

## Interesting Case:

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# Large mucocele with intracranial and intraorbital extension

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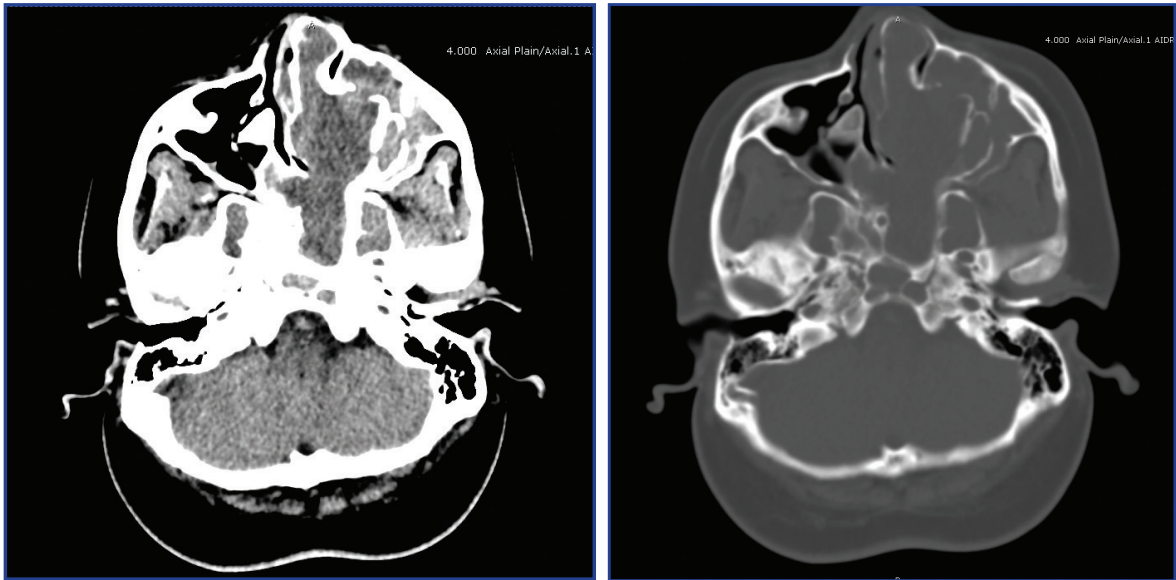
## Case summary

A 26-year-old female had had progressive right eye blindness for 13 years. She did not have any other complaints such as headache, seizure, or weakness. At that time she was admitted to a local hospital and Computed tomography (CT) imaging was performed, showing a mass at left maxillary sinus with extracranial extension. The caring physician advised surgery but her family refused due to financial problem. A year later, she developed complete bilateral eye blindness and the subsequent CT revealed larger size of the mass. She was referred to a tertiary hospital for surgery but she lost follow-up. In this visit, she had foul-smelling discharge per left nostril without fever for 2 weeks followed by seizure and headache for 2 days. She had no underlying disease, no history of seizure and head trauma.

Physical examination revealed that her vital signs were normal; body temperature of 36.7\*c, blood pressure of 100/69 mmHg, pulse rate of 110/ minute, and respiratory rate of 24/ minute. Both pupils were dilated (8 mm/ 8 mm). Light reflexes were diminished in both eyes as well as proptosis with more severity in the left eye. The fundus scope showed bilateral markedly pale discs. There was a mass protruding outside both nostrils. The cranial nerve examination and neurological examinations were normal.

Laboratory investigation for hormonal levels (Free-T4, TSH, HGH cortisol, FSH, LH, prolactin) were all in normal ranges.

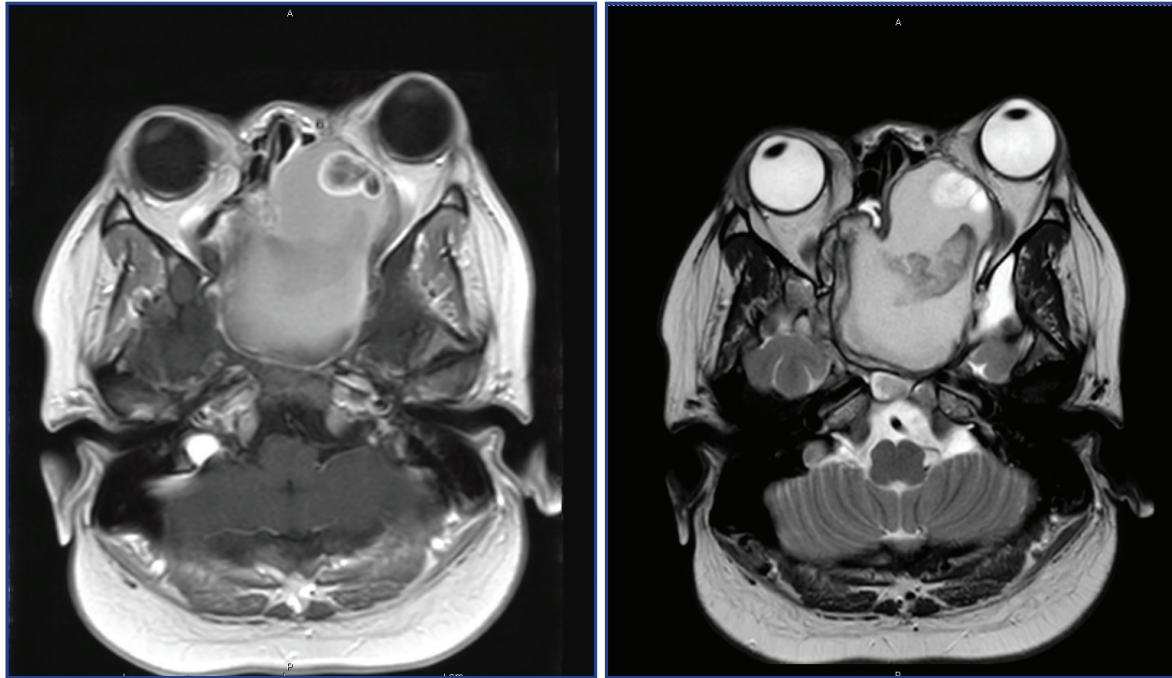
### *Imaging findings*



**Figure 1.** Non-enhanced cranial CT at the level of maxillary sinuses demonstrates large left sinunasal soft tissue with erosion/remodeling and expansion of their bony walls, better demonstrated in bone window (left).



**Figure 2.** Non-enhanced cranial CT in axial (right), sagittal (middle) and coronal (left) demonstrates a large left sinunasal soft tissue mass with intracranial extension and mass effects causing lateralization of the globe, narrowing retrobulbar space and proptosis.



**Figure 3.** (A) T1-weighted spin-echo magnetic resonance imaging after intravenous administration of gadolinium demonstrates a non-enhancing mass with hypointense rim in left sinunasal region with intracranial extension, displacing adjacent intracranial vessels. (B) The mass appears hyperintense on T2-weighted imaging.

## Diagnosis

A large mucocele with intracranial and intraorbital extension

## Treatment

The patient underwent bilateral tumor resection via an endoscopic sinus surgery (ESS). Intraoperatively, there was an expansile bony tumor with mucoid and soft tissue components occluding bilateral nasal cavities and extending into sphenoidal areas. The mass was removed and pathologically proven to be mucocele.

In spite of complete tumor removal, however, bilateral blindness was still present. The patient was discharged after 11 days of hospitalization. A 6-month follow-up MRI was planned to exclude any recurrence.

## Discussion

Mucoceleles are benign mucus-containing, epithelial lined cystic cavities, developing in paranasal sinuses as a result of chronic obstruction of the outlet ostia that connects the sinus to the nasal cavity.[1,2] They are characterized by a gradual expansion through a continuous mucous-secreting process, resulting in a compression and displacement of the surrounding soft tissues and erosion of the bone walls.[3]

Mucocele could be undiagnosed until symptoms due to compression of surrounding structures arise. The symptoms depend on area of extension: orbital extension (pain, proptosis, loss of vision, ocular motility disturbances, tearing), cranial extension (meningitis, headaches, epidural abscess, subdural empyema, brain abscess, cranial nerve palsies) and nasal expansion (nasal blockage and loss of sense of smell).

Plain skull radiography showing enlarged sinus walls and signs of bone erosion can be suspicious of a mucocele. CT is the most useful investigation in identifying a mucocele, that reveals a non-enhancing airless mass inside the sinus with mucous density.[4] MRI appears iso-hyperintense on T2-weighted imaging. The T1-weighted imaging can vary based on the proportion of mucus, water, and protein contents.[5-7] Showing no contrast enhancement helps differentiate a mucocele from other naso-sinusal tumors. CT scans are much better at delineating the extent of the lesion and its relations to the surrounding structures as compared to an MRI.[8]

Surgery is the required treatment for complete mucocele eradication. This procedure could be achieved with transnasal endoscopic surgery, which has recently become a preferred approach over the more invasive external craniotomy with craniofacial surgery. However, endoscopic transnasal approach is not recommended in cases with intracranial extension due to higher risk of intracavitary residual and post-operative recurrences.[9,10] The prognosis of visual function depends upon the period of visual impairment.

Devars du Mayne et al reported 23.5% recurrence rate with an average onset of recurrence being at 4 years, more frequently in patients with persistent inflammatory sinus diseases. They advised CT imaging every 2 years for at least 4 to 5 years after surgery. Despite a low recurrence rate, a long follow up is generally recommended.[11,12]

Although most cases in prior reports were largely with/without intracranial extension, few cases were presented with intraorbital extension. One large fronto-orbital mucocele causing proptosis of the right eye and diplopia was reported by Mohan S[13], but the visual acuity was still intact. The other case by Severino R et al[14], reported a fronto-orbital mucocele with 4-week history of left exophthalmos and sudden proptosis. Both cases had no visual complication after treatment. In this case, we would like to show the rare long-standing large mucocele with serious extension causing poor prognosis in treatment.

## References

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