



Unilateral Proptosis A Rare Presentation of Posterior Ethmoidal Sinus Mucocele

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Abstract

Paranasal sinus mucoceles are epithelium-lined cystic masses usually resulting from obstruction of sinus ostia. They most frequently occur in the frontal and ethmoid sinuses. While ophthalmologic symptoms are most common, patients also report rhinological or neurological complaints. The close proximity of paranasal sinus mucoceles to the orbit and skull base predisposes the patient to significant morbidity. Computed tomography displays a non-enhancing homogenous mass with expansion of bony walls. Magnetic resonance imaging reveals variable intensity of T1-weighted images and a hyperintense mass on T2-weighted images. Histopathologically mucoceles have features of respiratory mucosa with areas of reactive bone formation, hemorrhage, fibrosis, and granulation tissue. Surgical excision is the standard treatment with trends towards endoscopic technique.

INTRODUCTION

Mucocele is a clinical entity, which was first described by Langenbeckin 1818. Mucoceles are the most common lesions causing expansion of the paranasalsinuses. It is an epithelial lined mucus containing sac almost completely filling a paranasal sinus cavity & being capable of expansion. It is a relatively uncommon condition most frequently occurring in the fronto-ethmoidal region. The sinuses most commonly involved in decreasing order of frequency are the frontal,

ethmoidal, sphenoid & maxillary sinuses. Paranasal sinus mucoceles predominantly occur in the fronto-ethmoid region (64%), followed by the maxillary sinus (18.6%), the sphenoid sinus(8.4%) and the posterior ethmoid sinus (6.7%).¹ Mucoceles usually enlarge slowly over many years, frontal, ethmoid & sphenoidal mucoceles frequently causes visual disturbances. Ethmoidal mucoceles occur most often in the anterior ethmoid region & may be suspected on clinical examination by the presence of proptosis& lateral

displacement of the globe.²Posterior ethmoid-mucoceles may falsely give the impression of being localized in the sphenoid sinus because they impinge upon the apex of the orbit causing visual dysfunction including proptosis, diplopia, ptosis & impairment of visual acuity.¹

CASE REPORT

30 yrs female with no co-morbidities in past presented in medicine outpatient clinic with complaints of persistent frontal headache associated with drooping of left upper eyelid & difficulty in movement of left eye in all directions since 7 days with progressive dimension of vision over 10 days. Patient has no significant past medical history or of addiction or trauma to the head. On general examination patient was afebrile, Pulse=84 beats/min, Blood Pressure=114/70 mm of mercury

LOCAL EXAMINATION

Right eye -pupil normal in size, reacting to light, vision 6/6, corneal reflex present, fundus examination normal

Left eye -pupil normal in size, sluggishly reacting to light, vision- finger counting present at a distance of 0.5 meter, corneal reflex- absent, severe external ophthalmoplegia with severe ptosis present, fundus shows papilloedema

ENT examination - Anterior Rhinoscopy no blackish discoloration or mucopurulent discharge was seen. Diagnostic nasal endoscopy was normal. MRI Brain: 21.4 x30.2x23.4 mm (Trans x AP X CC) sized well-defined expansile lesion in posterior ethmoid air cells on the left side was seen. It was causing pressure erosion/thinning of wall of the air cells, predominantly on the lateral aspect.

Severe compression & displacement of medial rectus & superior oblique muscle and also mild compression on the posterior orbital segment of the optic nerve, minimal relative anterior displacement of the left globe which was mild hyper intense on T1 & intermediate in intensity on T2 suggesting organized debris within it.

Stagnated fluid signal was seen on its anterior aspect with minimal peripheral smooth enhancement on post gadolinium images.

Impression: Benign morphology expansile lesion in the posterior ethmoid air cells on the left side, possibly a Mucocele.

Patient was advised HRCT PNS from ENT side but was not done. Further, a neuro surgery opinion was sought but no active management was advised from their side, since there was no intracranial involvement, however a repeat scan was advised after 2 month or earlier if symptoms worsen but we lost contact with the patient there after.

DISCUSSION

A mucocele is defined as a mucous filled epithelium lined sac. Mucoceles commonly involve ethmoidal and frontal sinuses. Mucoceles are commonly caused due to obstruction to drainage channel of paranasal sinuses. These expansile cystic masses are sometimes filled with mucopurulent secretions.³ Sometimes, associated bone destruction is also evident.⁴ Mucoceles are rather common in frontal sinuses, next come the ethmoidal sinuses. Isolated mucoceles involving ethmoidal sinuses are rather rare. They always occur in combination with frontal / sphenoid mucoceles

Mucocele have been postulated to form due to obstruction of sinus ostia following chronic infections / allergic reactions involving paranasal sinuses. Previous trauma / surgery can also cause obstruction to sinus outflow⁵ channels causing formation of mucoceles. Ethmoidal mucoceles if present in isolation could be caused by endoscopic ethmoidectomy. Some studies have reported occurrence of isolated ethmoidal mucoceles even 10 years after surgery.⁶ Paranasal sinuses continues to expand slowly owing to pent up mucous secretions. Continuing expansion of this cyst puts pressure on the bony walls of paranasal sinuses, causing bony erosion and remodeling. Unchecked extension of sinus cavity can cause

extension of mucocele into the orbit, nasopharynx and cranial cavity.⁴ In addition to pressure changes, inflammatory mediators like prostaglandins, interleukins and tumor necrosis factor present within mucoceles also contribute to their expansion capability.

Clinical features

Usually patients with mucoceles involving paranasal sinuses do not have nasal / sinus symptoms. Only symptoms they present with are ophthalmological in nature.

1. Pain: This is commonly periorbital in nature, caused by inflammation and stretching of nasal and sinus mucosa, rarely from dura. Pain is usually transmitted by the trigeminal nerve.
2. Progressive proptosis
3. Visual disturbances (diplopia/blurring of vision): This is caused by erosion of the bony casing around optic nerve. Any further expansion of mucocele will cause compression of optic nerve compromising its blood supply. Infections from mucocele can reach the optic nerve when the bony casing around the optic nerve is breached by the enlarging mucocele.⁷
4. Epiphora
5. Impaired ocular mobility.

Diagnosis

Conventional radiography

Skull radiographs do not have a significant role to play in the diagnosis of mucoceles. If obtained, they demonstrate opacification and expansion of the affected sinus.

CT

The affected sinus is completely opacified, and the margins expanded and usually thinned. Areas of complete bony resorption may be present resulting in bony defect and extension of the 'mass' into adjacent tissues.⁸⁻⁹ Peripheral calcification is sometimes seen.⁸ Following administration of contrast only peripheral enhancement (if any) is seen.

The content of the sinus is variable, depending on the degree of hydration, ranging from near water attenuation to hyper attenuating as secretions become increasingly thick and dehydrated.⁹

MRI

MRI signal intensity is very variable and depends on the proportions of water, mucus and protein

- T1 -water rich content: low signal (most common),protein rich content: high signal
- T2-water rich content: high signal (most common),protein rich content: low signal
- T1 C+ (Gd)-enhancement if present, only occurs at the periphery
- DWI: variable

It should be noted that colonization with fungus could lead to very low signal on both T1 & T2 weighted sequences, mimicking a normal aerated sinus.⁹⁻¹⁰

TREATMENT

Treatment of mucoceles is surgical and the access routes may be either external or endonasal. External approach is made through fronto-ethmoidectomy (Lynch's procedure) or by osteoplastic flaps with or without frontal sinus obliteration and total excision of mucosa. For many years, these techniques were the only surgical alternative to treat fronto-ethmoidal mucoceles. They are aggressive procedures with high morbidity and currently reserved for extreme cases with significant intracranial or orbital extension.¹¹⁻¹² The current tendency is to conduct functional, little invasive and low morbidity procedure with nasosinus endoscopic surgery, with marsupialization and abundant drainage of the lesion, preserving the epithelium. Recent studies demonstrated that mucoceles does not affect the characteristics of the respiratory mucosa and that marsupialization and consequent improvement of local ventilation is possible to reverse epithelial metaplasia into normal respiratory epithelium or at least bring it close to normal.

CONCLUSION

Mucoceles can occur as primary lesions or they may emerge secondary to some other conditions. There are two modes of surgical treatment. The first is the endoscopic marsupialization and creation of a new drainage pathway. The second method is the standard external approach such as Lynch-Howarth external fronto-ethmoidectomy or frontal osteoplastic operation. There is increasing in the literature that endoscopic management of sinus mucoceles is successful, with low morbidity rates and recurrence. Rhinologic surgeons should consider the endoscopic technique as the surgical procedure of choice for management of paranasal sinus mucoceles.

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