral edema, nasal discharge, and nasal obstruction sensation. A visual acuity decrease is not usual in the frontal mucoceles, whereas it is common in the sphenoidal and posterior ethmoidal mucoceles as a consequence of lesions of the optic nerve for direct contact or for a protracted compression. In the literature there is no agreement concerning the best therapeutic approach to frontal mucoceles. If FESS (functional endoscopic sinus surgery) has become the main treatment of the ethmoidal, maxillary, and sphenoidal mucoceles, the therapeutic approach to the frontal mucocele is not unequivocal.

Surgical techniques comprise a preservative approach with an enlargement of the fronto-nasal duct and a marsupialization of the sinus or a radical approach consisting of obliteration with autologous tissue or through a frontal sinus cranialization.

In the current work, we examined the fronto-orbital mucoceles observed in the last 8 years to evaluate, in conformity with our results and a review of the literature, the most adequate surgical approach to the frontal mucoceles that present orbital invasion.

**Patients and Methods**

Between January 1995 and January 2003, 12 patients with frontal mucoceles with orbital extension were surgically treated in the department of Maxillo-Facial Surgery of “La Sapienza” University, Rome. Five were male patients and seven female; they ranged in age between 11 and 80 years (average, 54.4 years). In three patients the frontal mucocele also involved the ethmoidal sinus. Predisposing factors to the mucocele development were found in five patients: three of whom had previous surgery in the nasal-sinus region, one with concomitant osteoma, one with craniofacial trauma, and one with sinonasal polyposis. Two cases were relapsing mucoceles 6 and 9 months, respectively, after operation was performed in another structure. The most common symptoms were frontal headache (25.0%), pain in orbital region (16.6%), exophthalmus (83.3%), mass in preorbital region (41.6%), palpebral edema (16.6%), and diplopia (33.3%). No patient reported a reduction of visual acuity.

Preoperative diagnoses were determined with computed tomography (CT) scans of the paranasal sinuses (Fig 1). In three cases, a magnetic resonance imaging (MRI) scan also was executed. The invasion of the orbital cavity was radiologically demonstrated in all 12 patients: CT and MRI provided evidence of the size, shape, and extension of the mucocele and displacement of the ocular globe. In two patients the mucocele also presented an intracranial extension.

All patients underwent surgery. In nine, a coronal incision was performed with preparation of the frontal osteoplastic flap and complete removal of the fronto-orbital mucocele (Fig 2). This approach allowed separation of the mucocele from the dura in the two patients with an extradural intracranial invasion, in whom the posterior wall of the frontal sinus was eroded. A transfrontal approach with Lynch incision was chosen for three patients. It was considered the least invasive method for an 11-year-old patient and thought to avoid unpleasant aesthetic outcomes for two patients with a tendency to baldness. In only one patient, one with an ethmoido-frontal relapse caused by a fronto-orbital mucocele removal with the transfrontal approach, did we decide to proceed with an intranasal approach, restoring the patency of the frontal duct with the use of endoscopy.

**Results**

The 12 patients were regularly followed up during different periods, from 6 months to 8 years. Postoperative CT scan demonstrated the complete ablation of the lesion in all patients. The average period of hospitalization was 4 days. No patient experienced postoperative complications. Clinically, we noticed an immediate postoperative improvement to nor-
malization of the proptosis in 10 patients affected by this clinical sign. Patients with diplopia and headache reported the disappearance of such symptoms during the postoperative period.

The surgical approach to 12 fronto-orbital mucoceles was constantly based on the “open surgery”: the realization of an osteoplastic frontal flap, through a coronal incision or Lynch incision, allowed us to expose the frontal sinus, diagnose the mucocele intracranial invasion, completely remove the mucocele from ethmoidal cells when affected, and separate its mucosa from the periorbita.

The pathologic analysis of the operating specimen showed an ulcerative hyperplastic mucosa with low acute inflammatory or chronic nonspecific infiltrate.

Only one patient experienced recurrence, approximately 2 years after the surgical intervention. The patient underwent surgery again with an endoscopic nasal approach, and he has experienced no additional recurrence.

Excellent aesthetic results have been reached in all patients.

DISCUSSION

The frontal mucoceles were described by Dezheimeris for the first time in 1725. In 1818, Litersangeback described signs and symptoms of mucoceles, which he called “hydatidic cyst.” The term “mucocele” was first used by Rollet in 1896 to describe a lesion of the upper-internal part of the orbit.

Although several theories have been proposed, the physiopathology of mucoceles is mainly unknown. The two most important factors involved in the mucocele’s pathogenesis are mucosal inflammation and the obstruction of the fronto-nasal duct.

The feature of eroding the bony walls is connected both to the presence, inside the mucocele mucosa, of a large amount of inflammatory cytokines with osteolytic capability (PGE2), and to the endosinus pressure increase, as demonstrated by Fenton et al.

If the mucocele does not cross the borders of the frontal sinus, the most common symptoms are headache and a tension sensation in the frontal region. If the mucocele becomes infected, it is called mucopiocele. This formation has an extreme aggressiveness toward surrounding tissues, and it can lead to lethal complications, such as cerebral abscess or septic endophthalmitis.

The orbit is often affected by frontal sinus mucoceles, and the onset of the symptoms often is linked to that affliction. The most common sign of the orbital invasion is the exophthalmus (found in 83.3% of our patients). The exophthalmus is typically antero-inferior, often with combined lateral deviation of the ocular globe.

In the case of a medial deviation, a pathology of the lachrymal gland must be take into consideration, whereas the relief of an axial exophthalmus, especially if it’s bilateral, leads us toward the diagnosis of Graves-Basedow disease. Other signs and ocular symptoms are diplopia, conjunctival hyperemia, and blepharoptosis. Visual acuity decrease is not a characteristic of frontal-orbital mucoceles. In fact, the superior length of the optic nerve compared with the orbital axis allows a proptosis of a level relatively high, with a small effect on the visual acuity.

The noncomplicated mucocele has slow growth, and the orbital invasion development takes several months, sometimes years. During that period, a contemporaneous erosion of the posterior or the upper wall of the frontal sinus with an intracranial invasion is common. In literature, an intracranial infiltration percentage from the frontal mucoceles can vary from 27% to 61%. In our series, the intracranial invasion was present in 2 of 12 (17%) patients with fronto-orbital mucocele.

Careful radiologic preoperative research is fundamental for diagnosis of mucocele and for the differential diagnosis from other pathologies, including malignant tumors of the frontal sinus.

The CT scan is the best diagnostic instrument for
studying the mucocele, it shows the mucocele as a capsulated mass, with homogeneous density, isodense with the cerebral tissue, with peripheral enhancement after injection of contrast medium. The bony walls can be normal, swollen, or eroded. MRI scan is indicated in the differential diagnosis with a malignant neoplasia or with a brain prolapse. Nevertheless, the images of MRI mucoceles are variable: they can present a hypersignal both in T1, because of the strong proteic concentration in the mucus, and in T2, because of the large quantity of water. However, mucoceles can also present a hyposignal in T1 and absence of a signal in T2 because of the dry mucus.

Concerning the treatment of the mucocele of the frontal sinus, many different positions are reported in literature. Some authors support an endoscopic approach to the frontal sinus, whereas others think that the best therapeutic procedure for the frontal mucoceles is the “open surgery.” The endoscopic surgery of the paranasal sinuses has been gaining more approvals in recent years and it’s currently the must chosen procedure for the treatment of benign diseases such as sinusitis and mucoceles of maxillary, ethmoidal, and sphenoidal sinuses. However, with regard to the frontal sinus, the same supporters of endoscopic surgery often do not recommend the endoscopic approach to frontal mucoceles. For instance, according to Naudo et al., our school supports, whereas others support an endoscopic/open surgery approach in five of six cases. In addition, other supporters of the endoscopic method admit that it is not usable for all frontal mucoceles. For example, it has to be excluded in cases of combined osteoma or when frontal sinus floor drilling for widening the sinus ostium is needed because this drilling is at risk when approached via the endoscopic method. Other authors support the endoscopic approach to the frontal sinus, but they essentially refer to the treatment of the frontal sinus or mucoceles without orbital invasion. At the end, the endoscopic surgery appears suitable in the ethmoidofrontal mucoceles limited to the medial part of the frontal sinus: in that case, it is indeed possible to obtain complete mucocele removal and an orifice enlargement of the frontal duct via the endoscopic method.

According to Perié et al., our school supports, as in the past, the “open surgery” in the treatment of fronto-orbital mucoceles. The open surgery provides the choice of several types of incision: the Lynch incision, coronal incision, and paralateronasal rhinotomic incision are the most used. The paralateronasal rhinotomic incision was not used because, even if it allows a very good exposure of maxillary and ethmoidal sinuses, it’s less appropriate for frontal sinus. The incision that we prefer has been the coronal one because it allows us to perform a wide osteoplastic flap with the possibility of exposing a large part of the frontal sinus, together with excellent aesthetic results. However, for patients with a tendency to baldness, a Lynch incision was chosen to avoid unsightly scars.

With the open surgery, we can follow a conservative procedure, with the purpose of assuring a good drain to the frontal sinus after the mucocele has undergone complete resection, or a radical procedure, with the ablation of the frontal sinus and its filling up with autologous tissue (like abdominal fat, muscle, or galea-pericranium flap) or through cranialization.

The ablation/filling up surgery of the sinuses often is associated with numerous complications, such as infections in the region from which abdominal fat is taken or lesions of cephalic dura during the mucocele exeresis in the case of osteolysis of the frontal sinus posterior wall.

Another disadvantage of the sinus obliteration with fat tissue is that a careful radiologic postoperative follow-up can not be done. In fact, even with MRI scans, the fat is indistinguishable from a possible recurrence of mucocele.

The cranialization procedure of the frontal sinus is the most radical treatment of frontal mucoceles: it consists of a complete ablation of the frontal sinus mucosa, a closure of the fronto-nasal duct, and removal of the frontal sinus posterior wall.

In our opinion, the radical procedures, although associated with smaller rates of recurrence, are excessively invasive for mucoceles limited to the frontal sinus or interfering with the orbit. They have to be taken into consideration if there is an intracranial invasion or in the presence of recurrence.

We prefer a conservative procedure via the external method with osteoplastic flap after coronal incision. Such an approach has many advantages: (1) it allows the direct visualization and the complete removal of the mucocele from the frontal sinus and from the orbit; (2) it allows radiologic postoperative follow-up; (3) it is associated with a low rate of recurrence (in our experience only one recurrence in 12 patients); (4) invasiveness and complications are more limited than with the radical procedure, even if they are greater than with the endoscopic approach ones; and (5) the aesthetic results are excellent.

In conclusion, after a literature review and according to the results of our experience, we support...
open surgery in the fronto-orbital mucoceles treatment. The conservative approach through a coronal incision and a creation of osteoplastic flap is, in our opinion, preferable than both the endoscopic or the radical surgical procedures. With regard to the endoscopic procedure, the conservative approach allows a better visualization of the lesion, the possibility to directly visualize the orbital invasion and to remove without risks the mucosa of the mucocele from the periortbit. Compared with the radical approach, although associated in literature with rates of recurrence slightly higher, it is considered more suitable to the treatment of a benign lesion because it’s less invasive, assures a good drain of the frontal sinus secretions, and allows a careful radiologic post-operative follow-up.

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