Giant Frontal Mucocele with Extensive Bone Destruction: Case Illustration

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Abstract: Mucoceles are epithelial - lined cavities in the paranasal sinuses that contain mucus. They develop due to scar tissue and sinus ostium obstruction, either from chronic sinusitis, trauma, or surgery. Frontal sinus mucoceles can present with many different symptoms including eye disorders. A case of giant frontal mucocele with extensive bone destruction that reappears after four years is reported. Surgery to remove the mucocele and reconstruction of the skull would be performed, mucocele can cause bone destruction because these mucoceles have a tendency to develop by eroding the surrounding bone wall replacing and destroying the structure with pressure and bone resorption.

Keywords: Frontal mucocele, bone destruction, surgery

1. Introduction

Mucocele is a rare benign intracranial pathology arising from the slowly growing paranasal sinuses that develops after obstruction of the sinus ostium. The frontal and ethmoid paranasal sinuses are the sinuses most commonly affected (70% - 90%), and young adults (20 - 40 years of age) are the most commonly affected age group, with a slight predilection in male patients but no significant difference by gender.

Frontal sinuses mucoceles can have various sizes and configurations. Intraorbital degree involvement is not used to differentiate between the different types of lesions.

These are the following classification of mucoceles sinus:

Type 1: Limited to frontal sinus (with or without orbital extension).
Type 2: Frontoethmoid mucocele (with or without orbital extension).
Type 3: Erosion of the posterior sinus wall.
   A: Minimal or without intracranial extension.
   B: Major intracranial extension.
Type 4: Erosion of the anterior wall.
Type 5: Erosion of both anterior and posterior wall.
   A: Minimal or without intracranial extension.
   B: Major intracranial extension.

The diagnosis of mucocele is based on clinical examination carried out with the help of computed tomography (CT) and magnetic resonance imaging (MRI). The differential diagnosis includes paranasal sinus carcinoma, Aspergillus infection, chronic infection or inverting papilloma.

2. Case Illustration

A 63-year-old male patient, came with the chief complaint of a lump on the forehead. This lump on the forehead has been felt since 3 years ago. The patient feels that it is getting bigger within time. The patient also feels that over time this lump is getting bigger, the patient also feels a headache and the vision in both eyes is getting blurry. Headache is felt enough to interfere with the patient's daily activities. On eye examination found ODS KSI with visual acuity OD 20/150 and OS 20/100. The patient has had surgery 7 years ago with the same complaint. The patient has a history of diabetes mellitus uncontrolled.

MRI examination was found T1 - weighted MRI with contrast shows an increase in signal intensity around the area of the swollen lesion and shows an increase in the periphery, T2 - WI also shows an increase of signal intensity. The results of MRI with contrast found a cystic mass measuring 5.48 x 6.77 x 6.65 cm, originating from the right and left frontal sinuses that extended to the right and left extraaxial regions of the frontal region. Possibly accompanied by destruction of the posterior wall of the right and left frontal sinuses, the dominant left side pushing the frontal lobe posteriorly, extending to the right orbit to push the right eyeball to the lateral side. Then accompanied by mucosal thickening of the right and left ethmoidal, sphenoidal and maxillary sinuses. (Figure 1. a, b, and c)

Figure 1: Preoperative image head MRI (a, b and c) Axial, coronal and sagittal images (T1 - weighted with contrast) showing cystic mass in frontal region with bone destruction pushing frontal lobe and right eyeball.

Based on the symptoms and examination leading to a frontal mucocele that may reappear, then we performed surgery for tumor resection and skull reconstruction using bone cement. Opening from bifrontal incision behind hairline, skin dissection found mass with soft consistency surrounded by thick wall. After explored until edge around the mass, found bone destruction. Incision at the mass came out brown viscous fluid. The cyst wall that attached to the bone and dura mater was removed carefully and protect superior...
sagittal sinus with hemostate agent. The aspirated fluid and cyst wall tissue was sent for pathological examination.

Bone destruction found in frontal bone left and right side anterior posterior wall extended to the right orbital. The frontal sinus closed with muscle graft, and prepared bone graft from bone cement to close the skull defect. After surgery the patient's condition improved, the headache was getting better, the condition of the surgical wound was good, there were no signs of infection and cerebrospinal fluid leakage.

Cytological examination found smear consisted of a dense distribution of cyst macrophages, including degenerating cells. Histopathological examination shows the cyst wall tissues lined by flattened, pseudostratified, ciliated columnar epithelium with chronic inflammatory cells (Figure 2a and b). Based on the clinical, radiological, and histological findings of this patient, it can be confirmed that it is a frontal sinus mucocele.

![Image of histopathological photos](image)

**Figure 2:** Histopathological photos. (a) Sections of cyst wall consisting of surface epithelium and fibrous connective tissue (H&E stain, 200x) and (b) The inner surface of the cyst wall contains macrophages, lymphoplasmacytic, multinucleated giant cells (H&E stain, 200x).

3. **Discussion**

Mucoceles are most common in the frontal and ethmoid sinuses, rarely in the sphenoid sinus and rarely in the maxillary sinus. Proposis is the most common sign of a frontal mucocele. Other clinical features include a mass in the medial upper quadrant of the orbit, pain, vertical diplopia, limited upward vision, bifrontal headache and increased tearing. In this case from clinical examination there was big lump in forehead and difficulty to open the right eyelid also feel headache, blurry vision that can occur due to the spread of the mucocele to intracranial and from medial to lateral orbital. Mucoceles are expansive pseudocystic formations that secrete mucus and are capable of expansion based on the dynamic process of bone resorption and new bone formation. They result from sinus ostium obstruction and are often associated with previous conditions such as chronic sinusitis, trauma, surgery or widespread lesions. With continued mucus secretion and accumulation, the increased pressure causes sinus bony atrophy or erosion, allowing mucoceles to develop in less resistant pathways. It affects into the orbit, adjacent sinuses, nasal cavity, intracranially or through the skin. Occasionally, an aggressive mucocele will cause destruction of bone, especially the orbital wall and can then simulate a malignant neoplasm. Imaging result in this case shows bone destruction in frontal region left and right side extended to right orbita, it could be due to the unusually aggressive expansion of the mucocele, MRI examination was carried out because MRI was considered more useful in differentiating mucoceles from neoplasms through contrast enhancement. [14]

MRI is superior to CT in distinguishing mucoceles from soft tissue and in relation to adjacent soft tissues such as the brain and orbit. The classic radiographic appearance of a mucocele is generalized thinning and expansion of the sinus walls and there may also be evidence of sinus disease and bone erosion.

Mucoceles usually appear homogeneous and airless. MRI showed variable signal intensity on both T1 and T2 - weighted images, depending on the state of hydration, protein content and viscosity of the mucocele contents. [8] Contrast - enhanced MRI is useful in differentiating mucoceles from sinonasal tumors. [7] Mucoceles typically reveal thin peripheral linear enhancement with low central signal intensity on T1 - weighted images; and sinonasal tumors show a diffuse increase. In this patient, it was found that there were swelling lesions and peripheral enhancement in the T1 and T2 sections, as well as the process of destruction of the bone wall.

Diagnosis requires correlation between clinical, radiographic, and pathological findings, as histopathological
findings alone are nonspecific. Histopathological examination shows the cyst wall tissues lined by flattened, pseudostratified, ciliated columnar epithelium with chronic inflammatory cells. There is a typical histopathological findings in this patient which supports the diagnosis of frontal mucocele.

Other known complications of frontal mucoceles include erosion of the anterior wall, resulting in a gently fluctuating mass beneath the periostium of the frontal bone. Posterior wall erosion can lead to complications such as epidural abscess, meningeitis, subdural empyema and brain abscess. [4] Rarely, cranial nerve palsies may also occur. Intraoperatively there is extensive bone destruction on the posterior wall of the right and left frontal bone, mucocele extend intracranial and pushing the frontal lobe posteriorly, and classified as type 5B. [12]

Definitive treatment of mucoceles is primarily surgical, but frontal sinus mucoceles that extend into the orbit and anterior cranial fossa can, in certain cases, be difficult to manage therapeutically and can lead to lethal complications. [6] Traditionally, frontal mucoceles are treated with an external open oblitative procedure. Alternatively, functional endoscopic sinus surgery can be used to evacuate the mucocele, but this procedure becomes difficult if there is intracranial expansion. In this case there was extradural intracranial expansion, which made the external approach more suitable for frontal mucoceles, especially distally and in cases of recurrence and with bone destruction that need skull reconstruction.

The external open approach has advantages: being a direct approach, by allowing exposure of the entire sinus, providing complete removal of the sinus to prevent recurrence of sinus disease, and preventing blind curettage of any open dura mater. The prognosis for frontal sinus mucoceles is good with a chance of cure, and a low incidence of recurrence. [13] Although, to control relapse, long-term follow-up is recommended.

4. Conclusion

Giant Mucoceles in the frontal sinus area are rare and usually take a long time to enlarge, which if enlarged will cause extensive disruption of the orbit and anterior cranial fossa. Frontal mucoceles can be clearly diagnosed and cured, but early diagnosis and prompt treatment are essential. Surgery to remove the mucocele and skull reconstruction with bone cement can be chosen and long term follow up is recommended.

References