

Recurrent bilateral fronto-ethmoidal mucocoele with intracranial extension: A case report

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Summary

Mucocoeles of the paranasal sinuses are relatively uncommon and rarely bilateral, the frontal and ethmoidal sinuses being most commonly affected. Rarely, fronto-ethmoidal mucocoeles expand and extend into the anterior cranial fossa producing mass effect. We report a 66-year-old woman, who presented with a 20-year history of recurrent bilateral progressive proptosis and supra orbital swellings with blindness in the left eye. Ultrasound scan of the left eye showed a large cystic mass in the orbit due to extension of ipsilateral frontal mucocoele. Computed tomographic brain scan showed large bilateral fronto-ethmoidal mucocoeles, erosion of the right orbital roof, right posterior and left anterior sinus walls, and extension of the right frontal sinus into the anterior cranial fossa with mass effect. The patient had a bicoronal craniotomy and bilateral fronto-ethmoidectomy with direct drainage of independent right mucocoele and left mucopyocoele. This case illustrates complications that may result from chronicity and treatment failure in patients with sinus mucocoeles.

Key-words: *Mucocoeles, Recurrent, Bilateral, Fronto-ethmoidal*

Résumé

Sinus paranasaux de la mucocèle sont relativement peu commun et rarement bilatéral, le sinus frontal et ethmoïde sont le plus souvent concernés.

Les mucocèles fronto-ethmoïde rarement étendent et développe dans le fossa cranier antérieur ce qui produit un effet de masse. Nous rapportons le cas d'une femme âgée de 66 ans, qui s'est présentée avec une histoire de 20 ans de proptose progressive bilatérale récurrente et tuméfaction supra orbitale avec la cécité dans l'oeil gauche. L'échographie de l'oeil gauche avait indiqué un grand mass cystique dans l'orbite attribuable à l'extension de la mucocèle frontale ipsilatérale. Tomographique Examen au scanner du cerveau assisté par l'ordinateur avait indiqué une grande érosion de la mucocèle fronto-ethmoïdale de la voute orbitale du côté droite, postérieur du côté droite, et les parois sinus antérieur du côté gauche et extension du sinus frontal du côté droite dans le fossa cranier antérieur avec un effet de masse. Le patient avait eu une craniotomie bicoronale et un fronto-ethmoïdectomie bilatérale avec drainage directe de la mucocèle du côté droite indépendant et mucopyocèle du côté gauche. Ce cas présente une illustration des complications qui pourront être attribuable au échec du traitement et à la chronicité chez des patients avec le sinus mucocèles.

Introduction

Mucocoeles of the paranasal sinuses are relatively

uncommon and rarely bilateral¹⁻⁴; the frontal and ethmoidal sinuses being most commonly affected.¹⁻³ Characteristically, they evolve very slowly, but sometimes rapidly as acute inflammatory processes to become a mucopyocoele especially when secondarily infected.^{1,2,4,5} Most commonly, expansion of the sinus by a mucocoele or a mucopyocoele follows the route of least resistance into the orbit, causing lateral (2-13mm) or inferior (1-10mm) displacement in 55% and 59% of cases respectively, and resulting in limited ocular mobility in the upward gaze.¹ Majority (91%) of the patients with fronto-ethmoidal mucocoeles exhibit some degree of dynamic or adynamic proptosis (1-17mm) on account of which they are initially seen by the ophthalmologist.¹⁻⁴ Rarely, fronto-ethmoidal mucocoeles expand and extend into the anterior cranial fossa producing mass effect.⁵⁻⁸

To the best of our knowledge, this is the first case reported in Nigeria, of recurrent bilateral fronto-ethmoidal expansion with independent mucocoele and mucopyocoele and intracranial extension. This case illustrates complications that may result from chronicity and treatment failure in patients with sinus mucocoeles.

Case report

A 66-year-old woman, a farmer presented with a 20-year history of recurrent bilateral progressive proptosis and supra-orbital swellings. There was associated recurrent nasal blockage, nasal discharge, postnasal drip, excessive sneezing, bilateral anosmia and bifrontal headache. She had had a left external fronto-ethmoidectomy at another University Teaching Hospital 17 years before current presentation; and a repeat operation 3 years later at the same hospital. Proptosis in the left eye recurred a few weeks after each surgery and was progressive in the subsequent 14 years. One year prior to presentation she had a rapidly progressive proptosis, which eventually led to blindness in the left eye. There were no



Fig. 1 Preoperative clinical photograph of a 66 year old woman with recurrent bilateral fronto-ethmoidal mucocoele. Note bilateral proptosis and supra-orbital swellings.

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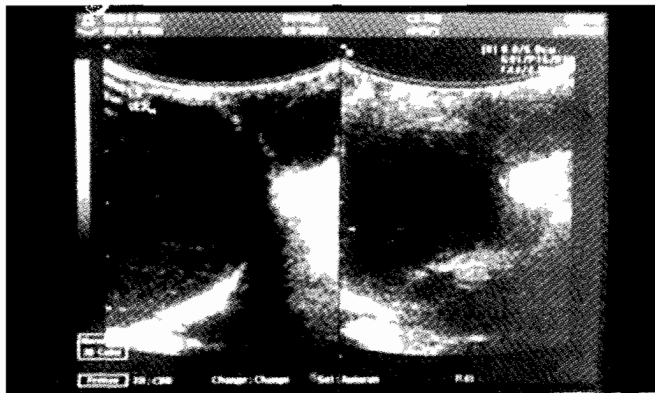


Fig. 2 Vertical and horizontal ultrasound scans of the left eye showing a large cystic mass with internal echoes in the superior aspect of the orbit due to fronto-ethmoidal mucocoele. The globe is seen displaced inferiorly in the vertical scan on the left.

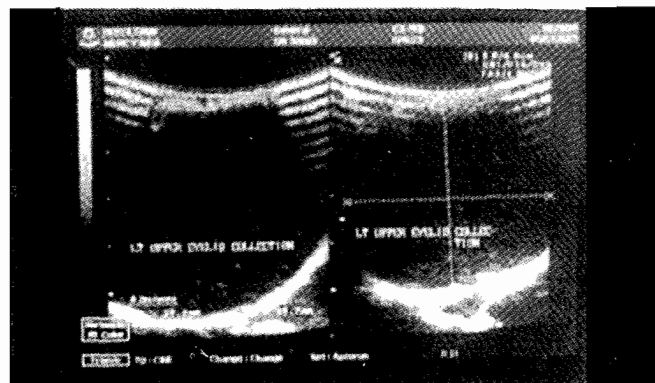


Fig. 3 Vertical and horizontal ultrasound scans of the left orbit showing a large cystic mass, which measures 39mm in diameter due to fronto-ethmoidal mucocoele.

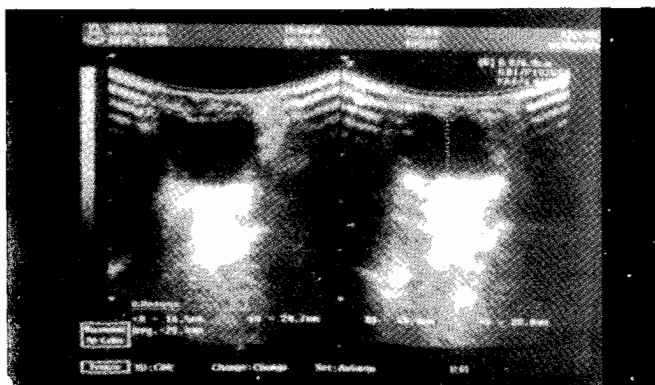


Fig. 4 Reduced antero-posterior diameter (14mm) of the left globe in the horizontal scan due to compression from fronto-ethmoidal mucocoele with orbital extension.

significant aural or throat symptoms and no significant weight loss.

Examination revealed bilateral non-axial proptosis (right = 23mm, left = 21mm), supraorbital swellings, phthisis bulbi and right exotropia. (Fig 1) Visual acuity was $\frac{6}{36}$ in the right eye and no light perception in the left. The left supra orbital skin, covering an 8x6x2cm supraorbital swelling was hyperpigmented with a transverse healed scar on its summit. In the right eye, the conjunctiva was normal, and the cornea was clear, but the anterior chamber was shallow with dilated

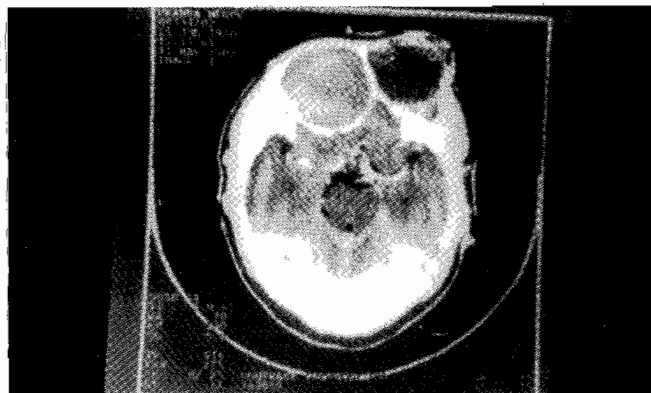


Fig. 5 Computed tomographic brain scan showing large bilateral fronto-ethmoidal mucocoeles with extension of right frontal mucocoele into the anterior cranial fossa and erosion of its posterior sinus wall. The anterior wall of the left frontal sinus is also eroded.



Fig. 6 Immediate postoperative clinical photograph showing sutured coronal scalp incision.

pupil, and a reactive pink disc (CD 0.3).

Attenuated vessels with widespread hard exudates suggestive of grade III hypertensive retinopathy were seen on dilated funduscopy in both eyes. The blood pressure, which was $\frac{160}{100}$ mmHg and associated displaced apex beat, cardiomegaly on chest radiograph, and an anterior hemiblock with non-specific ST-T changes demonstrated on ECG were suggestive of hypertensive heart disease. Aside from engorged nasal turbinates no other significant nasal, aural and throat signs were observed. There were no palpable cervical lymph nodes. Neurological assessment revealed multiple cranial nerve deficits of right I-IV and left I-II nerves without pyramidal signs. A clinical diagnosis of bilateral proptosis secondary to fronto-ethmoidal mucocoele in a hypertensive heart disease patient was made.

Haematological and biochemical profiles were within normal limits. Right orbito-ocular ultrasound revealed an increase in size of the orbital soft tissue space but no definite focal lesion was demonstrated in the retrobulbar soft tissue. Left orbito-ocular ultrasound (Figs 2-4) showed a large cystic mass, 3.9cm in diameter, superomedial to the globe and a partial vitreous detachment. Computed tomographic brain scan (Fig.5) showed large bilateral fronto-ethmoidal mucocoeles, erosion of the right orbital roof and posterior sinus wall, erosion of left anterior sinus wall and extension of the right frontal mucocoele into the anterior cranial fossa with mass effect

The patient had a bicoronal craniotomy and bilateral fronto-ethmoidectomy with direct drainage of mucocoeles. An expanded right frontal sinus compressed the ipsilateral frontal lobe. 70mls of glistening yellowish brown mucocoele was drained from this sinus. Its floor and posterior wall were reduced to a membrane like fibrinous shelf separating the cavity from the right frontal lobe. The smaller left sinus, with an eroded anterior wall but intact posterior wall and floor, contained 50mls of mucopurulent exudate. The inter-frontal sinus septum was however intact suggesting independence of the two sinus cavities. After thorough debridement, the bone defects were covered with methyl metacrylate, the frontal bones were replaced and the scalp was closed (Fig. 6). The pathological diagnosis of the curetted sinus linings was chronic inflammatory tissue and bacteriological study of sinus aspirates revealed no growth.

Discussion

Fronto-ethmoidal mucocoele is relatively rare and will cause visual impairment and blindness when management is belated or ineffective as in the case presented.^{1,5} The insidious onset and absence of pain in most mucocoeles of the paranasal sinuses particularly those of the fronto-ethmoidal region often lure those affected into a state of careless optimism.²

Fronto-ethmoidal mucocoeles are rarely bilateral; reported incidences vary from 0 to 4%.^{1,4} The fact that the left side in our patient was a pyocoele and the right a mucocoele confirms the finding that the inter-sinus septum was intact. It also shows that one is not an extension of the other but they probably developed independently of each other. The pathogenesis of each sinus disease may be similar, though independent and completely separate anatomically.

The formation of a mucocoele has traditionally been attributed to a combination of the obstruction and inflammation of affected sinus.¹ Previous surgery or trauma may contribute to the obstruction, but a significant proportion may not have such contributory factors.^{1,4,5,9} Development of the mucocoele would appear to depend upon the degree and duration of obstruction, the absence of alternative drainage routes and a process of bone resorption and expansion possibly initiated by infection.^{1,4} Histological studies confirm the presence of bone remodeling at the interface between mucocoele and sinus wall.^{1,10} Recent studies on bone-resorbing factors confirm the presence of prostaglandin E₂(PGE₂), collagenase, and cytokines such as interleukin-1 and tumour necrosis factor (TNF) compared with normal controls and chronically inflamed mucosa.¹

Intracranial extension, though rare, has been reported in fronto-ethmoidal and sphenoidal mucocoeles.^{1,7,8,11} At operation, Martinson², found small erosions in the posterior wall of about 60% of all of the frontal sinuses, but large erosions of about 2cms or more in diameter were found in 30% of the cases. Other reported complications of fronto-ethmoidal mucocoele include intracranial abscess, meningitis, cavernous sinus thrombosis, osteomyelitis, destroyed eye and even death.^{2,5,11} Martinson in Nigeria² reported two deaths in his series of 98, both had cerebral abscesses, one of which had ruptured into the lateral ventricle causing early

postoperative death.²

The principal presenting complaint in patients with fronto-ethmoidal mucocoele is proptosis, present in over 80%, with varying degrees of downward and lateral displacement of the globe.¹² Other ophthalmic morbidity occurs only after significant orbital extension leading to compressive optic neuropathy, or even compression of the chiasma and concomitant visual loss or bitemporal hemianopia.¹² Prompt and accurate clinical and neuroradiological diagnosis of fronto-ethmoidal sinus mucocoeles is essential to fostering early effective surgical therapy and prevention of permanent visual loss.^{4,9,12} About 5% of Martinson's series had permanent loss of vision.² The degree of improvement in visual acuity following surgical intervention depends on the patient's visual acuity before the procedure, and the time between onset of disease and surgery.¹² When monocular or binocular vision has been reduced to light perception before surgery, prognosis for visual recovery is poor and if surgical intervention is further delayed the chance of irreversible or permanent blindness increases.^{4,9,12} Because of this, early and correct diagnosis and early presentation to the Otorhinolaryngologist become pertinent.⁴ In this country, however, the patient often consults traditional healers and quacks before ever getting to a specialist hospital.^{2,13,14} This adds to delay in presentation and treatment as it occurred in our patient for 14 years after the second operative intervention.

Until recently, the majority of fronto-ethmoidal mucocoeles were treated by external fronto-ethmoidectomy employing a Lynch Howarth's approach.¹⁵ However, functional endoscopic surgery enhances the chance for excellent result in the treatment of most sinus mucocoeles.⁵ Open surgery remains a valid procedure in frontal mucocoeles with orbital and or cranial extension or osteomyelitis and in developing world where only few centers have facilities for endoscopic nasal surgery.⁶ In our patient, a bilateral Lynch Howarth's incision was avoided by using a bicoronal approach which allowed access to both mucocoeles, allowed room for sequestrectomy of osteomyelitic bone and gave opportunity for cranioplasty with methylmetacrylate. This approach is preferred in patients with bilateral fronto-ethmoidal mucocoele, with intracranial extension or recurrence after surgical treatment, or when cranioplasty with methylmetacrylate is required.^{2,6,11,16,17}

Recurrence could occur following satisfactory fronto-ethmoidectomy as observed in this case; the reported rates being about 4% or more within 1-30 years of initial operation.^{1,2,6,18} It is usually due to stenosis of the enlarged frontonasal opening and this requires revision.² The open approach to surgical management attempts to reduce this high rate of recurrence.

Conclusion

This case illustrates complications that may result from chronicity and treatment failure in patients with fronto-ethmoidal sinus mucocoeles.

Acknowledgement

We acknowledge the initial neuro-ophthalmic review of

this patient by Dr A.I. Uyanne at the hospital's Eye Clinic.

References

1. Lund VJ. The orbit. In Kerr AG; Mackay IS, Bull TR (Eds) Scott Brown's Otolaryngology Rhinology 6th Ed London: Butterworth/Heinemann 1997: pp 4/24/1-4/24/19.
2. Martinson FD. Mucocoeles of the paranasal sinuses in Nigeria. Ghana Med. J. 1974; 13: 192-198.
3. Martinson FD, Odeku EL. The Problem of Unilateral Proptosis in Nigeria: International Surgery. 1969; 51: 20-29.
4. Ologe FE, Odebode TO, Owwoeye JFA, Eletta AP. Ophthalmic Manifestations of fronto-ethmoidal mucocoeles: A report of five cases. Afr. J. Med. med. Sci. 2003; 32: 209 – 214.
5. Chen TM, Lee TJ, Huang TS. Endoscopic Sinus Surgery for the treatment of fronto-ethmoidal mucocoele complicated with orbital abscess: a case report. Chang. Keng. I. Hsueh 1997; 20: 39-43.
6. Pia F, Aluffi P, Borello G. Frontal mucocoele: Open Surgery. Acta-Otorhinolaryngol – Ital. 1999; 19: 155-159.
7. Voegels RL, Balbani AP, Santos-Junior RC, Butugan O. Fronto-ethmoidal mucocoele with intracranial extension: a case report. Ear Nose Throat J. 1998; 77: 117-120.
8. Steinberg CM, Bailey BJ, Calhoun KH, Quinn FB. Management of invasive Fronto-ethmoidal sinus mucocoeles. Arch. Otolaryngology. Head Neck Surg. 1986; 112: 1060-1063.
9. Selvapandian S, Rajshekar V, Chandy MJ. Mucocoeles: a neurosurgical perspective. Br. J. Neurosurg. 1994; 8: 57-61.
10. Lund VJ, Milroy CM. Fronto-ethmoidal mucocoeles: a histopathological analysis. J. Laryngol. Otol. 1991; 105: 921-923.
11. Perie S, Sequert C, Cabanes J, Visot A, Krastinova D, Deroine P, Chabolle F. Frontal mucocoeles with orbital or cerebral extension: therapeutic strategy. Ann-Otolaryngol – Chir-Cervicofac. 1996; 113: 384-391.
12. Gurard B, Chondart L, Hamelin N, Agbague I, Ibazizen MT, Brasnu D, Cabanis EA. Fronto-naso-ethmoido-sphenoido-maxillo-orbital mucocoele with ophthalmologic presentation. J. Fr-Ophthalmol 1999; 22: 536-540.
13. Lasisi AO, Ajuwon AJ. Beliefs and perceptions of Ear, Nose and Throat-related conditions among residents of a traditional community in Ibadan, Nigeria. Afr. J. Med. Med Sci. 2002; 31:45-48.
14. Ologe FE, Nwawolo CC. Identifiable risk factors for chronic suppurative Otitis Media (CSOM) in a rural community in Nigeria. Nigeria Quarterly Journal of Hospital Medicine 2000; 10: 239-242.
15. Rubin JS, Lund VJ, Salmm B. Fronto-ethmoidectomy in the treatment of mucocoeles: A neglected operation. Arch Otolaryngol Head Neck Surg. 1986; 112: 434-436.
16. Smoot EC-3rd, Bowen DG, Lappert P, Ruiz JA. Delayed development of an ectopic frontal sinus mucocoele after paediatric cranial trauma. J. Craniofac. Surg. 1995; 6: 327-331.
17. Fligny I, Lamas G, Aidan P, Fongeront B, Soudant J. Frontal Mucocoele; Clinical symptoms, treatment and results: apropos of 17 cases. Acta. Otorrhinolaryngol – Belg. 1993; 47: 429-434.
18. Valles H, Palomar A, Blanc J, Sevil J, Fumanal L. Irreversible bilateral amaourosis secondary to the course of ethmofrontal mucocoele. Acta – Otorrhinolaryngol. Esp 1990; 41: 257-260.