CASE REPORT

Endoscopic surgical management of the fronto-ethmoido-orbital mucopyocele: a case report and literature review

Daniela Cernev[1,2], Victoria Botan[2], Vasile Cabac[1,2]

[1]"Nicolae Testemitanu" State University of Medicine and Pharmacy, Chisinau, Republic of Moldova

[2]ENT Department, "Sfanta Treime" IMPH MCH, Chisinau, Republic of Moldova

ABSTRACT

Frontal sinus mucocele are benign, pseudocystic lesions that derive from the destruction of the sinus ostium, resulting in a continuous accumulation of mucus. The process of mucocele growth leads to a progressive enlargement of the sinus cavity, thickening and erosion of its bone walls until the invasion of surrounding tissues. Mucoceles are more common in the frontal sinuses, while involvement in the sphenoid, ethmoid and maxillary sinus is rare. The frontal sinus floor is divided by the upper orbital wall; thus, an early displacement of the orbit occurs in the growth of the frontal mucocele. This paper presents one of our experiences in endoscopic surgical management of frontal sinus mucopyocele with secondary orbital complications.

KEYWORDS: mucopyocele, frontal sinus, endoscopy.

INTRODUCTION

Mucoceles of the paranasal sinuses were first described by Langenbeck (1820) under the name of hydatids and later, in 1909, Rollet suggested the name mucocele[1]. The mucoceles are more common in the frontal sinuses, while involvement of the sphenoid, ethmoid and maxillary sinus is rare[2,3]. The mucocele is a chronic, expanding lesion, lining the mucosa of the paranasal sinuses, characterized by mucous retention that, when infected, leads to mucopyocele. Their primary cause is the obstruction of the sinus ostium caused by congenital anomalies, infections, inflammation, allergies, trauma (including surgery) or a benign or malignant tumor[1]. Even benign, it tends to expand by eroding the surrounding environment of bone walls, which move and destroy structures through pressure and bone resorption[2].

Symptoms and signs of frontal mucopyocele include pain, exophthalmia, diplopia and vision loss. If not diagnosed or left untreated, erosive mucopyocele can lead to meningitis, meningoencephalitis, pneumocephaly, cerebral abscess, convulsions or CSF fistulas. They occur with a similar frequency in adults of both sexes and rarely in children younger than ten years.

The imaging of mucopyocele appears as an expansive lesion in the sinus with no air access, with the thinning and sometimes erosion of its bone walls. Computed tomographic (CT) scanning has proved to be an excellent diagnostic tool, being essential in surgical planning. Magnetic resonance imaging (MRI) may provide additional information in the examination of the orbit and may be a preferable imaging technique in case of suspected soft tissue-involved tumors[2]. Mucopyocele can lead to various complications with expansions in the orbit, the nasal and intracranial cavity, so both the computed tomography (CT) and MRI are used in differential diagnosis and mucocele assessment[3,4]. The CT scan is used for regional anatomy evaluation, especially intracranial detection, and orbital expansion with or without bone erosion. The MRI is useful in differentiating mucopyocele from neoplasms[3,5].

Depending on their content, cysts are divided into: mucocele (fluid from the cyst of a viscous, mucous nature, represented by mucin); pyocele (represented by

Corresponding author: Daniela Cernev, PhD, University Assistant, "Nicolae Testemitanu" State University of Medicine and Pharmacy, 165 Stefan cel Mare si Sfant Blv., 2004, Chisinau, Republic of Moldova
ORCID: https://orcid.org/0000-0001-8388-0901
e-mail: daniela.cernev@usmf.md
Received for publication: May 24, 2023 / **Accepted:** October 3, 2023

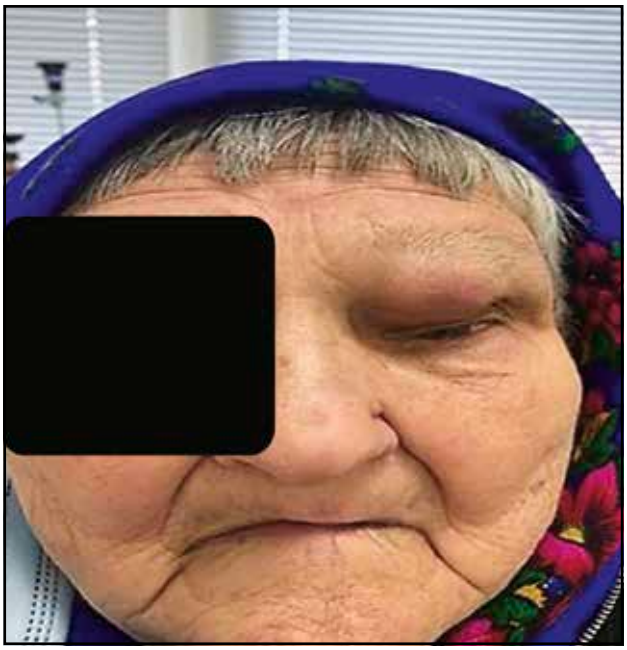

Figure 1. Image showing swelling in the left orbital superior internal angle.

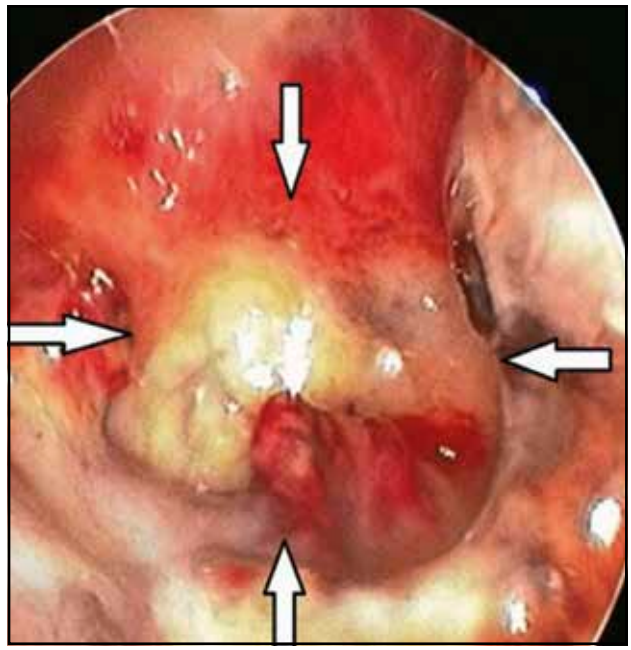

Figure 2. Endoscopy showing a reddish mass in the left nasal cavity.

pus-filled cyst); hydrocele (cyst content – serous liquid); pneumocele (air-filled formations are extremely rare and only at the frontal sinuses)[6].

The classification of frontal sinus mucopyocele is based on the orbital or endocranial extension that also determines the therapeutic approach: Type I – limited to the frontal sinus (+/- orbital extension); Type II – fronto-ethmoidal mucoceles (+/- orbital extension); Type III – erosion of the posterior wall of the frontal sinus (with subtype A – no/minimum intracranial extension and subtype B – significant intracranial extension); Type IV – erosion of the front and rear walls (with subtype A – no/minimum intracranial extension and Subtype B – with significant intracranial extension)[7].

The pathogenesis of the mucoceles of the paranasal sinuses has been described by authors via different "theories" on its appearance. The "monoglandular theory" explains the appearance of mucocele by clogging a single mucous gland, resulting in its extension, proliferation of the epithelial layer and the formation of a mucus bag. The "morphogenetic theory" refers to the congenital interruption of the development of cells of the lattice labyrinth, by analogy with odontogenic cysts. The third theory, also known as the "compression theory" states that it is a simple obstruction of the drainage channels, followed by the formation of the aggressive content of the mucocele and activation of osteoclasts, which lead to bone destruction[6].

Different surgical approaches should be considered in the treatment of mucocele and can be represented by: endoscopic approach used in small-sized mucocele with the wide opening of the ethmoidal bubble for drainage, or external surgical techniques used in bulky fronto-ethmoidal mucocele, with full excision of the capsule and insertion of a silicone tube in the frontonasal duct for epithelialization. The combined external and endoscopic endonasal approach is the method of choice under the current conditions, allowing an endoscopic control at the time of identification, enlargement and catheterization of the frontonasal duct. Drainage with marsupialization can sometimes be sufficient in particular clinical cases (e.g., old age, comorbidities, particular anatomical conformations, etc). Treatment of orbital and endocranial complications is performed in teams together with the ophthalmologist or neurosurgeon[8].

Differential diagnosis includes chronic infection, inverted papilloma, paranasal sinus carcinoma or aspergillus infection.

CASE REPORT

A female patient aged 80 years was admitted to the ENT Department of "Sfanta Treime" IMPH MCH with pronounced frontal headache, left eye pain, and progressive left exophthalmia for about 1 year. On examination, ENT clinical evaluation and nasal endoscopy, we identified a painful swelling in the left orbital superior internal angle, left exophthalmia, diplopia (Figure 1). An anterior rhinoscopy revealed a roomy nasal cavity with bilateral inferior turbinates atrophy, and a smooth reddish nasal mass was seen arising from the lateral nasal wall. The diagnostic nasal endoscopy disclosed a reddish mass in the left nasal cavity arising from the lateral nasal wall (Figure 2). The serologic blood results revealed an in-

Table 1. Serologic blood test results (complete blood count and biochemistry).

	Result		Reference values	Unit
Haemoglobin	13.5		11.5-15.0	g/dL
Red blood cells	4.73		3.8-5	x10^6/L
White blood cells	40	!!!	4.5-13	x10^/L
Lymphocytes (LY)	26.1	!!!	2.01-4.34	%
Platelets (PLT)	249		150-400	x10^L
Erythrocyte sedimentation rate (ESR)	56	!!!	0-20	mm/h
Creatinine	89	!!!	46-70	Mmol/l
Blood urea nitrogen (BUN)	11.8		18-64	Mmol/l
Alanine transaminase (ALT)	17		0-24	U/l
Aspartate transferase (AST)	20		0-25	U/l
Blood glucose test	5.5		3.3-5.6	mm/l

flammatory and infectious syndrome, leucocytosis with lymphocytosis (Table 1).

The ophthalmological examination revealed left eye diplopia, as a result of compressed nerves that control the movements of the eye globes; reduced left eye visual acuity, resulting from the pus accumulation in the frontal sinus area; left exophthalmia, caused by increased pressure of the inflamed mucosa, as well as pus accumulation in the frontal sinus; left palpebral edema due to inflammation of the frontal sinus; limited left eye movements, caused by inflammation of the frontal sinus and increased pressure posed on the nerves that control eye movements.

The patient underwent CT scanning of the paranasal sinuses (Figure 3) that showed: a cystic formation in the left frontal sinus, with expansion into the upper ethmoidal cells, with deformation and erosion of the anterior wall of the frontal sinus; enlargement in the left supraorbital region; deformation of the orbit medial wall with no involvement of the orbit; no evidence of intracranial extension; total opacification of the maxillary sinuses, and hypoplastic right frontal sinus.

Considering the CT scan result, we decided to perform an MRI scan (Figure 4) which identified a mucopyocele of the left frontal sinus – non-homogeneous mucopurulent content in the left frontal sinus and the bilateral ethmoidal cells (left > right), with erosive changes of the dorsal and lower wall of the left frontal sinus and of the right lateral wall of the left ethmoidal cells, with orbital extension (compression of the extracoronary space), with no certain

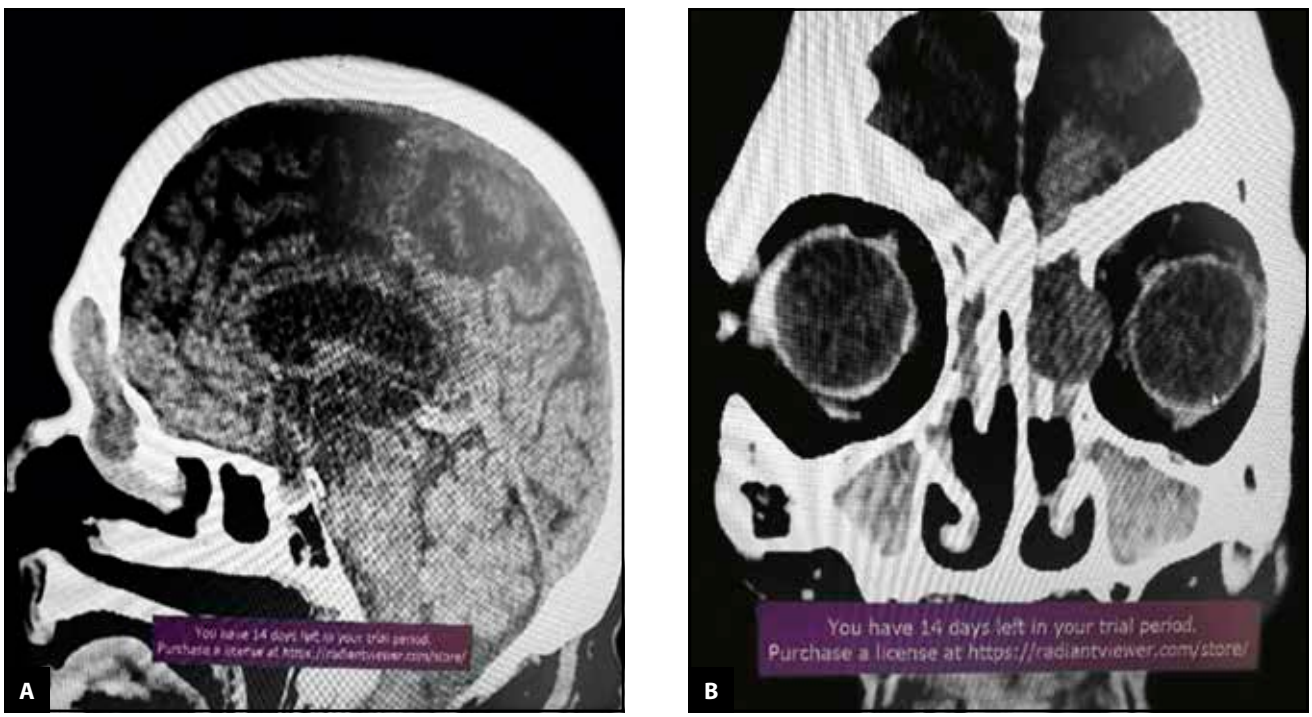

Figure 3. Cranio-facial CT scan: **A** – sagittal slice showing a convex space-occupying opacity with thinned and eroded bone walls displacing adjacent structures with a homogeneous content; **B** – coronal slice showing a left-sided well-circumscribed soft tissue mass with lateral displacement of the orbit.

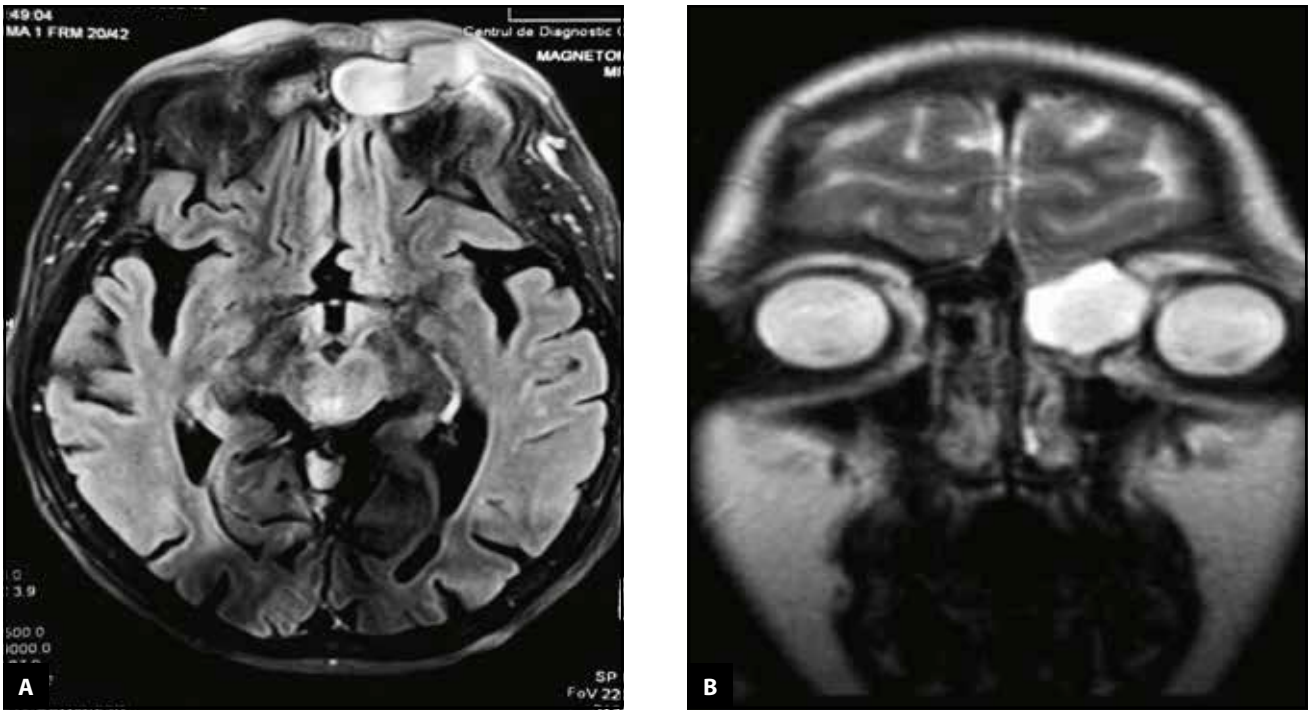

Figure 4. Cranio-facial MRI scan: **A** – axial slice shows the suspected mucocele in the left frontal sinus and ethmoidal cells; **B** – coronal slice identified a hyperattenuated homogenous expansile lesion originating from the left ethmoid sinus with left intraorbital extension.

data for cranial intra-expansion; opacification of the cavities of the maxillary sinuses.

The diagnosis was based on the objective clinical examination and the imaging evaluation results. The elective treatment in this case was the surgical one. Anterior left ethmoidectomy was performed, followed by a subsequent left frontal sinus drainage. The surgical technique involved: infundibulotomy, unciformectomy, resection of the ethmoidal bubble and other anterior ethmoidal cells, highlighting the left frontal sinus recess and removal of pathological tissues from this level to widen the frontonasal duct, as well as aspiration of the pathological content. The left maxillary sinus ostium was widened to ensure adequate frontal sinus drainage and prevent a recurrence (Figure 5).

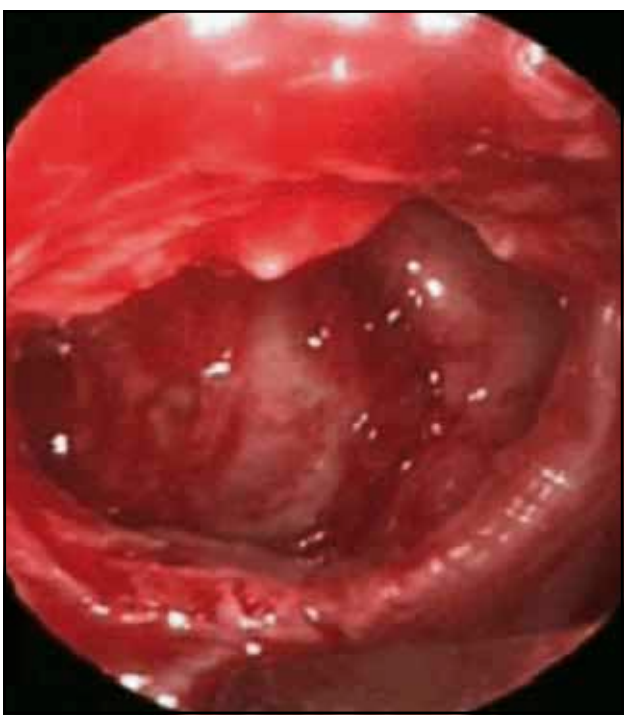

Figure 5. The postoperative nasal cavity (endoscopic view).

The treatment also included administration of NSAIDs for 5 days, antibiotics (third-generation cephalosporins (ceftriaxone 1 gram intravenously) 7 days, metronidazole 100 ml x 2 times a day 7 days), nasal decongestants 7 days.

The palpebral edema decreased over 24 h to 7 days, and the patient was discharged with satisfactory condition.

At the 1-month follow-up, the patient's overall condition was satisfactory – pink skin, no fever, painless eye opening, with no left palpebral edema and exophthalmia, absence of diplopia and improvement of visual acuity (Figure 6). At the endoscopic nasal examination, the nasal mucosa was pink, clean, without pathological eliminations (Figure 7).

DISCUSSIONS

Mucopyoceles are benign, slow-growing lesions that occur more frequently in the frontal or ethmoidal sinus, being characterised by the presence of a sac which can be

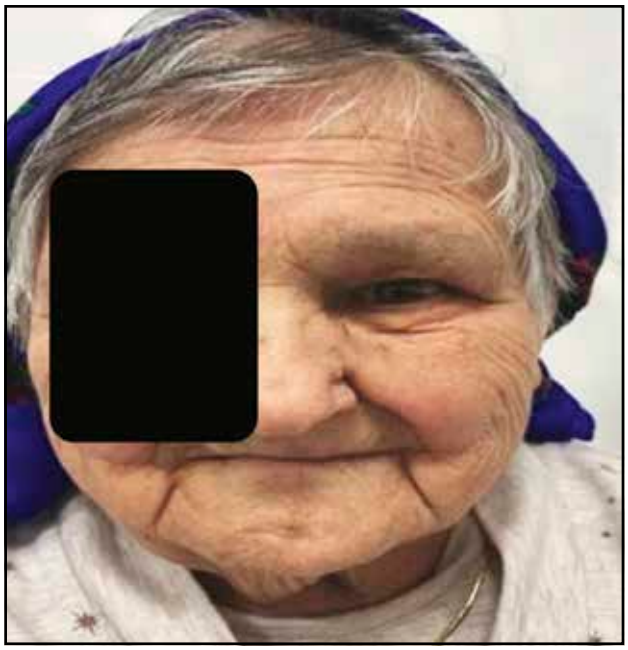

Figure 6. Postoperative photo after removal of a left fronto-ethmoido-orbital mucopyocele.

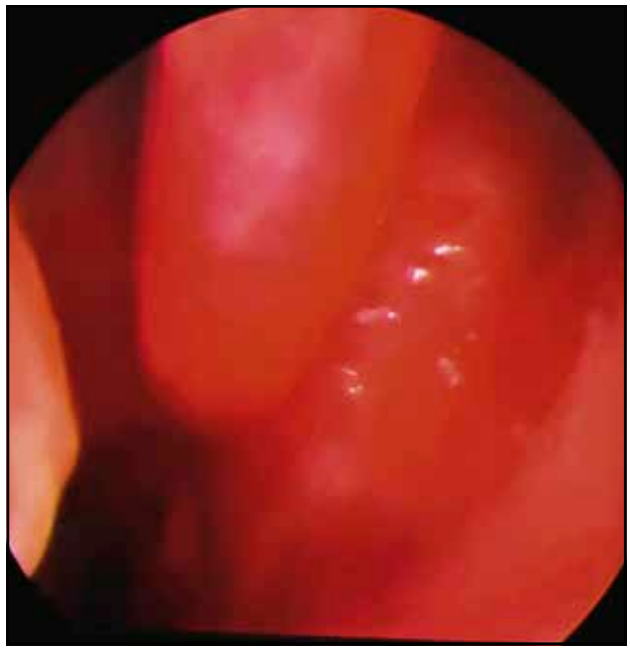

Figure 7. Endoscopic examination after 1 month.

filled with pus as a result of chronic infection. Mucopyocele expansion in the orbit due to the destruction of the orbital wall leads to proptosis and diplopia; occasionally, it can compress the optic nerve, leading to loss of vision[8].

About 60-89% of mucopyoceles occur in the frontal sinus, followed by 8-30% in the ethmoid sinuses and less than 5% in the maxillary sinus, the sphenoid sinus being less implicated. They might occur at any age, but it is mostly diagnosed in patients aged between 40 and 60; men and women are equally affected[2,3].

The etiology of the mucopyocele of the frontal sinus is multifactorial and has not yet been fully elucidated. The considerable pathological changes can lead to drainage disorders through the nasofrontal duct, which is a dominant factor in their appearance. Most often, these include chronic sinusitis, allergic reactions to the mucosa of the paranasal sinuses, lesions, anatomical developmental abnormalities of the paranasal sinuses and excretory canal, tumors, etc.[9].

Detailed histopathological studies have shown that following the obstruction of the frontonasal duct and subsequent infection in the frontal sinus cavity, a continuous stimulation of monocytes and lymphocytes leads to the production of cytokines by mucosal fibroblasts. These cytokines promote bone resorption and remodelling, leading to mucocele expansion[10]. When compared to normal frontal sinus mucosa fibroblasts, cultivated fibroblasts derived from the fronto-ethmoid mucocele have been shown to produce significantly increased levels of collagenase and prostaglandin E2[11]. There are several studies which state that the high levels of prostaglandin E2 play a major role in the osteolytic process, thus exhibiting local aggressive behaviour of these expanding masses[12].

The culture isolated from mucoceles can sometimes confirm the presence of infection. Brooks et al. showed in one study from 2001 that the most common involved bacteria were *Staphylococcus aureus*, alpha-haemolytic streptococci, *Haemophilus* species, and gram-negative bacilli, while the predominant anaerobic isolates were the species of *Propionibacterium acnes*, *Peptostreptococcus*, *Prevotella* and *Fusobacterium*[13].

The clinical presentation varies depending on the anatomical location of the mucopyocele, and it ranges from asymptomatic headaches to disabilities and visual disturbances[14,15]. Usually, the onset of symptoms is insidious. Patients with fronto-ethmoid mucocele may present frontal headache, facial asymmetry or swelling, as well as ophthalmological manifestations such as impaired visual acuity, reduced eye mobility or proptosis. Proptosis (83%) and diplopia (45%) are the most common complaints. On the objective exam, periorbital sensitivity, swelling and low visual acuity can be determined by restricted extraocular movements.

Intracranial extension due to erosion of the posterior wall of the frontal sinus can lead to meningitis, meningoencephalitis, pneumocephalus, brain abscess and seizures[15,16]. The posterior wall of the sinus is particularly prone to erosion because it is inherently thin. The tendency for bone erosion and intracranial extension has been more commonly reported in cases of infection.

The patient under the study had a mucopyocele extended to the orbit resulting in proptosis, and posterior erosion of the frontal sinus. When a mucocele becomes infected, it becomes a pooch and gives rise to symptoms

and signs of inflammation, resulting in painful symptoms present in the frontal region, which developed in the patient a year earlier. In our patient, proptosis occurred due to an increased pressure exerted by the inflamed mucosa and the accumulation of pus in the left frontal sinus. Diplopia was caused by the compression of the nerves that control the movements of the eyeball, a reduced visual acuity resulting from the accumulation of pus in the frontal sinus area. Exophthalmia was caused by an increased pressure exerted by the inflamed mucosa and the accumulation of pus in the frontal sinus. Palpebral edema occurred due to an inflamed frontal sinus. Limited eye movements were caused by inflammation of the frontal sinus and increased pressure on the nerves that control eye movements[17].

Computed tomography (CT) is currently the method of choice, showing the coronal and axial views with three-dimensional reconstruction with or without bone erosion. Moreover, the surgeon obtains important information on the individual anatomy of the frontal sinus and the other paranasal sinuses. This knowledge is an important prerequisite for planning surgical interventions. However, magnetic resonance imaging (MRI) may be helpful whenever there is extension beyond the sinus boundaries and in differentiating mucocele from meningoencephaloceles and other expansile sinonasal lesions[18].

Magnetic resonance imaging is useful when the diagnosis is uncertain and it is necessary to differentiate between different types of soft tissues within the sinonasal cavities, especially if the mucocele formed secondary to a neoplasm. Additionally, when the mucocele extends intracranially, the MRI offers superior imaging of the surrounding brain. The usual signal characteristics for a mucocele are low T1 and high T2, but variations commonly occur depending on the presence of blood and the water content of the mucocele[19].

Surgery is the treatment of choice in the frontal mucopyocele. The surgical approach depends on the extension of the mucopyocele. Since most frontal sinus mucopyocele are medially located, they can be treated by transnasal endoscopic approach in order to restore the anatomical and functional integrity of the frontonasal duct and frontal sinus. Depending on the size of the process and the involvement of the surrounding anatomical structures, the extent of a surgical intervention ranges from minimally invasive endoscopic surgery to craniotomy with or without sinus obliteration[10]. Attention to postoperative nasal hygiene, including nasal irrigation and topical steroids, is critical. If the contents of the mucocele are purulent or if the microbiological cultures are positive, oral antibiotics are used. A close endoscopic follow-up postoperatively should be continued until the cavity heals and mucociliary clearance re-establishes[20].

However, there is a number of relative contraindications for an endonasal endoscopic approach, such as the presence of any sinonasal involvement that prevents the drainage of the ostium (e.g., osteoma), the onset of mucocele in the outermost and posterosuperior region of the sinus; and the presence of major sclerosis on the sinus floor. In cases where intranasal treatment is challenging, an external pathway[21] or an approach combined with external treatment under endoscopic control may be applied. The combined approach should be used in severe cases where the anatomy, the disease progression or previous surgery might restrict the endoscopic view and access to the frontal sinus, as well as in cases where a fistulous tract is already present[22].

CONCLUSIONS

Mucoceles are benign lesions of expansive characteristic that may cause severe complications at orbital and intracranial levels. Frontal mucoceles may occasionally present with ophthalmic manifestations such as proptosis. Being benign and curable, early recognition and management of mucoceles is of paramount importance. A high index of suspicion and appropriate imaging studies are necessary for the diagnosis of mucocele. The endoscopic endonasal approach is the treatment of choice as this is a safe and effective technique in most of the cases of mucocele.

Conflict of interest: The authors have no conflict of interest.

Contribution of authors: All the authors have equally contributed to this work.

Financial discloser: There are no financial disclosures of the authors.

Fundings: There are no funds for this article.

Authors' information:

Daniela Cernev, PhD, University Assistant, "Nicolae Testemitanu" State University of Medicine and Pharmacy, Chisinau, Republic of Moldova. E-mail daniela.cernev@usmf.md. ORCID: https://orcid.org/0000-0001-8388-0901.

Victoria Botan, ENT Practioner, ENT Department, "Sfanta Treime" IMPH MCH, Chisinau, Republic of Moldova. E-mail: botan.vica@gmail.com.

Vasile Cabac, PhD in Medical Sciences, Associate Professor, "Nicolae Testemitanu" State University of Medicine and Pharmacy, ENT Department, "Sfanta Treime" IMPH MCH, Chisinau, Republic of Moldova. E-mail: vasile.cabac@usmf.md. ORCID: https://orcid.org/0000-0003-4899-7375.

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