Frontoethmoidal mucocoele with orbital and intracranial extension

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Abstract

We report a case of a giant frontoethmoidal mucocoele extending to the orbit and anterior cranial fossa. Clinically, the patient had an insidious onset of clinical presentation with no intracranial symptoms. We present its CT, MRI and histopathological features with a brief review of literature. Various surgical approaches are also discussed.

Keywords Frontoethmoidal mucocoele · Paranasal sinuses

Introduction

Mucocoele of the paranasal sinuses is a slowly expanding benign lesion that develops when the physiological drainage of the mucous produced by the epithelial lining of the paranasal sinuses is impeded. The occlusion of the ostia may be secondary to infection, allergy, trauma, benign neoplasm (osteoma or fibrous dysplasia), malignant or metastatic tumours [1]. It has also been suggested that a cystic degeneration of a goblet cell gland could result in a mucocoele. Rarely the cause is not known. They present in the third or fourth decade, affecting men and women equally.

Case report

A 58-year-old woman presented with complaints of a swelling above the medial aspect of the right eye, heaviness of the right upper eyelid and epiphora of 2 years duration. She gave history suggestive of occasional sinusitis, but she had no persistent nasal block. She had undergone cataract surgery of the right eye 2 years back. On examination, the right eyeball was displaced downwards and laterally with restricted upward eye movement

The swelling which was smooth and nontender, was found extending from over the right frontal sinus upto the medial canthus of the right eye. This had the typical egg shell crackling consistency. Anterior rhinoscopic examination was normal. Ophthalmological examination revealed pseudo aphakia of the right eye with a vision of 6/24, and immature cataract of the left eye with only perception of hand movements at half a metre distance.

CT scan of the head showed a large soft tissue density mass of +20–40 HU filling the expanded right frontal sinus and anterior ethmoid cells, eroding the superomedial wall of the right orbit, displacing the eyeball downwards and laterally (Fig. 1). Both the anterior and posterior tables of the frontal sinus were eroded with the lesion extending to the anterior cranial fossa (Fig. 2). There was no significant enhancement of the lesion after contrast administration. No calcification was noted. Provisional diagnosis of mucocoele was made.

Surgery was carried out by a team of ENT surgeons and Neurosurgeons. The osteoplastic flap was raised by an eyebrow incision which was then curved down and extended 1.5 cm below the medial canthus of right eye. Bony defect over the frontal sinus was widened and the mucocoele was removed from the bony wall. The mass was seen extending into the ethmoids. Anterior ethmoidectomy was performed and the capsulated mass was completely stripped from the involved sinuses. Dura could be seen through the defect in the posterior wall of the frontal sinus and was found to be intact. Frontonasal drainage was fashioned through the frontal sinus floor, using an endotracheal tube of size 4.5 mm internal diameter. The flap was replaced and sutured. Post operative period was uneventful. Injection cefuroxime was given for 10 days. She was discharged 2 weeks later.

Discussion

The term mucocoele was first described by Rollet in 1896 [2]. Commonest sinus to be involved is the frontal sinus, frequently in association with anterior ethmoids (64%), and others in the order of frequency are maxillary sinus, sphenoid and posterior ethmoid sinus.

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Stenosis of the ostium is most likely seen in the frontal sinus where the narrow duct which is around 2mm wide passes for a distance of 5–10 mm through the ethmoid bone in close relationship to the ethmoid sinuses [3]. Eventually the mucocoele expands out of the sinus into the neighbouring structures such as orbit, cranial vault and facial tissues. Erosion of the bony walls occurs at the thinnest portion, being the orbital wall of the horizontal part of the ethmoid in 90% of the cases. It may remain uninfected and asymptomatic till there is considerable displacement of the eyeball. In children, a mucocoele from the ethmoid is more common

Fig. 1 CTscan Head with contrast, showing a soft tissue density mass filling the expanded right frontal sinus, anterior ethmoidal cells, eroding the superomedial wall of the right orbit, displacing the eyeball laterally and downwards, and extending to anterior cranial fossa.

Fig. 2 Axial CTscan Head with contrast, showing erosion of the anterior and posterior wall of the frontal sinus with soft tissue lesion involving the frontal sinus and anterior cranial fossa.

and expands into the orbit by eroding through the lamina papyracea [3].

Histological findings in a mucocoele are of an ulcerated hyperplastic mucosa with chronic nonspecific inflammation, malpighian metaplasia, surrounded by a reactive fibrous capsule [2].

Erosion of the sinus walls are caused by the mass effect of the lesion and by the osteolytic activity of the cytokines such as interleukins IL-1 and IL-6 from the fibroblasts of the mucocoele lining [1].

Clinically, the most common feature is a swelling which develops above and medial to the eye. Such a swelling occurs slowly and takes years to become evident. The eye is slowly pushed downward and outward, later causing diplopia. Palpation discloses a nontender, smooth, partially compressible mass. Large lesions produce an increased feeling of pressure and mass effect. Eyeball movements get restricted on upward and inward motion. Epiphora, astigmatism, chemosis, eyelid oedema, diminished vision, ophthalmoplegia and optic neuritis are the other ocular features [2]. Sphenoidal mucocoeles can cause ocular motor nerve palsy [4]. Due to these symptoms, patients may at first present to the ophthalmologist before they are referred to the ENT surgeon.

In 10% of the cases, the initial presentation is forehead swelling. On palpation, one can elicit the typical egg shell crackling sensation. Incidence of skull base erosion is around 10 to 20% [2]. Erosion of the posterior wall of the frontal sinus can lead to meningitis, meningoencephalitis, CSF fistula etc [2]. Headache and rarely, nasal obstruction with nasal mass may be seen.

Radiologically, x-ray films show loss of continuity of the bony supraorbital ridge or displacement of the bony floor of the frontal sinus. CT scan features include a homogenous expansile mass filling the sinus cavity and surrounding structures. The expansile appearance is due to the gradual pressure atrophy and erosion of the bone. In CT studies with contrast injection, there is no increase in the intrinsic density of mucocoele due to its avascular mucoid content. Rim enhancement is seen in capsular inflammation. On MRI scan, the mass is heterogenous, being hypointense on T1 weighted images and hyperintense on T2 weighted images. MRI features vary depending on the stage of development and the protein content. High intensity on T1 weighted images indicate high protein content and hypointensity on T2 weighted images shows thick mucous [4].

Differential diagnosis include orbital abscess (inflammatory swelling with fever), dacryocystitis (swelling below the medial canthal ligament), pneumosinus dilatans (abnormal enlargement of the frontal sinus which is aerated and has well defined margins), frontal sinus tumours like osteomas, (persistent feeling of pressure in the frontal region), meningocele, dysmormonal exophthalmos, orbital tumours etc [3]. Further radiological studies help in differentiating these various conditions.

A pyocele is an infected mucocoele. A delay in the treatment of a pyocele may lead to complications like orbital abscess, meningitis, subdural empyema or cavernous sinus thrombosis

Prior to endoscopic sinus surgery, patients with frontal sinus mucocoele were treated with an external surgical approach where the sinus mucosa was removed and either the sinus was obliterated using fat or in a more conservative manner, a drainage tube was placed after removing the sinus floor [1]. The osteoplastic flap technique is the procedure of choice in extensive frontal sinus disease. The flap is raised either through a bicoronal or eyebrow incision. This technique provides good cosmetic approach, opening and drainage of the frontal sinuses, and a clear frontonasal drainage [1].

Endoscopic sinus surgery is now gaining popularity in the management of paranasal sinus mucocoeles. Being minimally invasive, this has reduced the operative morbidity of external surgery. Ethmoido-frontal, maxillary, sphenoidal and ethmoidal mucocoeles are excellent indications for an exclusively endoscopic marsupialization [5]. A medially localised frontal mucocoele may be managed endoscopically. The presence of bony erosion of the orbital and posterior walls of the frontal sinus does not preclude an endoscopic approach as long as there is no intracranial complication [1]. But for large, laterally localised lesions with orbital and intracranial extension, an external approach combined with endoscopic surgery is preferred for both wide exposure and reconstruction. Modified endoscopic Lothrop technique which involves removal of the frontal sinus floor with frontal septectomy and resection of superior part of the nasal septum to create a large nasofrontal communication is now fast replacing the open surgical frontal sinus obliteration technique [6].

In this case, the patient had a large frontoethmoidal mucocoele with erosion of both the anterior and posterior walls of the frontal sinus and superomedial wall of the orbit which justified an open surgical approach and a wide nasofrontal communication. After 6 months of follow up there has been no recurrence.

Conclusion

With CT and MRI guided images, endoscopic sinus surgery and marsupialisation of mucocoeles is gaining acceptance as an alternative choice to the traditional open surgical techniques. While this remains as a viable alternative in the management of paranasal sinus diseases, an external approach combined with endoscopic sinus surgery seems to be the choice of treatment in extensive frontal sinus disease.

References

1. Molteni G, Spinelli R, Parigatti S, Colombo L, Ronchi P (2003) Voluminous frontoethmoidal mucocoele with epidermal involvement. Surgical treatment by coronal approach. *Acta Otolaryngol Ital* 23:185–190
2. Cansiz H, Yener M, Guvenc MG, Canbaz B et al (2003) Giant frontoethmoid mucocoele with intracranial extension: Case report *Ear, Nose, Throat* 82:50–52
3. Scot-Brown's text book of Otolaryngology; 1987; 5th edition- Butterworth – Heinmman; 206–207
4. Akan H, Cihan B, Celenk K (2004) Sphenoid sinus mucocoele causing third nerve paralysis: CT and MR findings. *Dento-maxillofacial Radiology* 33:342–344
5. Venail F, Marlier F et al (2003) Combined approach (endoscopic and external) for the treatment of sinus mucocoele. *Rev Laryngol Otol Rhinol (Bord)* 124(3):165–170
6. Kontakis SE, Gross CW (2003) Long-term results of the Lothrop operation. *Curr Opin Otolaryngol Head Neck Surg* 11(1):37–40