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Синдром верхушки глазницы, вызванный доброкачественным мукоцеле задней решетчатой кости, приводящий к необратимой слепоте: описание случая

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Резюме

Мукоцеле – доброкачественное хроническое кистозное поражение, возникающее преимущественно из лобных и решетчатых пазух, в редких случаях из клеток, расположенных сзади. Они могут вызвать компрессию зрительного нерва, синдром верхушки орбиты, компрессионную нейропатию зрительного нерва и слепоту. 71-летняя пациентка обратилась в офтальмологическое отделение скорой помощи с опущением правого века и нечеткостью зрения. При компьютерной томографии выявлено поражение мягких тканей в правой задней решетчатой пазухе, затрагивающее верхушку глазницы. При магнитно-резонансной томографии – объемное образование овальной формы, разрушающее границы кости пазухи. Выставлен диагноз «синдром верхушки правосторонней орбиты, обусловленный мукоцеле задней решетчатой кости». Пациенту проведено лечение преднизолоном, а также функциональная эндоскопическая хирургия носовых пазух. Через год после операции острота зрения пациента составила 0/20, отсутствовали светоощущения, диагностирован афферентный зрачковый дефект правого глаза.
Мукоцеле диагностируется с помощью магнитно-резонансной томографии. Может привести к потере зрения из-за передачи давления на зрительный нерв. Для лечения этого заболевания и предотвращения необратимой потери зрения используются марсупиализация и кортикостероиды. Мукоцеле решетчатой пазухи, хотя и встречается редко, может привести к необратимой слепоте, что требует сотрудничества между рентгенологами, лор-хирургами и офтальмологами для раннего выявления заболевания.
Ключевые слова: слепота, острая потеря зрения, мукоцеле, мукоцеле задней решетчатой кости, синдром верхушки орбиты

■ INTRODUCTION

A mucocele is a cystic lesion that develops in the sinus mucosa. It is benign, long-lasting and lined with epithelial cells. Due to the relatively narrow ostia, Mucoceles usually form in the frontal sinuses (65%) and ethmoid sinuses (25%) [1]. In rare cases, mucoceles

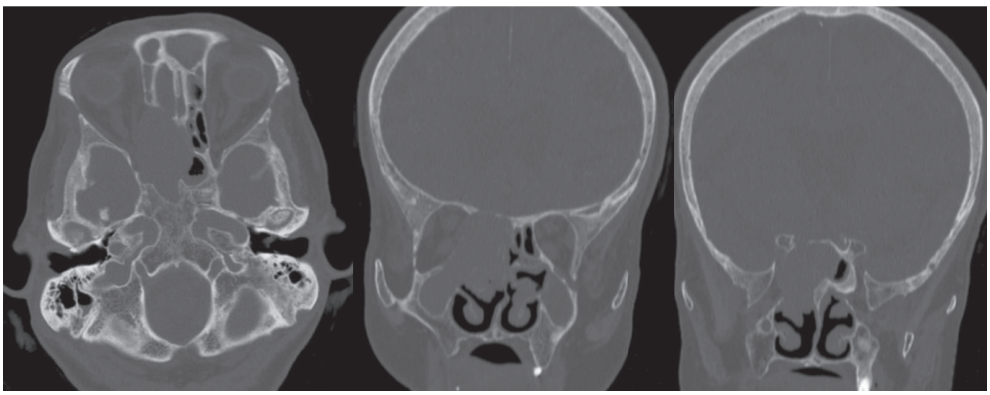
originating from posteriorly located cells can compress the optic nerve and surrounding structures, leading to blindness and compressive optic neuropathy [2].

■ CASE PRESENTATION

A 71-year-old lady presented to the ophthalmology ER complaining of a right-sided droopy eyelid and blurry vision that started 30 days back and progressed to complete vision loss 5 days before her presentation. On examination, VA was 0/20 in the right eye, and 20/20 left eye. Frozen globe right eye, normal EOM left eye. APD right eye, reactive pupil left eye. Decreases sensation of the face over the V2 distribution. An emergent CT brain was requested and reported as "well-defined soft tissue density lesion centered in right posterior ethmoid sinus closely abutting and encroaching orbital apex and extending to the nasal cavity measure overall about 2.1×3.7×3.0 cm (Fig. 1).

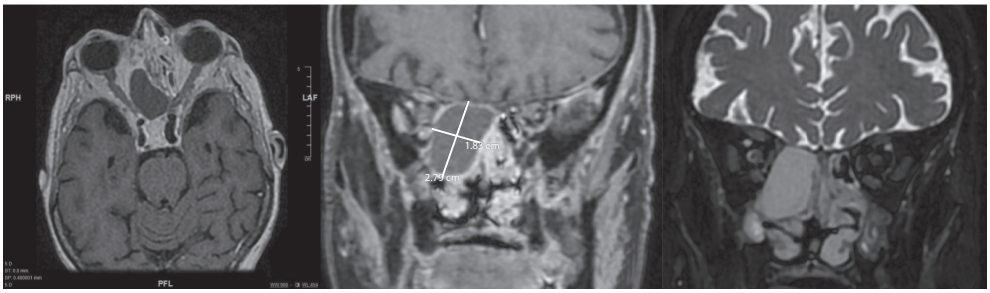
MRI was requested to delineate this pathology further and was reported as "evidence of oval shape mass lesion measuring 2×2.8×3.53 cm centered within the right ethmoid air cells extending posteriorly to the sphenoid sinus eroding the anterior and anterior superior sinus bony boundaries posteriorly while medially it erodes the right lamina paprecea and medial wall of ipsilateral right optic canal abutting the optic nerve but no significant compression or signal alteration of the nerves. The lesion exhibits high T2 and flair and low T1 signal intensity with no enhancement in post-contrast images. Unremarkable cavernous sinuses". Findings probably represent a longstanding ethmoid mucocele with erosion of the adjacent bones as described earlier (Fig. 2).

The patient was diagnosed with right-sided orbital apex syndrome, which was caused by a posterior ethmoid mucocele. The patient was referred to an otorhinolaryngology specialist. After further inquiry, the patient denied any history of sinusitis, nasal obstruction, hyposmia, rhinorrhea, or recent URTI. The patient also reported no history of headache, nausea, vomiting, or any other facility, apart from a positive history of a motor vehicle accident that occurred 40 years ago. Examination revealed no proptosis but a droopy eyelid covering half of the iris. The patient had cranial nerves 2, 3, 4, and



A **B** **C**

Fig. 1. CT SCAN showing posterior ethmoid homogenous mucocele encroaching orbital apex and extending to the nasal cavity axial View (A). Mass Effect causing Thinning and dehiscence of the medial orbital wall and optic nerve as shown in the coronal views (B, C)



A **B** **C**

Fig. 2. MRI Showing oval-shaped mass lesion centered within the right ethmoid air cells extending posteriorly to the sphenoid sinus. The lesion exhibits low T1 signal intensity with no enhancement in post-contrast images (A, B). High T2 and flair signal (C)

V1 palsy with a frozen globe right eye. An intranasal examination with a rigid scope of 2.6 mm 0-degree revealed a right-sided polypoidal Middle meatus with synechia but was otherwise unremarkable. The patient was immediately admitted and started on prednisone 50 mg od. And booked for navigation-guided functional endoscopic sinus surgery for right mucocele marsupialization vs biopsy. During surgery, 30–40 cc of copious mucous discharge was drained from the posterior ethmoid air cell and sent for analysis. The other sinuses were inspected for any other pathology and were unremarkable. Analysis revealed that the mucocele content comprised fibro-collagenous tissues lined by respiratory epithelium with chronic inflammatory cells and granulation tissue. Post-operatively, the patient gradually improved with complete resolution of extraocular movement of the right eye at 3 months post-op, although their vision did not improve. The patient's last follow-up was one year after surgery, during which their examination revealed VA 0/20, no light perception, and APD right eye.

■ DISCUSSION

One uncommon cause of lifelong unilateral vision loss and blindness is ethmoidal sinus mucocele. There is no age bias and both genders are equally impacted [3–5]. Clinical signs of sphenoethmoidal mucoceles typically involve the growth of the sinus and the lesion's extension beyond of the sinus's boundaries. Usually, the lesion extends along the least resistant course [6].

For the diagnosis of mucocele, localizing the lesion, and evaluating potential intraorbital and cerebral extension, magnetic resonance imaging is essential. The degree of variability in MRI signal intensity is contingent upon the tissue's water content, mucus viscosity, and protein composition [7]. On T1-weighted imaging, mucoceles typically exhibit a thin peripheral linear enhancement with a central low-signal intensity, and on T2-weighted and FLAIR images, a high-signal intensity. Acute sinusitis (no bony expansion), mucus retention cyst (does not fill the sinus and does not exhibit bony expansion), paranasal sinus tumors (mostly exhibit diffuse contrast enhancement), antrochoanal polyp (locally protrudes through the ostiomeatal complex), and Aspergillus sp. infection (displays a low signal on both T1- and T2-weighted sequences, mimicking a normal aerated sinus) are among the differential diagnoses.

It is currently not fully understood which specific physiological processes are responsible for causing ocular neuropathy and vision loss that are typically associated with mucocele [8]. There is no fatty or other soft tissue surrounding the optic nerve within the optic canal, which means that the mucocele pressure is directly transferred to the optic nerve. This can compromise the blood supply, leading to the appearance of optic atrophy. Another possible reason for visual abnormalities could be ocular neuritis, which is caused by an inflammatory response. Inflammatory factors, rather than mechanical compression, are often linked to an early onset and rapid progression of visual abnormalities. Gradual emergence of clinical symptoms is typically associated with mechanical compression [9].

The primary objectives of managing mucocele are to decompress the sinus and relieve pressure on the orbit, brain, and optic nerve. This can be achieved through the use of corticosteroids and mucocele marsupialization. Early intervention is crucial in lowering the risk of irreversible vision loss and aiding patients in regaining their vision. Studies have demonstrated that decompression surgery can be postponed for up to three weeks following the initial diagnosis while still achieving a full visual recovery [10, 11]. Wu et al. also found that even with a delay of five weeks, their patient had postoperative visual acuity of 20/20. Nonetheless, in rare cases, despite early surgical decompression and intravenous treatment with steroids and/or antibiotics, there may be no visual improvement [12].

■ CONCLUSION

Though rare, ethmoidal sinus mucoceles pose a serious risk of irreversible blindness. As such, it is imperative to take early surgical intervention to prevent permanent damage. Delaying the treatment may lead to severe and potentially irreversible consequences.

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