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Ophthalmic manifestations of fronto-ethmoidal mucocoeles: a report of five cases

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Summary

Fronto-ethmoidal mucocoele is rare and will cause visual impairment and blindness when management is belated. In order to improve the awareness of its ophthalmic manifestations and problems associated with its management we retrospectively analyzed clinical, radiological and histological features in five illustrative cases. They all presented with unilateral supra-orbital swellings and proptosis, antedated by head injury in two cases. Other ophthalmic manifestations included eye lid swelling [3], progressive visual loss [2], unilateral ptosis [2], exotropia [2] and diplopia [1]. Only two patients had nasal symptoms. In all cases, x-rays of the paranasal sinuses revealed radiolucent frontal sinuses with eroded anterior walls, floors and midline septa. Brain scan in one case demonstrated opacification and enlargement of the corresponding sinus. Fronto-ethmoidectomy in four cases confirmed erosion of sinus' walls and floors and in one case of the contiguous supra-orbital ridge in addition to colored fluid aspirates. In all cases, the curetted sinus lining comprised of chronic inflammatory tissue. Surgical intervention was followed by prompt postoperative resolution of ophthalmic manifestations. A high level of suspicion is essential for early diagnosis and treatment of fronto-ethmoidal mucocoeles and will assist in preventing irreversible visual loss.

Keywords: Fronto-ethmoidal mucocoeles, ophthalmic manifestations

Résumé

Le mucocoede fronto-ethmoidal est rare, mais en cas de délai dans la gestion, il peut causer un faiblissement visuel et même un aveuglement complète. Pour méliorer son connaissance optalmique et des problèmes de sa gestion, nous avons analysé des cinq caractères clinique radiogique et historique dans les cas explicatifs. Tous les cas se manifestent de grosseurs supra orbitales et de proptosis, surtout a cause d'un injure de la tête dans deux cas. Les autres manifestations optalmiques sont: l'inflammation de la paupière de l'oeil (3) perte visuelle progressive (2) ptosisunilateral (2) et diplopie (1) Il y'a seulement deux malades qui avaient des symptômes nasaux. Dans tous les cas, les X-rays des sinus de para nasale on révèle les sinus fronteaux radiolucent et les parties antérieures érodés, en bas et au milieu les septa. Dans un cas, le scanneur du cerveau a démontre l'opacification et l'agrandissement du sinus correspondant. L'ethmoidectomie-fronto dans quatre cas afferme une erosion invétérée de murs du sinus' et sols et dans un cas de la corniche supra orbitale contiguë en plus de fluide coloré aspire. Dans tous les cas, l'intérieure des sinus curetés avait de tissu provocateur chronique. L'intervention chirurgicale a été suivie par une résolution Postopératoire de manifestations ophtalmiques. Un haut niveau de soupçon est essentiel pour diagnostic tôt et traitement de mucocoedes du fronto-ethmoidal pour prévenir la perte visuelle irrévocable.

Introduction

A mucocoele is an epithelial-lined, mucus containing sac which completely fills a sinus and is capable of expansion [1]. It is relatively uncommon and rarely bilateral [1-5]. The frontal and ethmoidal sinuses are most commonly affected [1,2] When it is secondarily infected it becomes a pyocoele. Generally, the evolution of a mucocoele is slow, but sometimes they evolve rapidly as acute inflammatory processes, occasionally discharging through the upper eyelid [1,2,5,6]. Paranasal sinus mucocoeles present with a multitude of symptoms including ophthalmic disturbances [6]. They are benign and typically cause ophthalmic morbidity after significant orbital extension. Expansion of the mucocoele generally follows the route of least resistance, into the orbit; such that majority of the patients are initially seen by the ophthalmologist on account of proptosis [1-3,5,7] Mucocoeles with intracranial extension are often reported by Otorhinolaryngologist and Neurosurgeons [6,8]. The neurosurgical and ophthalmic manifestations of sinus mucocoeles have been well documented with the frontoethmoidal and sphenoidal varieties presenting with

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proptosis and visual loss respectively [9]. Majority (91%) of the patients with fronto-ethmoidal mucocoeles exhibit some degree of proptosis (1-17mm), 55% exhibit lateral displacement (2-13mm) while 59% show inferior displacement (1-10mm) resulting in limited ocular mobility in the upward gaze [1]. The most common ophthalmic manifestations include lid swelling, ocular pain, pseudoptosis, proptosis, and displacement of the eyeball. Others include epiphora, blurring of vision, decreased visual acuity, visual field abnormality, diplopia, disc oedema, retino-choroidal folds and increased intraocular pressure [1-3,5,10,11].

A few cases of fronto-ethmoidal mucocoeles have been reported in Nigeria [2,3] but this is the first report from the University of Ilorin Teaching Hospital, Ilorin, Nigeria, a referral center which serves a large population of the middle belt of the country. This report illustrates its rarity, slow evolution, rapid evolution following secondary infection, ophthalmic manifestations, delay in presentation to the Otorhinolaryngologist and the need for team approach for accurate diagnosis and prompt treatment.

Patients and methods

We retrospectively studied five patients, three males and two females referred for further management from the Eye Clinic to the Otorhinolaryngology Clinic of this hospital between 1996 and 2001. Their case records were analyzed for age, sex, symptoms referable to mucocoele, eye symptoms and duration, presence of nasal symptoms, predisposing factors and duration, source of referral and clinical findings. Other analysis includes otorhinolaryngologic and ophthalmic findings, imaging and operative findings, histological diagnosis and postoperative ophthalmic status. Each patient had X-rays of the paranasal sinuses. One case had in addition, a cranial computed tomographic scan. At the time of this report, four of the patients had had an external frontoethmoidectomy via a Lynch Howarth's incision while surgical procedure is still being delayed in the fifth case because of financial constraints. The Neurosurgeon who is one of the authors participated in the management of the patients when intracranial extension was diagnosed or suspected or in anticipation of inadvertent penetration of the posterior wall of the frontal sinus during curettage.

Results

Clinical, ophthalmic, radiological and operative findings in all cases are summarized in Table 1. All the cases presented with a medial supra-orbital swelling of varying durations (6 months to 15 years) involving the left and



Fig.1: Case 1: A 42 year old female with left fronto ethmoidal mucocoele (post-operative status) and associated immature cataract. Note a healed Lynch Howarth's incision over the medial aspect of left supraorbital ridge.



Fig. 2: Case 2: A 42 year old male with left fronto ethmoidal mucocoele (pre-operative status). Note left supraorbital swelling (extending to the midline), Eyelid swelling and proptosis.

right sides in three and two cases respectively (Figs 1&2). Two of these cases presented with nasal symptoms including recurrent nasal blockage and discharge (cases 3&4) and another two (cases 2&3) with antecedent knife and hammer injury to the ipsilateral forehead occurring 10 and 15 years respectively prior to onset of initial symptoms. The most common ophthalmic manifestations were proptosis in all cases, eye lid swelling in 3 (fig.2), 20°

CASES	1	2	2	4	
Age(Years)/Sex (a) Signs/symptoms:	42/Female	42/Male	5 60/Female	4 17/Male	5 52/Male
Supra-orbital swelling/size (cmxcm) Headache Sinus discharge Duration	Left / 2x1.5 Present Present 2yrs	Left / 4x2 Present Absent 15yrs	Right / 4x4 Absent Absent 2yrs	Right / 5x1 Absent Absent 6 months	Left / lx2 Absent Absent 5yrs
(b) Eye symptoms/signs: Proptosis Ocular pain Epiphora Visual Acuity Lid swelling Ptosis Diplopia Exotropia	Present Present Absent Hand movement Present Complete Nil 20°	Present Present 6/6 Present Partial Nil Nil	Present Absent Present 6/24 Absent Absent Nil 15º	Present Absent Absent 6/6 Present Absent Nil Nil	Present Absent Absent 6/6 Absent Absent Present Nil
(c) X-Ray findings :: Radio-Lucency Erosion of ♦ Sclerosis	Present Anterior wall/ floor marginal	Present Anterior wall/ floor/septum nil	Present Anterior wall/ nil	Present Expanded sinus nil	Present Anterior wall/Floor nil
(d) Operative findings: I) Erosion of:	- Anterior wall & Floor	- Anterior wall +Supra-orbital	- Floor	- Anterior wall	Awaiting • operation
ii) Volume of fluid aspirate (mls) =	40	ridge 100	25	15	

Table 1: Clinical, ophthalmic, radiological and operative findings in Frontoethmoidal mucocoeles.

and 15° exotropia in 2, ptosis in 2 cases as well as diplopia and sinus discharge in 1 case each. Visual acuity was reduced in two cases to hand movement and 6/24 respectively. None of the patients was blind at presentation (Table 1). In addition, Case-1 presented with an ipsilateral mature cataract (fig.1). X-rays of their paranasal sinuses revealed radiolucency and erosion of the anterior walls, floors or midline septa of the frontal sinuses (Figs.3 & 4, Table 1). In addition, brain scanning in one patient (case 1) showed enlargement and opacification of right frontal and ethmoidal sinuses (Fig 5) in addition to erosion of the posterior and inferior walls of the frontal sinus (fig 6). Operative findings in four patients included erosion of the anterior wall (1 case), floor (1 case), anterior wall and ipsilateral supra-orbital



Fig. 3: Antero-posterior view of the paranasal sinuses in Case 2 showing enlargement and radiolucency of the left frontal sinus



Fig.4: Lateral paranasal sinus film in Case 2 showing erosion of the anterior wall and floor of the frontal sinus.



Fig. 5: Cranial computed tomography (CT) in Case 1(Table 1), showing left proptosis and opacification of left frontal sinus.



Fig. 6: CT scanogram of Case 1 showing crosion of the posterior wall and floor and widening of the frontal sinus, with suspected intracranial extension.

ridge (1 case), and anterior wall and floor (1 case) of the frontal sinuses. Sinus aspirates were greenish, straw colored or brownish mucous fluids with volumes ranging from 15 to 100 cm³. Histology of curetted sinus linings revealed chronic inflammatory tissue. There was prompt resolution of ophthalmic manifestations in all operated cases within six months of surgery.

Discussion

The formation of a mucocoele has traditionally been attributed to a combination of the obstruction of the affected sinus and inflammation[1]. Previous surgery or trauma may contribute to the obstruction, but a significant proportion may not have such contributory factors[1,6,10,11]. 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 infections. Histological studies confirm the presence of bone remodeling at the interface between mucocoele and sinus wall [1]. Recent studies on boneresorbing factors confirm the presence of prostaglandin E, (PGE,), collagenase, and cytokines such as interleukin-1 and tumour necrosis factor (TNF) compared with normal controls and chronically inflamed mucosa [1].

Generally, the evolution of a mucocoele is slow, but sometimes they evolve rapidly as acute inflammatory processes, occasionally discharging through the upper eye lid [1,2,5,6] Expansion of the mucocoele generally follows the route of least resistance, into the orbit; hence majority of the patients are initially referred to the ophthalmologist on account of proptosis [1-3,5,8]. There may also be intracranial extensions [6,8].

Para-nasal sinus mucocoeles are relatively rare. In a one year review of 2,890 patients presenting to the ENT clinic in Jos, Nigeria, Bhattia and Varughese⁴ found no case of sinus mucocoeles. And when they do occur they are uncommon in children [2]. There is a male preponderance with a male to female ratio of 3:1 [2]. Fronto-ethmoidal mucocoeles causing unilateral proptosis as in the cases presented, form the bulk, sometimes up to 80%, of the cases of paranasal sinus mucocoeles [2,5]

Trauma or paranasal sinus operations in young patients may lead to mucocoele formation due to scarring in the wound [10]. Such mucocoeles develop 15 to 25 years after the initial trauma or surgical procedure [6,11]. Only two of our patients (cases 2 & 3) sustained head trauma antedating symptoms by 15 and 10 years respectively. Case 2 illustrates the characteristic slow evolution while Case 1 shows the occasional rapid

evolution that follows acute secondary infection [6]. While many cases of fronto-ethmoidal mucocoeles may not have nasal symptoms as in three of our cases, some have accompanying symptoms of sinusitis (cases 3&4) [13]. A small percentage (10-15%) may actually present with nasal polyp [7]. Lack of nasal symptoms delays referral to the Otolaryngologist and prolongs the time lapse to diagnosis. Ophthalmic symptoms are the most dominant features of fronto-ethmoidal Mucocoeles [2,3,5,7,10]. This explains why patients, as in this series, presented first to the Ophthalmologist. A discharging sinus is a unique feature which hastens early search for medical help in this environment. It usually involves the upper eye lid but occasionally the forehead [2,5] (Case 1). It is the result of spontaneous rupture of the mucocoele in most cases, but in a few cases as in our setting, it follows deliberate incision by local herbalists and rarely by medically qualified persons [2]. Scarring around the discharging sinus in the upper eye lid could lead to varying degrees of retraction or even eversion (ectropion) [2]. The principal presenting complaint is proptosis, present in over 80%, with varying degrees of downward and lateral displacement of the globe [7,12]. Other ophthalmic morbidity occurs only after significant orbital extension leading to compressive optic neuropathy, or even compression of the chiasma and concomitant visual loss [12]. It is important to promptly and correctly diagnose mucocoeles on the basis of clinical and neuro-radiological findings in order to foster early surgery and prevent permanent visual loss [12]. 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 [11]. 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. Because of this, early and correct diagnosis and early presentation to the Otorhinolaryngologist become pertinent. In this country however, the patient often consults local herbalists, chemists and quacks before ever getting to a specialist hospital [2]. And when they choose orthodox medicine they usually pass through the general practitioner and other specialists including the Ophthalmologist, Neurologist, or Neurosurgeon before final referral to an Otorhinolaryngologist. This delays both diagnosis and treatment. A team approach involving the Ophthalmologist, Radiologist, Otorhinolaryngologist, and Neurosurgeon is essential for accurate diagnosis and treatment.. The differential diagnoses of fronto-ethmoidal mucocoele include frontal osteoma, fibro-osseous

dysplasia, subcutaneous tumours, fronto-nasal encephalocoele, meningioma, neurilemomma and malignant diseases which could be primary or metastatic commonly from the kidney, breast or lung [7].

Computerized tomographic (CT) scanning or magnetic resonance imaging (MRI) have been found to be more sensitive than plain radiography in establishing diagnosis of sinus diseases including mucocoeles. Unfortunately, CT and MRI facilities are not available in most hospitals in developing countries. Where they are available, the cost is far beyond the reach of the average patient and his relatives. Only one of our patients could afford a CT brain scan. Plain radiography therefore remains a relevant tool in the diagnosis of fronto-ethmoidal mucocoeles in our environment. It has the advantage of lower cost and easy availability over CT and MRI, but is open to observer errors in interpretation [13].

Until recently, the majority of fronto-ethmoidal mucocoeles were treated by external frontoethmoidectomy, via a Lynch Howarth approach, as we have done for our patients [2]. However, functional endoscopic marsupialization is gaining popularity [6] Endoscopic surgery is an effective treatment which avoids a scar and is associated with less diplopia as the trochlea is not disturbed [1]. In this center, as in most centers in developing countries, there is no facility for endoscopic sinus surgery. Although functional endoscopic surgery enhances the chance for excellent result in the treatment of most sinus mucocoeles, open surgery remains a valid procedure in frontal mucocoeles with orbital and/or cranial extension or osteomyelitis and in situations where facilities for endo-nasal approach is not available [8].

Conclusion

Our experience suggests that ophthalmic manifestations of fronto-ethmoidal mucocoeles are common in Nigeria. The problem is that many cases are not recognized, and suspected cases are characteristically diagnosed and treated late. A high level of suspicion of mucocoeles as a cause of unilateral proptosis or progressive visual loss is therefore essential for making early diagnosis and effecting prompt surgical treatment. This is the key to prompt resolution of established ophthalmic manifestations [9] and prevention of irreversible visual loss and blindness.

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