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A Primary Ethmoid Mucocele in a 9-Year-Old Child: A Case Report and Literature Review

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Abstract

A mucocele is a mucous-filled cavity in the paranasal sinuses due to obstruction or compression of neighboring structures by inflammatory processes, trauma, or prior surgery. It can be indolent, locally expanding, and destructive. Mucoceles are extremely rare in children. We report a nine-year-old boy with sudden-onset left orbital pain and proptosis. Computed tomography showed a well-defined soft tissue lesion originating in the left ethmoid sinuses, suggestive of a mucocele. The mass was marsupialized by endoscopic sinus surgery. Mucoceles should be a part of the differential diagnosis of a child presenting with proptosis. Imaging studies are helpful in establishing the diagnosis.

Keywords: Child • Pediatrics • Ethmoid • Sinus • Mucocele

Introduction

A mucocele is mucous-containing cavity in the paranasal sinuses. Complete obstruction of the natural ostium of the affected sinus causes increased pressure and a mass effect on the surrounding structures. Mucoceles grow slowly, and years may pass between sinus obstruction and the appearance of symptoms. Mucoceles are rarely seen in children, and when they are present, they are secondary to an inflammatory process, tumor, trauma, or previous surgery [1]. Cystic fibrosis has also been linked to mucocele formation due to ciliary motility impairment [2]. A primary sinus mucocele in children is exceedingly rare. The presenting symptoms of a sinus mucocele depend on the site of origin and the structures affected by the expanding mass. Computerized tomography (CT) is the modality of choice for the diagnosis of sinonasal masses in general and mucoceles in particular. The purpose of the current report is to describe this rare entity and present a review of the literature.

Literature Review

The English literature was searched through PubMed using the keywords “mucocele”, “sinus”, “pediatric”, and “children”. The titles of all the retrieved papers were read, and the abstract of the article was also read if it described a mucocele in children. All abstracts of papers on patients older than 18 years or on mucoceles secondary to a trauma, tumor, or inflammatory disease were excluded. The remaining papers were read in their entirety. All papers with incomplete data on the patient’s age or whether or not the mucocele was a primary lesion were also excluded. The literature review yielded 13 children with primary mucoceles, and their demographics, operative data, and outcome were recorded (Table 1) [1-12].

Case Presentation

A nine-year-old male presented to the ear nose and throat clinic with a history of persistent unilateral left nasal obstruction for the preceding few weeks. Frontal headache, left orbital pain and proptosis appeared two days before admission. The past medical history was unremarkable, and there had been no previous head trauma or nasal surgery. The patient did not complain of fever, cough, rhinorrhea, or epistaxis. The physical examination revealed left eye proptosis and inferolateral displacement of the globe. The patient had no diplopia, the pupils were equal and reactive, and visual acuity and color vision were normal. The nasal endoscopic exam revealed edema and mucopurulent secretions in the left middle meatus. No mass was visible in the nasal cavity. The findings of a complete head and neck examination were within normal limits. A CT demonstrated a smooth-bordered expansile soft tissue lesion centered at the posterior ethmoid complex causing inferolateral displacement of the globe, compatible with an ethmoid mucocele (Figure 1).

Since the radiological appearance of the lesion was highly suggestive of mucocele, a decision was made to open the lesion and drain its content. A frozen section and excision of the mass with clear margins was planned in the unlikely possibility of the content being solid. The child's parents received a full explanation about the surgical plan and provided their signed consent. Following a course of antibiotic therapy of 10 days, the patient underwent endoscopic sinus surgery. The maxillary and ethmoid sinuses were opened and the medial anterior and posterior walls of the mucocele were removed, effectively marsupializing it into the nasal cavity (Figure 2). The left nasal cavity was packed with biodegradable/fragmentable, synthetic polyurethane foam (NASOPORE®, Polyganics, Groningen, and the Netherlands). Postoperative cleaning of the nasal cavity was performed until a clean and healthy cavity was achieved. The postoperative CT examination that was performed 8 months after surgery demonstrated an open and self-draining cavity with no evidence of recurrence (Figure 3). The patient was followed up endoscopically for 24 months with no evidence of recurrent disease.

Discussion

A mucocele is a mucous-filled sinus cavity secondary to complete obstruction of its opening. The involved sinus gradually expands via bone erosion with resultant pressure on surrounding structures. Most commonly occurring in the frontoethmoid area [13], mucoceles may affect any sinus and even the concha bullosa of the middle turbinate [4]. Most mucoceles occur in adults and are secondary to trauma, previous surgery, or other sinus pathologies, such as sinonasal polyposis and tumors [14]. Primary mucoceles in children are exceedingly rare, with few reports in the English literature. This unusual condition has been described in the middle turbinate [4], as well as the sphenoid [5,7], maxillary [8], and frontal [10] sinuses. Headache, pain,

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Table 1. The literature review yielded 13 children with primary mucoceles, and their demographics, operative data, and outcome were recorded.

Case No	Author	Age	Sex	Location	Signs and Symptoms	Duration of symptoms	Surgical Technique	Outcome	Follow-up
1	Al-Dousary	2.5	Female	Maxillary	Nasal obstruction purulent rhinorrhea cheek swelling	4 weeks	Endoscopic	Complete recovery	Not stated
2	Aslan1	5	Male	Middle Turbinate	Nasal Obstruction	3 years	Endoscopic	Healthy	18 months
3	Casteels2	10	Female	Sphenoid	Sudden Blindness	5 days	Endoscopic	Lt eye – regained vision6/12 Rt eye -blindness	3 months
4	Ceylan3	11	Female	Ethmoids	Left Nasal Obstruction, frontal headache and orbital pain	3 years	Endoscopic	Healthy	6 months
5	Diaz	11	Female	sphenoid	Proptosis visual loss diplopia and cheek swelling	7 years	Temporal Craniotomy	Improved diplopia and cheek swelling no visual improvement	Not stated
6	Haloi	5	Male	sphenoid	Headache lethargy Bilateral visual loss	2 weeks	Endoscopic	Complete recovery	2 months
7	Martinez	14	Female	Maxillary	Uepper molar tooth pain cheek swelling and facial pain	3 months	Endoscopic	Complete recovery	6 months
8	Nishie	12	Mele	Frontal	Frontal swelling	3 weeks	Not stated	Complete recovery	Not stated
9	Plikaitis	9	Female	Frontal	Frontal mass	1 year	Bicoronal incision and osteoplastic flap	Complete recovery	6 months
10	Skoulakis	8	Male	Maxillary			Open approach		Not stated
11	Suri	10	Female	Fronto-ethmoid	Frontal swelling proptosis nasal obstruction	Incidious onset – no specific time stated	Open approach	Complete recovery	Not stated
12	Suri	16	Female	Frontal	Frontal swelling proptosis	Incidious onset – no specific time stated	Open approach	Complete recovery	Not stated
13	Suri	16	Female	Frontal	Frontal swelling proptosis diplopia	Incidious onset – no specific time stated	Open approach	Complete recovery	Not stated

[1]Aslan G, Ugur MB and Bassullu N. "Giant Mucocele Originating from the Middle Concha in a 5-year-old Child: A Case Report." J Med Case Rep 7 (2013): 246.
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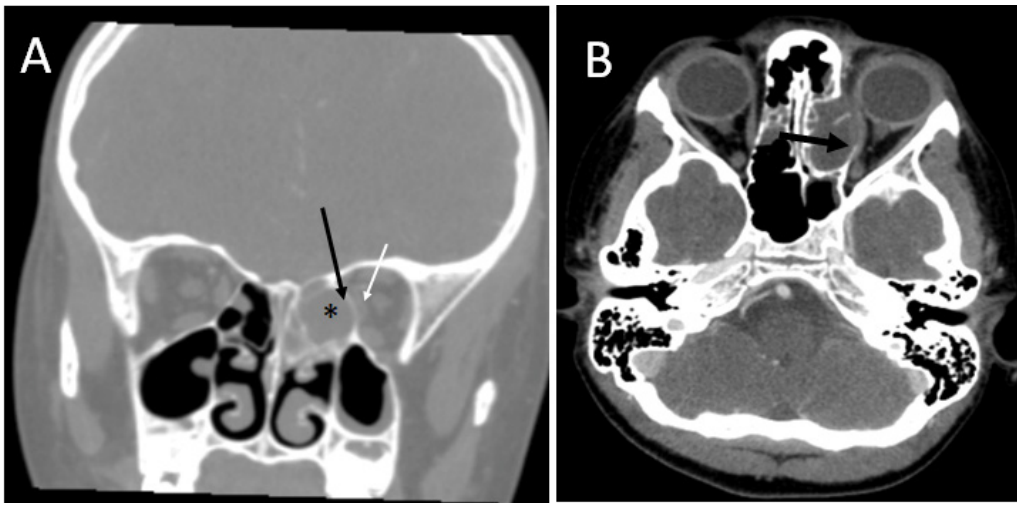

Figure 1. Pre-operative coronal (A) and axial (B) CT scan. A soft tissue mass (asterisk) is centered at the posterior ethmoid complex causing displacement of the lamina papyracea (black arrow) and medial rectus muscle (white arrow).

diplopia, and proptosis are the most common symptoms of mucoceles [15]. When a mucocele develops in the ethmoids, displacement of the lamina papyracea might result in proptosis and diplopia. A maxillary mucocele might cause bulging of the medial maxillary wall and nasal obstruction, while a frontal mucocele might press against the posterior wall and cause brain compression and cerebrospinal fluid leak [16]. A sphenoid mucocele might cause optic nerve disfunction that is usually reversible after surgery [17]. When situated in the posterior ethmoids, a mucocele can also create optic nerve pressure mimicking a sphenoid mass [18]. The radiological appearance of mucocele is that of an expansile lesion in an airless sinus, with thinning and erosion of its bony walls. Magnetic resonance imaging provides information on soft-tissue contents of the mass and on any involvement of the brain and orbit surrounding it.

The differential diagnosis of mucoceles includes encephaloceles, cholesterol granulomas, epidermoid cysts, meningiomas, chordomas, neurofibromas, salivary adenomas, paragangliomas, angiofibromas, and malignant neoplasms [19]. Moriyama et al. [18] found that in the time from surgery to the appearance of symptoms varies from 11 to 49 years in postoperative mucoceles. Due to such slow expansion, mucoceles may become very large before the patient seeks medical attention. Orbital pain and proptosis were the presenting symptoms of an ethmoid mucocele in the child we describe, leading us to recommend that the differential diagnosis of orbital pain in children should include the possibility of ethmoid expansile pathology, however rare. In the past, the surgical approach for mucoceles was mainly external and performed by a Lynch-Howarth external frontoethmoidectomy or a frontal osteoplastic operation. They have been replaced by endoscopic endonasal approaches for most patients, and endoscopic wide mucocele marsupialization was reported to have a 99% cure rate in a large series of patients [13]. Similarly, Moriyama et al. [18] published a series of 47 patients

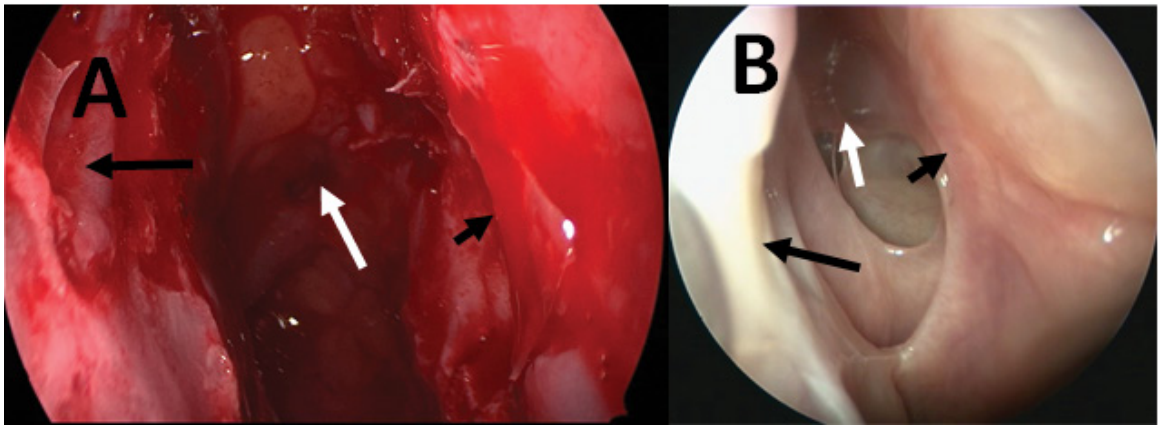

Figure 2. Endoscopic intraoperative (A) and postoperative (B) pictures of an open ethmoid cavity. The lamina papyracea (short black arrow), middle turbinate (long black arrow) and skull base (white arrow) represent the anatomical boundaries of the ethmoid complex.

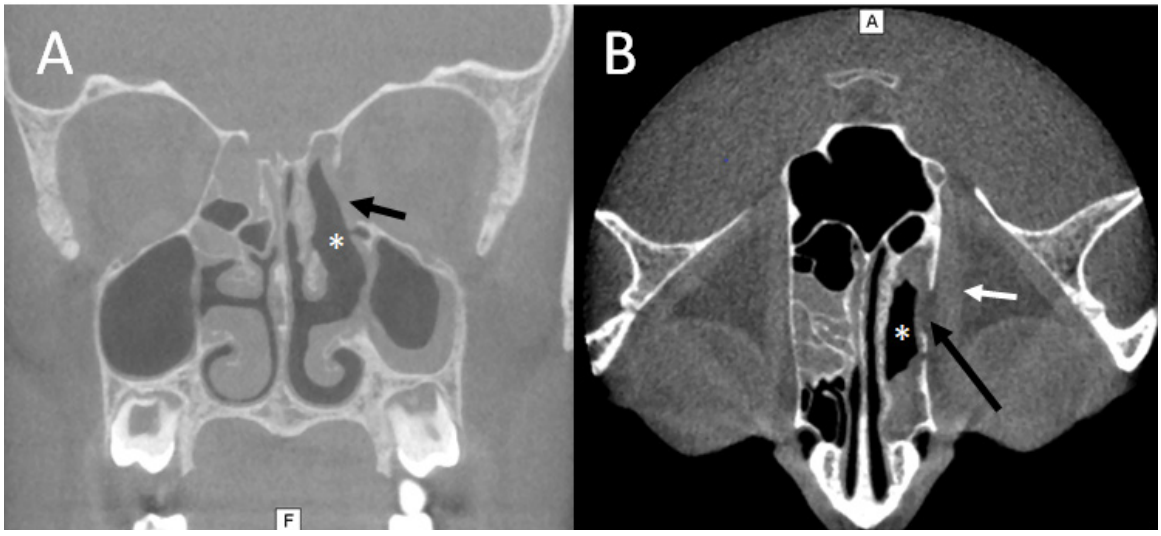

Figure 3. Post-operative coronal (A) and axial (B) CT scans. The previously mucous-filled ethmoid cavity is well-aerated (asterisk). The lamina papyracea (black arrow) and medial rectus (white arrow) have reverted to their normal anatomic position.

with ethmoid and sphenoid mucoceles, and reported that wide marsupialization was sufficient to prevent recurrence to obviate the need for complete excision. Moreover, endoscopic techniques can be used in conjunction with external approaches in cases with large and high or lateral frontal mucocele [6].

Conclusion

Mucoceles can be primary lesions or they can develop secondary to sinus pathology or trauma. Although exceedingly rare, they should be considered in the differential diagnosis of children presenting with orbital pain, proptosis, and visual disturbances. The endoscopic approach yields a high cure rate and is considered the surgical procedure of choice for the management of sinonasal mucoceles.

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