An unusually giant frontoethmoidal mucopyocele in a child – A case report

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A B S T R A C T

Mucoceles are expansile benign mucus-filled cystic lesions of the paranasal sinuses as a result of the chronic obstruction to sinus ostia. It is rarely seen in children. Secondary infections of the mucoceles result in the formation of a mucopyocele. The proximity of these lesions to the orbit and brain may cause morbidity and potential mortality, if left without intervention. Here we came across a 12-year-old child who had a giant right side frontoethmoidal mucopyocele causing proptosis and lateral displacement of the right eye with normal visual acuity. Here, the case, the experience and the outcome of the endoscopic marsupialization of the frontoethmoidal mucopyocele, are being discussed.

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Introduction

Mucocele is a benign and expansile cystic lesion lined with respiratory mucosa of pseudo stratified columnar epithelium [1]. Mucoceles arising inside the paranasal sinus are expansile in nature, slow in growth and may affect neighboring structures such as orbit and the intracranial structures [2]. Mucoceles of the paranasal sinuses were first documented by Langenbeck (1820) under the name of hydatids and Rollet (1909) coined the name mucocele [3]. An infected mucocele is called a pyocele or muco-pyocoele. The incidence of mucopyoceles of paranasal sinuses in general population is 0.4–0.8% [4].

Any paranasal sinus can form a mucocele but frontal and ethmoid sinuses are most commonly affected followed by sphenoid and maxillary sinus. Expansion of mucocele may take place over several years whereas it expands rapidly when associated with secondary infection leading to pyocele. Frontoethmoidal, ethmoidal, maxillary and sphenoid sinus mucopyoceles are excellent indications for exclusive endonasal endoscopic surgery. The endoscopic approach avoids external incision and associated morbidity and it has distinct advantage over the external frontoethmoidectomy. We report an unusual giant mucopyocele arising from the bulla ethmoidalis and frontal sinus invading into the orbit of a 12-year-old boy managed by endoscopic marsupialization.

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Case report

A 12-year-old boy presented at the Otorhinolaryngology outpatient department with complaints of gradually progressive right nasal obstruction, frontal headache and right side orbital proptosis since last 2 years. He had complaints of right side facial pain with reduced sense of smell. There was no history of nasal discharge, nasal bleeding, loss of vision or epiphora. He has no history of allergic rhinitis with no other medical co-morbidities. There was no previous history of trauma or surgery. He was taking treatment from local physician for proptosis of eye for over two years but no improvement. On examination, there was proptosis in the right side with downward and outward displacement of right eye with normal eye ball movements (Fig. 1). Diagnostic nasal endoscopy showed a large smooth bulging over the right lateral wall of nose above inferior turbinate with presence of pus points in the middle meatus (Fig. 2). The swelling seemed to impinge the septum. Computed tomography (CT) scan of the nose and paranasal sinus showed a large soft tissue density lesion of size 12 × 7 × 5 cm in the right frontoethmoid area (Fig. 3). The patient underwent endoscopic excision of the mucocele with a wide middle meatal antrostomy and opening of the frontal recess area (Fig. 4). Purulent fluid was seen intraoperatively from the mucocele. A bacteriological swab taken from the pus inside the pyomucocele revealed growth of Gram positive cocci (Staphylococcus albus and Staphylococcus viridians) in the culture. Post-operatively, the patient is symptom free.

Discussion

The long-standing obstruction of sinus ostia or compartment of septate sinus helps in the formation of benign,
mucus-filled cystic, and expansile lesion of the paranasal sinus is called as mucocele. Mucoceles usually occur between the age of 40 and 70 years and it is rarely seen in children [5]. It has a slight male predominance but right and left sides are equally involved. Mucocele can occur in any of the paranasal sinuses but frontal sinus (60–89%) is most commonly involved followed by ethmoidal sinus (8–30%), maxillary sinus (<5%) and is rarely seen in the sphenoid sinus [6]. The mucoceles usually behave like space occupying lesions that leads to bone erosion and the displacement of the surrounding structures.

The exact etiology for origin of mucocele is not known but one proposed mechanism for formation of mucocele is cystic degeneration of a seromucinous gland, resulting in a retention cyst. There are certain risk factors associated with mucocele formation such as chronic low grade infection/inflammation, previous sinus surgery, allergy, radiotherapy, trauma and tumors [7]. The mucocele in the paranasal sinuses is formed when its draining mucus gets blocked by an obstruction at its ostium. Rarely, benign lesion like cystic fibrosis or malignant neoplasm resulting in obstruction of ostium can lead to formation of the mucocele [6]. The osteoclastic bone resorption enhanced by release of Prostaglandin (PGE2) and proinflammatory cytokines (IL-1 and TNF-α) into the wall of the mucoceles has been suggesting inflammatory etiology of mucocele [8].

Common clinical presentations of the mucocele are headache, facial pressure, facial swelling or deformity, dental pain, nasal obstruction, orbital manifestations like periorbital pain, proptosis, impaired orbital mobility and diplopia. Mucocele may compress the optic nerve, resulting in loss of vision. Intracranial extension by eroding the posterior wall of the frontal sinus can lead to CSF rhinorrhea, meningitis and brain abscess [9]. The patient we reported presents with right side proptosis without visual disturbance, nasal obstruction and right side facial pain with reduced sense of smell.

The expansile growth of the mucocele usually follows the path of least resistance that is into the orbit. So, in majority of the patients, lateral and inferior displacement of the orbit, diplopia, limited ocular motility in upward gaze, sometimes with a characteristic egg shell cracking sensation due to thinning of the overlying bone. Secondary infection of mucocele leads to pyoceles, which causes rapid expansion, compromising the vision [10]. Studies have shown that high levels of prostaglandin E2 play an important role in the osteolytic process in mucoceles and explain the locally aggressive behavior of the expansile lesions [11]. The differential diagnoses of mucoceles are encephaloceles, cholesterol granuloma, epidermoid cyst, meningioma, chordoma, neurofibroma, salivary adenoma, paraganglioma, nasoanangiofibroma, and malignant neoplasms [12].

The diagnosis of the mucopyoceles is based on the clinical features and radiological tests like computed tomography (CT) and magnetic resonance (MR) imaging. CT and MR are both considered as the gold standard for diagnosis for these lesions. The imaging of the choice is CT scanning in both axial and coronal planes [13]. It clearly evaluates the amount of expansion of the lesion and surrounding bone erosion [12]. MRI is usually identify the relationship between the lesions and surrounding vital structures like brain, orbit and affected other soft tissues. Histopathology examination shows they are cyst like structures lined with respiratory epithelium and filled with mucus.

The mainstay of the treatment of mucocele or mucopyocele is surgery, which ranges from endoscopic sinus surgery to craniotomy, and craniofacial exposure, with or without obliteration of the sinus [13]. The purpose of the surgical treatment is to clear the lesion, radically remove the paranasal sinus mucosa to prevent recurrence and re-establishment of a plane of separation between the extracranial and intracranial space and at the same time to achieve the cosmetic results.

Intranasal endoscopic marsupialization was first reported by Kennedy et al. in 1989 and presently this surgical approach is the treatment of choice as it is less invasive and has less morbidity in comparison to conventional technique like external frontoethmoidectomy and osteoplastic flaps with or without obliteration of the sinus [14]. Transnasal endoscopic approach and marsupialization give complete recovery in 82–96% of cases [15]. It is important to understand that mucoceles are prone to recurrence if inadequate marsupialization is done. Endoscopic approach has now become a standard treatment but occasionally very large size complex one needs an open external approach to widen the drainage and prevent recurrence. After surgery, the displacement of the orbit to normal position takes several months while bone remodeling occurs. In our case, the patient had undergone endoscopic marsupialization which is considered as a safe and effective approach with less postoperative complications. The patient is now on follow-up and he is symptom free without any evidence of recurrence.

**Conclusion**

A giant frontoethmoidal mucopyocele in a child is an uncommon diagnosis. The diagnosis is confirmed by radiological
tests. Surgery with endoscopic marsupialization is a safe and
effective procedure with less recurrence. Accurate and early
diagnosis with appropriate surgical intervention and
a collaboration among otolaryngologist, ophthalmologist and
neurosurgeons are always crucial. Delayed diagnosis of muco-
pyocele of paranasal sinus leads to paranasal sinus expansion
and invasion of vital structures such as skull base and the
orbit. Early intervention to mucopyocele leads to significant
reduction of orbital and intracranial complications.

Authors’ contributions/Wkład autorów

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Conflict of interest/Konflikt interesu

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Ethics/Etyka

The work described in this article have been carried out in
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