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Case Report

Longstanding giant frontoethmoidal mucocele with orbital and intracranial extension: challenges of surgical approach in a developing nation

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ABSTRACT

Mucoceles of the paranasal sinuses are expansile, benign mucus filled epithelial lined cystic-like lesion resulting from chronic obstruction to sinus ostia. They erode bony walls by pressure and bony resorption with the frontal and ethmoid sinuses predominately affected. The expansile lesion can involve surrounding structures like the orbit, intracranium or nasopharynx. Diagnosis is based on history, examination and radiological assessment with Computerized tomography scan being the imaging of choice. Surgery is the mainstay of treatment and would require collaboration with the neurosurgeons or the ophthalmologist when it extends to the brain or orbit respectively. It can be done via an external approach, endoscopic approach or a combination of both. If left without intervention, it may cause morbidity and potential mortality due to its proximity to the brain and orbit. We report a rare case in our setting of a 65-year-old male farmer with a painless huge left sided cystic fronto-orbital swelling of 8 years that mimicked a benign orbital tumor. The mass was non-tender and not pulsatile measuring about 20×18 cm with normal visual acuity. Computed tomography (CT) scans of the brain and paranasal sinus revealed a large well-defined non-contrast enhancing homogenous lesion involving the left fronto-ethmoidal sinuses with orbital and intracranial extension as well as inferolateral displacement of the left eye globe. He was reviewed by the ENT surgeons, neurosurgeons and ophthalmologists and subsequently had frontoethmoidectomy via an external approach.

Keywords: Rare giant mucocele, Frontoethmoidal, Paranasal sinuses, Nigeria

INTRODUCTION

Mucoceles of the paranasal sinuses is a rare condition characterized by expansile, benign mucus filled epithelial lined cystic-like lesion resulting from chronic obstruction to sinus ostia.[1] Obstruction of the ostia may occur as a result of inspissated secretions from infections or allergy, trauma, metastatic or malignant tumors or benign lesions affecting the sinuses.[2] It erodes bony walls by pressure and bony resorption. If left unchecked, it may result in lethal complications due to its proximity to the brain and orbit.[2] Mucocoeles are common in the third and fifth decade of life and has equal sex predilection.[3] The sinuses commonly affected are frontal and ethmoid sinuses and rarely the maxillary and sphenoid sinuses. Skull base erosion may occur in 10- 20% of cases.[4]

Treatment of mucocele of the paranasal sinuses is mainly surgical and could be challenging.[5] This could be excised via external approach, endoscopic approach or a combination of both.[6] The choice of approach depends on the extent of the lesion, expertise of the surgeon and available resources. When there is invasion of the orbit or anterior cranial fossa, a more radical approach may be

preferred and this is usually done in collaboration with the ophthalmologist or the neurosurgeons as the case may be.[7,8] In this article, we report a rare case in our setting, of a longstanding giant frontoethmoidal mucocoele presenting with displacement of the left eye globe.

CASE REPORT

A 65-years old male farmer in Northwest Nigeria presented with a painless large left sided frontal swelling of 8 years with slow progressive enlargement that mimicked the behavior of many benign orbital tumors. No significant nasal symptoms or features of frontal lobe syndrome. Examination revealed a large cystic mass involving the left fronto-orbital region with severe inferior lateral displacement of the left eye globe and normal visual acuity. The mass was non-tender and not pulsatile but measures about 20×18 cm with right eye lens opacity (Figure 1).

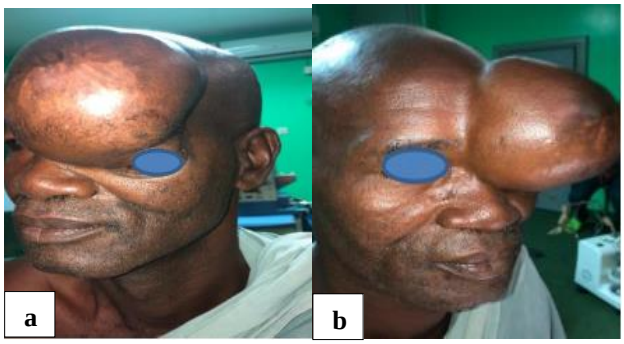

Figure 1: Frontolateral view of giant frontoethmoidal mucocoele (a) left and (b) right.

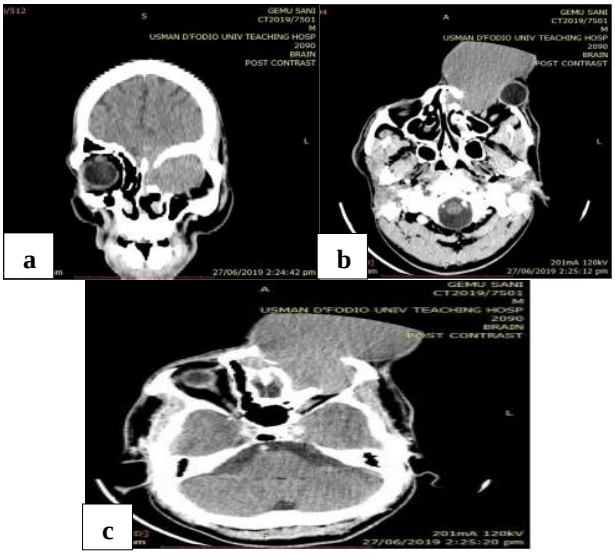

Figure 2: CT scan of the paranasal sinus and brain (a) coronal view of isodense lesion in orbit, frontal sinus and intracranial extension, (b) axial view with isodense lesion displacing the globe, and (c) isodense lesion involving the left ethmoid and globe on axial view.

Computed tomography (CT) scans of the brain and paranasal sinus revealed a large well-defined non-contrast enhancing homogenous lesion involving the left fronto ethmoidal sinuses with orbital and intracranial extension as well as inferolateral displacement of the left eye globe (Figure 2). Informed consent was given by the patient and he subsequently had frontoethmoidectomy via an external approach. Histopathological diagnosis was in keeping with frontoethmoidal mucocele. The patient was asymptomatic at 6-month follow-up (Figure 3).

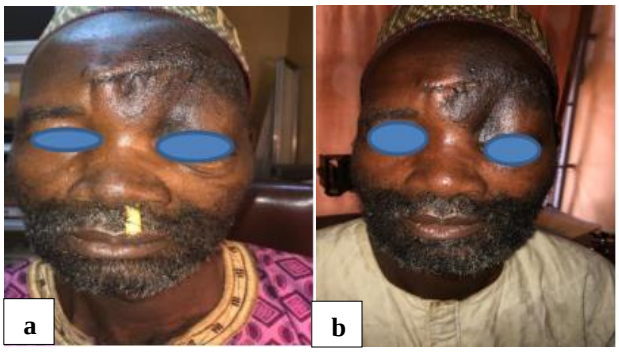

Figure 3: Photograph of patient at (a) one week postoperative showing frontonasal catheter in situ, and (b) six months follow up visit.

DISCUSSION

Mucocoeles are benign mucus filled epithelial lined cystic-like lesion resulting from chronic obstruction to sinus ostia. These cysts like lesions are lined with columnar epithelium or pseudo stratified epithelium containing goblet cells.[1,3] Continuous expansion of the cysts occurs as the goblet cells keep secreting mucus creating pressure effect on the surrounding bony structures causing bony erosion and bone remodeling. In addition, the presence of inflammatory mediators like interleukins and tumor necrosis factor also contribute to the expansion capability of mucocoeles while prostaglandins and collagenases secreted by fibroblasts increases bone resorption.[9,10] The expansile lesion may progress to involve the orbit, intracranium or nasopharynx.[11,12] The frontal and ethmoid sinuses are predominantly affected and less commonly the maxillary and sphenoid sinuses.[1,6,8,12-14] Rollet in 1896 first used the word mucocele while Onodi was the first to describe the histology of Mucocele in 1901.[1,15]

Mucoceles of the paranasal sinuses affect both sexes in equal proportion, occur frequently in the third to fourth decades of life and rarely in children.[3,12,14,15] Common predisposing factors related to developing paranasal sinus mucoceles are metastatic or malignant tumors or benign lesions affecting the sinuses, previous history of sinusitis, history of nasal or sinus surgery, allergy, and trauma.[2] All these lead to obstruction of the sinus ostium or entrapment of the sinus mucosa and it may take several years before patient develops symptoms. The symptoms depend on the sinuses affected, however the commonest are facial swelling and asymmetry, headache, proptosis,

exophthalmos, epiphora, impaired vision, and diplopia.[2,3,14] Meningitis, convulsions or cerebrospinal fistula may occur when the cyst erodes the posterior table of the frontal sinus.[16] They rarely present with nasal symptoms.[17] Mucoceles are termed giant when there are intraorbital and intracranial extensions or are greater than 5 cm in the reported dimension.[18,19]

Diagnosis of mucocele of the paranasal sinus is based on history, examination and radiological findings.[2] Plain X-rays of the sinuses though rarely done, may show soft tissue density mass obliterating the affected sinus, expansion, thinning and erosion of the sinus.[12] CT scan of the paranasal sinuses and brain is considered the imaging of choice for the diagnosis of mucoceles.[3] It clearly delineates the expanded airless sinus filled with homogeneous material. The affected sinus walls may be thickened due to sclerosis or thinned and eroded at places. It also clearly delineates skull base erosions. Mucocele appear as non-enhancing homogenous isodense lesions on CT as seen in this patient's CT scan while infected mucoceles (mucopyoceles) are contrast enhancing.[20] Magnetic resonance imaging (MRI) is helpful in cases of intracranial extension and to differentiate mucoceles from malignancy.[12,21] Mucoceles are characteristically fairly bright on T1-weighted images compared with the brain, and isohyperintense on T2-weighted images. In the index case patient couldn't afford MRI due to financial constraints and non- availability in our resource poor setting.

Treatment of mucoceles of the paranasal sinuses is mainly surgical and could be challenging.[22] It could be excised via external approach, endoscopic approach or a combination of both. The choice of approach depends on the site and extent of the lesion, expertise of the surgeon and available resources.[6,7] In our setting, despite the availability of skilled surgeons, an external approach was preferred as wide exposure would be needed in order to completely excise the cyst hence, preventing recurrence in this patient. Moreover, most patients cannot afford a second surgery here due to high cost of surgery hence a radical approach at the first surgery is advocated. In addition, when there is invasion of the orbit or anterior cranial fossa, a more radical approach may be preferred and this is usually done in collaboration with the ophthalmologist or the neurosurgeons as the case may be.

The goal of treatment is to ensure complete excision of the cysts with minimal morbidity as well as preventing recurrence and an overall satisfactory cosmetic outcome. In the case under review, the breach of the posterior table of the frontal sinus and orbital involvement in addition to the above-mentioned factors necessitated an external approach.

CONCLUSION

Giant longstanding frontoethmoidal mucocele is an unusual pathology in our center and is the first reported case in this environment to the best of our knowledge. Accurate, early diagnosis with appropriate surgical intervention based on availability of skills, funds and resources as well as multidisciplinary collaboration is advocated.

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