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Recurrent of a Giant Fronto-Ethmoidal Mucocele with Orbital Extension: A Case Report

N. Yassine[1*], A. Hebbezni[1], B. Boutakioute[1], M. Ouali Idrissi[1], N. Cherif Idrissi El Ganouni[1]

[1]Department of Radiology Arrazi, Mohammed VI University Hospital center, Marrakesh, Morocco

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***Corresponding author:** N. Yassine
Department of Radiology Arrazi, Mohammed VI University Hospital center, Marrakesh, Morocco

Abstract **Case Report**

Mucoceles of the paranasal sinuses are benign expansive pseudo-cystic lesions, affecting mainly adults. These rare lesions are usually evolving at low noise and are most often revealed by neurological or ophthalmological complications. We report a case of a recurrent fronto-ethmoidal mucocele with orbital extension. This was a 69-year-old patient with a previous history of endoscopic marsupilization of mucocele by a private practitioner 10 years back, who presented frontal headaches associated with a left-sided forehead swelling and chronic left proptosis evolving for 3 years, in whom clinical examination revealed fluctuant swelling present in the forehead region above the left eye and unilateral left sided proptosis. Cranio-facial computed tomography (CT) and magnetic resonance imaging (MRI) were in favor of a left fronto-ethmoidal mucocele with homolateral intra-orbital extension. The patient was operated by an endoscopic surgical approach. The outcomes were favorable with regression of headaches and resolution of proptosis. The fronto-ethmoidal mucocele, although benign, has an aggressive potential in the absence of treatment either towards the endocranium or the orbit. Imaging plays a crucial role in the diagnosis, preoperative planning and monitoring of mucoceles and CT and MRI should be used as complementary investigations of sino-nasal pathology. Treatment of mucoceles is surgical and recurrences are uncommon. Although the fronto-ethmoidal is a benign lesion, it is may be responsible for complications involving the functional or even vital prognosis. An early diagnosis and treatment allow to improve mucoceles outcomes.

Keywords: Fronto-ethmoidal mucocele, Orbital extension, Imaging, CT, MRI.

INTRODUCTION

Mucoceles of the paranasal sinuses are benign expansile cyst-like lesions of the sinuses that are caused by ostial blockage and gradual accumulation of mucoid secretions and desquamated epithelium [1]. The main cause of sinus mucocele is represented by iatrogeny in particular the endonasal surgery [2].

Mucoceles can occur at any age but mostly observed between the fourth and seventh decades and both sexes are equally affected [3]. They commonly develop in the frontal and the ethmoid sinuses [4, 5). Despite their histological benignity, they are aggressive and destructive lesions and may extend to the orbital and intracranial structures.

The clinical presentation of mucoceles varies according to the sinus affected, the size, the local extension and the involvement of adjacent tissues as well as the complications caused [6]. Their progression is very low and are often asymptomatic. Therefore, these rare tumors are often associated with a diagnostic delay as they are diagnosed at the stages of serious orbital or endocranial complications [7].

Computed tomography (CT) and magnetic resonance imaging (MRI) are the methods of choice for the diagnosis of mucoceles and the cause of obstruction. CT and MRI seem to play complementary roles in mucocele imaging. CT can determine the regional anatomy and extent of the lesion, specifically the intracranial extension and the bone erosion, while MRI is useful in differentiating mucoceles from neoplasms or identifying an underlying tumor as the cause of obstruction [8].

Surgery is the treatment of choice and consists of decompression, ideally endoscopic, and carrying out a drainage and a wide marsupialization of the mucocelic cavity. Recurrences of mucoceles after treatment can be seen after 3 to 4 years [9].

We present a case of a 69-year-old patient with a recurrent mucocele of the fronto-ethmoidal sinus with orbital extension.

CASE PRESENTATION

A 69-year-old man presented with a 3 years history of worsening frontal headaches associated with a left-sided forehead swelling and chronic left proptosis. The patient denied clinical symptoms such as diplopia, decreased vision, nasal obstruction and nasal discharge. He had no significant of history of general disorder or orbito-facial trauma or rhinosinusitis.

There was previous history of endoscopic marsupilization of mucocele by a private practitioner 10 years back. The patient had no particular family history.

On clinical examination, we noticed a fluctuant swelling present in the forehead region above the left eye. There was unilateral left sided and non axil proptosis and the eyeball was pushed inferiorly and laterally. Vision was normal in left eye with full extraocular movements in all directions. Pupillary reaction to light and accommodation were normal. Fundus examination was normal. Anterior rhinoscopy revealed a nasal septal deviation to the left.

The rest of the general examination including the neurological examination was normal. A facial CT after the injection of an intravenous contrast agent was performed and revealed a well-defined mass centered on the left frontal hemisinus, with oval shape and polylobed contours, spontaneously isodense, with discreetly high-density areas after injection of contrast, measuring approximately 64 x 53 x 70 mm. The mass seemed to extend to the anterior ethmoidal cells and the homolateral frontal sinus (Figure 1).

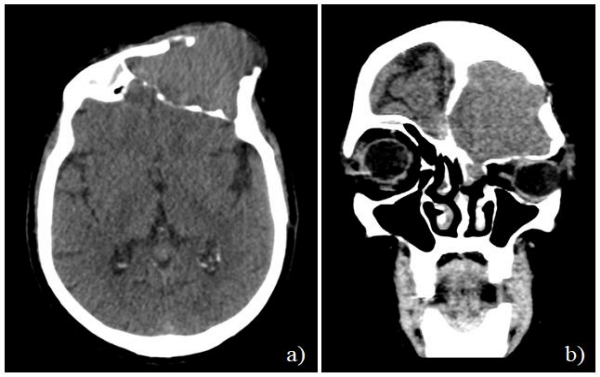

Figure 1: Facial CT without (a) and with contrast (b), soft-tissue window, axial (a) and coronal (b) sequences: mass centered on the left frontal hemisinus, spontaneously isodense (a), with discreetly high-density areas after injection of contrast (b), with extension to the homolateral frontal sinus (a) and the anterior ethmoidal cells (b)

It was responsible for a bulging of the cortical bones with bone erosion of the anterior and the posterior walls of the homolateral frontal sinus, the orbital roof, the cribriform plate of the ethmoid bone, the frontal intersinus septum, the homolateral nasal process of the frontal bone and the lamina papyracea (Figure 2).

The mass appeared also to invade the homolateral orbit, lying in apposition to and pressing on the superior rectus muscle of the left eye. Thus, it was responsible of an inferolateral left globe displacement and a proptosis scaled as degree I.

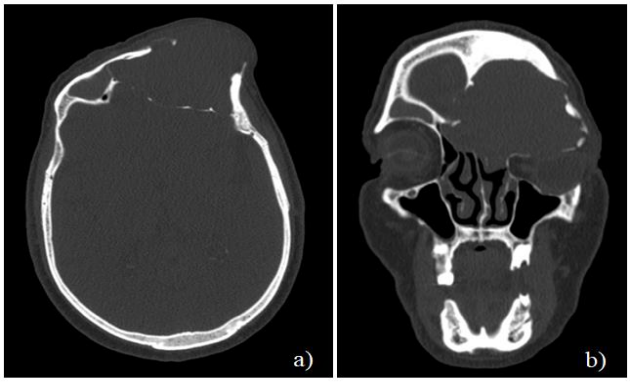

Figure 2: Facial CT, bone window, axial (a) and coronal (b) sequences: the mass is responsible of for a bulging of the cortical bones with bone erosion of the anterior and the posterior walls of the homolateral frontal sinus, the frontal intersinus septum and the orbital roof

The mass seemed also to be in close contact with the cerebral parenchyma which appeared normal and was suggestive of a recurrent fronto-ethmoidal mucocele with orbital extension.

To better characterize the mass and to study the intra-orbital and intracranial extension, a facial MRI was performed and revealed a well-limited left fronto-ethmoidal mass with moderate high-signal in T1 and FLAIR sequences (Figure 3). The mass showed heterogenous hypo-signal intensity in T2 sequence and hypo-signal intensity on diffusion sequence (Figure 4). It was not enhanced on gadolinium-enhanced MRI.

The mass was responsible for - as the facial CT showed- an extension to the homolateral orbit with scalloping on the superior rectus muscle and a proptosis scaled as grade I.

It extended endocranially with a scalloping on the left frontal parenchyma with discreet deviation of the midline and without any signal abnormality or detectable contrast (Figure 5).

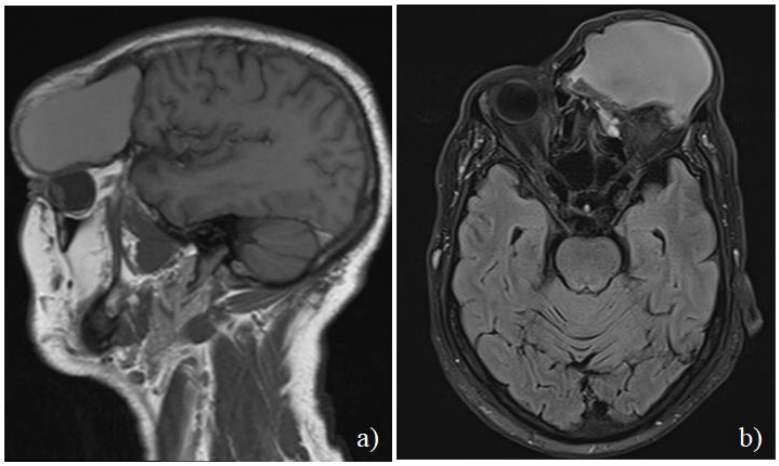

Figure 3: Facial MRI, T1 (a) and FLAIR (b) weighted sequences, sagittal (a) and axial (b) sections: a well-limited left fronto-ethmoidal mass with moderate high-signal intensity

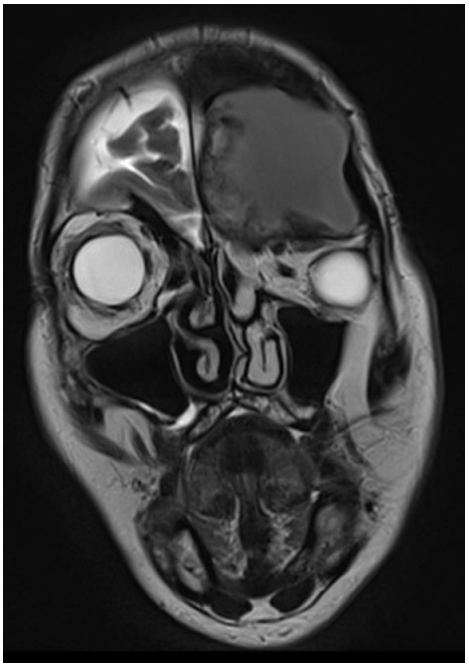

Figure 4: Facial MRI, T2 weighted sequences, coronal section: The mass showed heterogenous hypo-signal intensity in T2 sequence

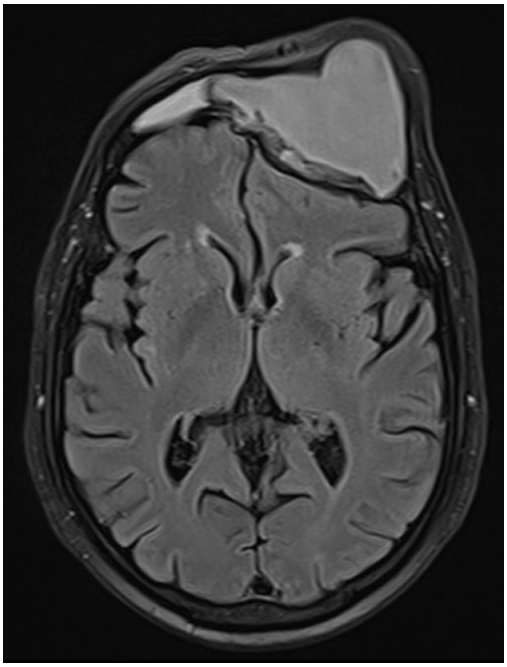

Figure 5: Facial MRI, FLAIR sequence, axial section: the mass extended endocranially with a scalloping on the left frontal parenchyma

The radiologist concluded to a left recurrent fronto-ethmoidal mucocele with orbital extension and according to the classification proposed by Thiagarajan [10], it was categorized as type IIIa .The patient was

referred to otorhinolaryngology department for surgical management. After endoscopic examination, a fronto-ethmoidal mucocele with orbital extension was diagnosed and endoscopic sinus surgery with frontal-ethmoidectomy, mucocele excision and drainage was performed. Histopathological examination confirmed the diagnosis. In the follow-up visits, the evolution was favorable with good healing of the operative wound, regression of headaches and resolution of the proptosis. One year after surgery, there was no recurrence of the mucocele. The patient is now being monitored on a yearly basis.

DISCUSSION

The term "mucocele" was first introduced by Rollet in 1896 [11]. Mucoceles of the paranasal sinuses are slow growing benign cyst-like lesions lined with respiratory mucosa and filled by mucoid substance [1].

Their etiology is unclear but they usually develop due to sinus ostium obstruction, preceded by chronic inflammation, previous craniofacial trauma or surgery, nasal polyps, benign neoplasm (osteoma or fibrous dysplasia), and malignant or metastatic tumors [8, 12]. An interesting study by Raynal *et al*., [2] showed that iatrogeny is the leading cause of sinus mucocele, with a particular and new incidence for endonasal surgery. However, spontaneous forms have been described [13].

Mucoceles represent a rare condition [14], although their incidence seems to be increasing significantly in recent years, due development of imaging techniques and nasal endoscopy for diagnostic purposes. They can develop at any age, but the majority of mucoceles are diagnosed between ages of 40 to 70 years old, as in our patient's case. Males and females are equally affected [3].

They most commonly develop in the frontal sinus (70–80%), followed by the ethmoid (25%), fronto-ethmoidal (10–14%), and maxillary (3% or less) sinuses. Sphenoid sinus mucoceles are rare [15]. Mucoceles are more frequently located in the anterior ethmoid sinuses because they are smaller, with smaller ostia and more numerous cells than posterior ethmoid sinuses [16]. Mucocele of the anterior ethmoid sinus frequently coexists with frontal mucocele (fronto-ethmoidal mucoceles) [17].

As for the pathogenesis, ostial obstruction and chronic inflammation are the two most commonly accepted factors in the genesis of mucoceles. However, it would appear that an additional infection may precipitate the formation of a mucocele [2]. Chronic inflammation leads to the release of some mediators at the capsule of the mucocele which degrades the bone [14]. In addition, the continuous production and the gradual accumulation of thick mucus within the mucocele cavity is responsible for the expansion of the affected sinus, the blowing and the erosion of the adjacent bones and the extension to the adjacent anatomical structures (sinuses, orbit, endocranium, skin), which can be responsible for complications involving the visual or vital prognosis [18].

The clinical presentation of mucocele varies depending on its location, the size and the degree of extension to the adjacent anatomical structures. The onset of symptoms is usually insidious. When the mucocele exceeds the limits of the sinus involved, it becomes symptomatic. The diagnosis is made at this stage in the majority of cases. The clinical signs of discovery are represented by oculo-orbital, naso-sinusal or neurological manifestations [19].

Patients with frontoethmoidal mucoceles may develop frontal headaches, facial asymmetry, or swelling, as well as ophthalmological manifestations, such as impaired visual acuity, reduced ocular mobility or proptosis. Proptosis and diplopia seem to be the commonest ocular manifestation of the fronto-ethmoidal location [20].

The direction of proptosis helps in localizing the sinus involved. Lesions near the orbital apex push the globe forward, and lesions arising from the fronto-ethmoidal complex push the eyeball forward, laterally, and downward [3].

Intracranial extension through erosion of the posterior wall of the frontal sinus can lead to meningoencephalitis, pneumocephalus, brain abscess, seizures or cavernous sinus fistula [21]. The posterior sinus wall is particularly prone to erosion because it is inherently thin. The tendency for bony erosion and intracranial or orbital extension is greater in the presence of acute infection of mucoceles (mucopyocele).

Radiologically, imaging plays a crucial role in the diagnosis, preoperative planning and monitoring of mucoceles [22].

The CT scan is the first-line imaging procedure to be performed in case of clinical or endoscopic suspicion of a mucocele. It remains the gold standard for the diagnosis and the surgical planning because it can visualize bone involvement and extension of the lesion in details. MRI is indicated to differentiate between mucocele and other tumors or inflammatory lesions through its signal intensity and enhancement characteristics or in case of intra-orbital or intracranial extension and in case of sphenoidal location [8].

On CT, mucoceles are generally isodense to the brain parenchyma without peripheral enhancement and with attenuation values ranging from 10 to 40 HU, which reflects the hydration and the protein

concentration of the mucoid contents [8]. Higher attenuation values are indicative of chronic lesions [17]. The presence of a thin peripheral enhancement line is indicative of infected mucoceles [23].

In addition, mucoceles may contain non enhancing hyperdense stippled areas, looking like multiple fine calcifications. They represent inspissated, dehydrated mucocele content [24].

CT represents the preferred tool to evaluate bone erosions and is an useful method to evaluate intracranial and intra-orbital extension [22].

In total, there are three criteria for CT diagnosis of a mucocele: homogeneous isodense mass, clearly defined margin, and patchy osteolysis around the mass. Erosion of the sinus wall with marginal sclerosis is also an indicative finding [22].

As mentioned above, MRI is crucial for differentiating a tumor from a mucocele, delineating the extension of the lesions, and ruling out an underlying tumor causing ostium obstruction [8].

Mucoceles usually exhibit T2-weighted hyper-intensity (indicative of high water content of the lesions), and T1-weighted hypo to high-intensity (indicative of low to high protein concentration and mucus viscosity), which reflect the increased water concentration, the different proteinaceous sinus contents or the hemorrhagic content of the lesion [24]. This aspect represents a pathognomonic MRI finding for mucoceles [25].

Young mucoceles contain mucus material rich in water, while chronic mucoceles can have any combination of signal intensities, as signal intensities in T1WI and T2WI, reflecting the degree of hydration of the mucus content, and the concentration of proteins [17].

Gadolinium enhanced MRI should always be performed when dealing with sino-nasal pathology, as there is a significant overlap in signal intensities of mucoceles, tumors and obstructed sinuses on unenhanced MRI. Mucoceles should strictly be devoid of enhancement after administration of contrast agent and the presence of a central enhancement within the lesion or even nodular peripheral enhancement should suggest a coexisting tumor [26].

In cases of mucoceles with proteinaceous content, mucoceles may become almost void of signal on T1W and T2W images, like that of air. CT helps in establishing the correct diagnosis, as the inspissated content would be of high density [25]. Thus, CT and MRI are complementary in such complicated cases.

The radiological differential diagnoses of mucoceles include: dermoid cysts, histiocytosis, fungal and tuberculosis infections, fronto-orbital cholesterol granuloma, and other uncommon neoplasms. Because of higher hyper-intensity from other lesions on T1W images, the differentiation is usually easy on MRI [25].

Finally, Thiagarajan [10] proposed a classification of frontal and fronto-ethmoidal mucoceles. They have been classified into 5 types depending on their extent:

- **Type I:** the mucocele is limited to the frontal sinus only with or without orbital extension.
- **Type II:** the mucocele is involving the frontal and ethmoidal sinuses with or without orbital extension.
- **Type IIIa:** the mucocele erodes the posterior wall of the frontal sinus with minimal or no intracranial involvement.
- **Type IIIb:** the mucocele erodes the posterior wall with major intra cranial extension.
- **Type IV:** the mucocele erodes the anterior wall of the frontal sinus.
- **Type Va**: there is erosion of both anterior and posterior walls of frontal sinus without or minimal intracranial extension.
- **Type Vb:** there is erosion of both anterior and posterior walls of frontal sinus with a major intracranial extension.

Treatment of mucoceles is surgical and is based on external or endonasal access routes [27]. Transnasal endoscopic surgery of mucoceles is currently accepted as the method of choice in the management of mucoceles due to its low iatrogenicity, its excellent efficiency and less morbidity. External approaches are aggressive procedures with high morbidity and are currently used only in the case of highly invasive lesions [17]. The surgical approach is based on the size, location, and extent of the mucocele. In the presence of infection, adjuvant antibiotic treatment is indicated.

Surgery involves decompression, ideally endoscopic, and consists of drainage and wide marsupialization of the mucocele cavity. A good resection of the sinus wall must be performed to ensure good drainage and avoid recurrences [22].

Recurrences of mucoceles are uncommon and can be seen after after 3 to 4 years (an average of 3.8 years) [9], and intra-orbital extension is considered the most frequent risk factor [1].

When diagnosed and treated in time (marsupialization), the prognosis is good. Delayed treatment can lead to complications that can be major: intra-orbital rupture of the lesion, frontal epidural abscess … [28].

CONCLUSION

Mucoceles of the paranasal sinuses are rare benign lesions. Frontal and fronto-ethmoidal mucoceles are the most frequent. Their severity is due to their aggressive potential towards adjacent anatomical structures which may be associated with functional or even vital prognosis.

CT and MRI are the methods of choice for diagnosing mucoceles of the paranasal sinuses and are of major importance for the treatment plan. Each method seems to have its own advantages, and they should be used as complementary investigations of sino-nasal pathology. On CT, mucocele appears as a homogeneous isodense mass without peripheral enhancement and with clearly defined margin and patchy osteolysis around. MRI usually demonstrate T2-weighted hyper-intensity and T1-weighted hypo to high-intensity without enhancement after injection of contrast.

Their management was improved with the advent of imaging and endonasal surgery which is considered as the gold standard.

REFERENCES

1. Scangas, G. A., Gudis, D. A., & Kennedy, D. W. (2013, September). The natural history and clinical characteristics of paranasal sinus mucoceles: a clinical review. In *International forum of allergy & rhinology* (Vol. 3, No. 9, pp. 712-717).
2. Raynal, M., Peynegre, R., Beautru, R., & Coste, A. (1999). Mucocèles sinusiennes et iatrogénie chirurgicale. In *Annales d'oto-laryngologie et de chirurgie cervico-faciale* (Vol. 116, No. 2, pp. 85-91).
3. Courson, A. M., Stankiewicz, J. A., & Lal, D. (2014). Contemporary management of frontal sinus mucoceles: a meta-analysis. *The Laryngoscope*, *124*(2), 378-386.
4. Natvig, K., & Larsen, T. E. (1978). Mucocele of the paranasal sinuses: A retrospective clinical and histological study. *The Journal of Laryngology & Otology*, *92*(12), 1075-1082.
5. Rao, V. M., & El-Noueam, K. I. (1998). Sinonasal imaging: anatomy and pathology. *Radiologic Clinics of North America*, *36*(5), 921-939.
6. Kim, Y. S., Kim, K., Lee, J. G., Yoon, J. H., & Kim, C. H. (2011). Paranasal sinus mucoceles with ophthalmologic manifestations: a 17-year review of 96 cases. *American Journal Of Rhinology & Allergy*, *25*(4), 272-275.
7. Zainine, R., Loukil, I., Dhaouadi, A., Ennaili, M., Mediouni, A., Chahed, H., ... & Besbes, G. (2014). Complications ophtalmiques des mucocèles rhino-sinusiennes. *Journal français d'ophtalmologie*, *37*(2), 93-98.
8. Tsitouridis, I., Michaelides, M., Bintoudi, A., & Kyriakou, V. (2007). Frontoethmoidal mucoceles: CT and MRI evaluation. *The Neuroradiology Journal*, *20*(5), 586-596.
9. Galiè, M., Mandrioli, S., Tieghi, R., & Clauser, L. (2005). Giant mucocele of the frontal sinus. *Journal of Craniofacial Surgery*, *16*(5), 933-935.
10. Thiagarajan, B. (2012). Fronto-ethmoidal mucocele with intracranial extension, an interesting case report and literature review. Internet version 6. ENT Scholar.
11. Alberti, P. W., Marshall, H. F., & Black, J. I. (1968). Fronto-ethmoidal mucocoele as a cause of unilateral proptosis. *The British Journal of Ophthalmology*, *52*(11), 833-888.
12. Molteni, G., Spinelli, R., Panigatti, S., Colombo, L., & Ronchi, P. (2003). Voluminous frontoethmoidal mucocele with epidural involvement. Surgical treatment by coronal approach. *Acta otorhinolaryngologica italica*, *23*(3), 185-190.
13. Klossek, J. M., Minet, A., & Fontanel, J. P. (1996). Les mucocèles sinusiennes: aspects cliniques: Les mucocèles sinusiennes. *Les Cahiers d'oto-rhino-laryngologie, de chirurgie cervico-faciale et d'audiophonologie*, *31*(7), 413-414.
14. Kim, D. W., Sohn, H. Y., Jeon, S. Y., Kim, J. P., Ahn, S. K., Park, J. J., ... & Hur, D. G. (2013). Ethmoidal mucocele presenting as oculomotor nerve paralysis. *Clinical and Experimental Otorhinolaryngology*, *6*(2), 103-106.
15. Arrue, P., Kany, M. T., Serrano, E., Lacroix, F., Percodani, J., Yardeni, E., ... & Manelfe, C. (1998). Mucoceles of the paranasal sinuses: uncommon location. *The Journal of Laryngology & Otology*, *112*(9), 840-844.
16. Som, P. M., & Shugar, J. M. (1980). The CT classification of ethmoid mucoceles. *Journal of Computer Assisted Tomography*, *4*(2), 199-203.
17. Lloyd, G., Lund, V. J., Savy, L., & Howard, D. (2000). Optimum imaging for mucoceles. *The Journal of Laryngology & Otology*, *114*(3), 233-236.
18. Hurley, D. B., Javer, A. R., Kuhn, F. A., & Citardi, M. J. (2000). The endoscopic management of chronic frontal sinusitis associated with frontal sinus posterior table erosion. *American Journal of Rhinology*, *14*(2), 113-120.
19. Benedikt, R. A., Brown, D. C., Roth, M. K., Geyer, C. A., & Ghaed, V. N. (1991). Spontaneous drainage of an ethmoidal mucocele: a possible cause of pneumosinus dilatans. *AJNR: American Journal of Neuroradiology*, *12*(4), 729-731.
20. Slootweg, P. J., & El-Naggar, A. K. (2018). World Health Organization 4th edition of head and neck tumor classification: insight into the consequential modifications. *Virchows Archiv*, *472*(3), 311-313.
21. Nakayama, T., Mori, K., & Maeda, M. (1998). Giant Pyocele in the Anterior Intracranial Fossa—Case Report—. *Neurologia medico-chirurgica*, *38*(8), 499-502.

22. Arnavielle, S., Vignal-Clermont, C., Galatoire, O., Jacomet, P. V., Klap, P., Boissonnet, H., ... & Morax, S. (2010). Compressive optic neuropathy related to an anterior clinoid mucocele. *Journal francais d'ophtalmologie*, *33*(3), 208-e1.
23. El Kettani, N. E. C., El Quessar, A., El Hassani, M. R., Chakir, N., & Jiddane, M. (2010). Fronto-ethmoidal mucocele. *Presse Medicale Paris Fr 1983*, 39(5), 612-613.
24. Van Tassel, P., Lee, Y. Y., Jing, B. S., & De Pena, C. A. (1989). Mucoceles of the paranasal sinuses: MR imaging with CT correlation. *American journal of neuroradiology*, *10*(3), 607-612.
25. Morgan, D. E. (2007). Clinical Magnetic Resonance Imaging, 3rd ed., vol. 3. *Am J Roentgenol*, 188(2), W216-W216.
26. Lanzieri, C. F., Shah, M., Krauss, D., & Lavertu, P. (1991). Use of gadolinium-enhanced MR imaging for differentiating mucoceles from neoplasms in the paranasal sinuses. *Radiology*, *178*(2), 425-428.
27. Khong, J. J., Malhotra, R., Selva, D., & Wormald, P. J. (2004). Efficacy of endoscopic sinus surgery for paranasal sinus mucocele including modified endoscopic Lothrop procedure for frontal sinus mucocele. *The Journal of Laryngology & Otology*, *118*(5), 352-356.
28. Herndon, M., McMains, K. C., & Kountakis, S. E. (2007). Presentation and management of extensive fronto-orbital-ethmoid mucoceles. *American journal of otolaryngology*, *28*(3), 145-147.