Middle turbinate primary mucocele in a child masquerading as a nasal tumour

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Abstract. Middle turbinate primary mucocele in a child masquerading as a nasal tumour. Introduction: Nasal obstruction in children is a very common complaint in daily ENT consultations. The common cold, rhinosinusitis, allergic rhinitis, hypertrophy and inflammation of the adenoid pad are very common causes of nasal obstruction, but there are numerous other causes for which the clinician must be aware. Methodology: The authors report a very unusual case of a primary mucocele of the middle turbinate of a 7-year-old girl masquerading as a nasal tumour. Results: The mucocele was successfully marsupialized surgically using an endonasal endoscopic approach. The authors report the case, imaging, and review the pertinent literature. Conclusion: Clinicians must be aware of less common aetiologies of nasal obstruction in order to orientate the diagnostic work-up and to propose adequate management.

Introduction

Nasal obstruction in children is a common complaint in daily ENT consultations. Although the common cold, rhinosinusitis, allergic rhinitis, hypertrophy, and inflammation of the adenoid pad are the most common causes of nasal obstruction in the paediatric population, there are numerous other causes. The latter can be classified into congenital, inflammatory, traumatic, iatrogenic, and neoplastic causes. The clinician must be aware of these less common aetiologies in order to orientate the diagnostic work-up and to propose adequate management.

We report a very unusual case of a primary mucocele of the middle turbinate masquerading as a nasal tumour in a child. The imaging, management and a review the pertinent literature are presented.

Case report

A 7-year-old girl was referred to the outpatient clinic with suspicion of a nasal tumour. She complained of progressive bilateral nasal obstruction, which had been present for a couple of months. She had no rhinorrhea, epistaxis, or pain. Her medical history was uneventful. She had no history of recurrent upper respiratory tract infections, nasal trauma, or sinonasal surgery. She had no pulmonary disease, and there was no known cause for any immunodeficiency.

Nasal endoscopy (Figure 1A) revealed a large lesion that completely obliterated the right nasal cavity and displaced the nasal septum to the left. A computed tomography (CT) scan showed a rounded, expansile lesion of soft tissue density involving the middle turbinate and extending to the anterior ethmoid, with no evidence of anterior skull base defect. Thin compact bone was present at the margin. Figures 1B and 1C show the lesion in the coronal and axial incidences.

Magnetic resonance imaging (MRI) (Figure 1D) clearly demonstrated the cystic nature of the lesion on T2 sequences. MRI definitively excluded a meningocele, meningo-encephalocele, or solid tumour. The diagnosis of a mucocele of the middle turbinate was then made.

Endoscopic endonasal surgery, performed under general anaesthesia, allowed a complete marsupialization of the lesion by resecting the lateral lamella of the middle turbinate. The mass was filled with mucoid secretion. Culture did not yield any pathogens. Histologic examination showed that the mucocele lining consisted of ciliary respiratory epithelium.

The postoperative period was uneventful. With a minimum 1-year follow up, the patient is still free of disease. A post-operative CT scan confirmed the
Figure 1
Nasal endoscopy; an expansile process completely fills the right nasal cavity.
Legend: (A) Nasal endoscopy; (B) CT: coronal incidence; (C) CT: axial incidence; (D) MRI: T2 sequence; (F) postoperative CT.
Primary mucocele of the middle turbinate

absence of residual pathology (Figures 1E, Figure 1F).

A septoplasty will be scheduled when the child reaches 17 years of age.

Discussion

The middle turbinate is a bony structure belonging to the ethmoid bone. It is attached anteriorly and posteriorly to the lateral wall of the nose. Superiorly, it has a vertical attachment to the skull base at the lateral border of the cribriform plate. The basal lamella of the middle turbinate separates the anterior ethmoid from the posterior ethmoid.2

Embryologically, nasal turbinates derive from a series of outgrowths from the foetal lateral nasal wall. These outgrowths form a series of ridges referred to as “ethmoturbinals”. The middle turbinate develops from the third ridge, and the superior turbinate from the fourth ridge.3

The middle turbinate may have some anatomical variations: concha bullosa (CB), paradoxically bent middle turbinate, or hypoplastic middle turbinate. CB is the most common anatomic variant of the middle turbinate. It is also called a pneumatized or aerated middle turbinate. It is absent in newborns, but its incidence increases with age. In the general adult population, its incidence varies from 14% to 53.6%, but there are conflicting reports of the prevalence of CB in the literature, with a wide discrepancy.4,5 CB is usually bilateral. When unilateral, it is often associated with a significant contralateral nasal septum deviation.6

The exact reason for middle turbinate pneumatization is still unknown.

Bolger et al classified patients with CB into three groups according to pneumatization of the middle turbinate and localization; these are lamellar, bulbous, and extensive types. The lamellar type consists of pneumatization of the vertical lamella of the turbinate. The bulbous type forms from pneumatization of the bulbous (inferior) segment, and the extensive CB is due to the pneumatization of both the lamellar and bulbous parts.

When the pneumatization is significant, it is considered to be pathogenic in two ways. First, it may directly or indirectly by pushing the uncinate process laterally block the drainage of the anterior ethmoid, frontal, or maxillary sinuses. Second, enlargement of the middle turbinate may increase the likelihood of mucosal contact between the turbinate and lateral structures. This may lead to dysfunction of the mucociliary clearance mechanism and the release of neuropeptides that cause local inflammation.3,5

From a clinical point of view, the lamellar and bulbous types are generally asymptomatic, while the extensive type is frequently associated with nasal congestion, headache, and impaired sense of smell. CB is also regarded by some authors as a factor favouring recurrent or chronic rhinosinusitis, but this continues to be debated.4,4

Mucocele of the middle turbinate was first described by Badia in 1994.9 Since then, many case reports from all over the world have been published; however, this remains an exceptional finding in children, with only a few reports in the literature.10-13

A mucocele is the most common, benign, cystic, slowly expansile, and locally destructive lesion. In the vast majority of cases, it occurs in the frontoethmoidal region. It has no connection with the nose. It contains mucus and is lined with ciliated respiratory epithelium.14 A mucocele of the middle turbinate always develops from a CB. Mucoceles can be idiopathic, the consequence of nasal trauma or surgery, or associated with a nasal tumour.

The pathogenesis of the mucoceles is unknown, but obstruction of the drainage pathway associated with chronic inflammation is generally considered a factor promoting them. Moreover, osteoclastic bone resorption provoked by release of prostaglandin (PGE2) and proinflammatory cytokines (IL-1 and TNF-α) into the wall of the mucocele has been demonstrated.14

In our case, there was no previous nasal surgery, nasal trauma, or nasal tumour. A congenital origin of this case is the most likely one.

The symptomatology of a mucocele of the middle turbinate is dominated by a progressive uni- or bilateral nasal obstruction, snoring, and headache. Rhinorrhoea is very rarely reported. Retention sinusitis may occur.11-13

When the mucocele becomes secondarily infected, it is called a mucopyocele. This situation is extremely rare.9 It can manifest as an intranasal mass, migraine headache, or be complicated by subdural empyema or orbital invasion.15-18

The diagnosis of a mucocele is based upon nasal endoscopy and imaging results. In our case, nasal endoscopy revealed a mass that completely blocked the nasal cavity and displaced the nasal septum contralaterally. The nasal mucosa looked normal.
These findings mimicked the aspect of a nasal tumour.

The differential diagnosis included a dacryocoele, meningocele, meningoencephalocele, intranasal glioma, mucocele, mesenchymatous tumour (i.e., chondro-osseus hamartoma), and osseous tumour (i.e., cemento-ossifying fibroma, fibrous dysplasia, or osteoma). A malignancy was a priori excluded because there was no necrosis, crusting, or epistaxis and no cervical lymphadenopathy.

The definitive diagnosis was made by imaging (CT and MRI). On the CT scan, a mucocele presents as a non-enhancing, homogeneous, hypodense, well-defined, rounded, expansile lesion. Typically there is rim enhancement due to inflammatory and hyperplastic mucosa. A mucocele can be locally destructive, with displacement, thinning, erosion, or dehiscence of the bony walls. CT is therefore of upmost importance when evaluating these bony changes, and to rule out orbital or intracranial extensions.19

In our case, the CT scan showed an expansile process with extension to the anterior ethmoid, with no orbital or intracranial expansion. Nevertheless, we could not differentiate between a solid or cystic lesion.

The MRI provided this information. The process gives a signal of varying intensity on both T1- and T2-weighted sequences depending on the protein concentration in the mucocele.19 In our case, there was a high, homogeneous signal on T2 sequences. The diagnosis of a mucocele of the middle turbinate was then made.

The management of such a lesion requires surgery. With advances in endonasal endoscopic surgery, the modality of the treatment has changed. In the past, completely removing the lining of the mucocele was recommended. Nowadays, in most cases surgery consists of marsupialization of the lesion. That means that the lesion is reintegrated into the upper respiratory tract (nose or sinus). The marsupialization stops the expansion of the lesion and, after a couple of months, reossification of the bony framework is observed.

Surgery on a CB can be performed in different ways; conchoplasty, crushing, resection of the lateral lamella, resection of the medial lamella, or transversal transsection.20 In our case, surgery consisted of a resection of the lateral lamella of the CB and suction of the mucoid secretion. The postoperative outcome was unremarkable.

Conclusion

Mucocele of the middle turbinate is an exceptional finding in a child. It must be included in the differential diagnosis of a mass filling the nasal cavity. Nasal endoscopy and a CT scan must be ordered. MRI may be necessary to confirm the cystic nature of the lesion. Marsupialization of the lesion, performed under general anaesthesia with endoscopic guidance, is currently the treatment of choice to stop the expansion of the lesion and to re-establish the nasal airflow. This surgery has no deleterious impact on the facial growth of the child.

References

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