The management of rare nasal mass-nasal dermoid sinus cysts: open rhinoplasty

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Abstract

The differential diagnosis of midline nasal masses includes inflammatory lesions, post-traumatic deformities, benign neoplasms, malignant neoplasms, and vascular masses. Midline congenital lesions of the nose are rare congenital anomalies. Their incidence is estimated at 1 per 20,000 to 40,000 births consisting of gliomas, encephaloceles, and nasal dermoid sinus cysts. Nasal dermoid sinus cysts account for 1-3% of dermoid cysts overall and 11-12% of head and neck dermoids. Most lesions are diagnosed within the first three years of life but in some cases the diagnosis can be prolonged. We present an 18-year old patient with nasal midline deformities, recurrent infections, airway obstruction and intracranial complications as occurring when the diagnosis has been prolonged.6

Introduction

Midline congenital lesions of the nose are a rare congenital anomaly. Their incidence is estimated at 1 per 20,000 to 40,000 births.1-3 The differential diagnosis of midline nasal masses includes inflammatory lesions, post-traumatic deformities, benign neoplasms, malignant neoplasms, and vascular masses.4,5 Gliomas, encephaloceles and nasal dermoid sinus cysts are the main part of the congenital midline lesions of the nose.4,5 The central nervous system’s (CNS) involvement in gliomas and encephaloceles is well known. Nasal dermoid sinus cyst (NDSC) has a potential for intracranial involvement. NDSC originates from ectoderm that forms from neuroectodermal and ectodermal inseparation.1 Nasal dermoid sinus cysts account for 1-3% of dermoid cysts overall and 11-12% of head and neck dermoids.2,3 NDSCs account for 61% of all midline nasal lesions in children.4,5 Early diagnosis is made in the first three years after birth in most cases. But in some cases the diagnosis may be prolonged.6 Nasal mid-

Case Report

Case #1

An 18-year old patient with chronically draining sinus opening after his birth and recurrent nasal midline infection history has been seen in our clinic. He had no other complaints. He had no specific history within his family and no maxillofacial trauma. In his examination there was widening at nasal dorsum and draining sinus opening on the skin of the nasal tip (Figure 1). In anterior rhinoscopy nasal mucous membrane was normal. In the cranial computed tomography (CCT) scan there was no intracranial extension. We performed an open technique rhinoplasty in local anesthesia. During elevation of the skin of the columna, the infratip, the tip and the supratip as a flap, we entrenched a catheter to the opening of the fistula which was located above the septum and between the two alar cartilages (Figure 1). Elevation was performed around the catheter and tract of the fistula. The fistula tract was extended to the nasion. The tract was continuing with a cyst. The skin of the opening of fistula, the tract of the fistula and the cyst were excised without rupturing. Specimen was sent for pathological examination (Figure 1). There was no problem in the

Figure 1. Draining sinus opening on the skin of nasal tip. The catheter entrenched to the opening of the fistula, which was located above the septum and between the two alar cartilages. Specimen was sent for pathological examination.
post-operative period and the patient was discharged after one day. The post-operative pathology report was epidermal inclusion cyst. The patient had no complaints in the one year follow-up and the esthetic result was satisfying.

**Case #2**

A two and a half-year old child with chronically draining sinus opening after his birth and recurrent nasal midline infection history was seen in our clinic. He also had no specific history within the family and no maxillofacial trauma. In his examination there was a 1x1 cm immobile subcutaneous mass with opening of the fistula at rhinion of the nasal dorsum. The mass was unable to be compressed and did not increase with crying. The anterior rhinoscopy was normal. A paranasal sinus computed tomography (CT) and magnetic resonance imaging (MRI) showed a cyst with no intracranial extension (Figure 2).

We performed an open technique rhinoplasty in general anesthesia (Figure 3). The tract was continuing with a cyst, which elongated cephalically between separated nasal bones and eroded bony septum. The fistula tract and the cyst were excised without rupturing. After cyst removal, the defect was filled with surgi-cell. Specimen was sent for pathological examination (Figure 3).

There was no problem in the post-operative period and the patient was discharged after three days. The post-operative pathology report was epidermal inclusion cyst. The patient had no complaints in the 6-month follow-up and the esthetic result was satisfying.

**Discussion**

NDSCs are the most frequent congenital midline lesions. The first report about NDSC was published by Bramann in 1890. There are lots of theories like sequestration, trilaminary and prenasal, about NDSCs. The most accepted theory is Pratt’s prenasal theory. Pratt described the common embriologic pathway of gliomas, encephaloceles and nasofrontal dermoid sinus tract. During the extension of dura between the unconnected bones in the skullbase to nasal region the dura is related with dermis in the nasal tip. If the bone tissue could not separate the dura from dermis during the ossification, anomalies occur.

For the congenital nasal masses, which originate from ectoderm and mesoderm, the term NDSC was first used in 1982. The diagnosis for most of the NDSCs is made in the first three years of childhood. But in some cases, like our first case, the diagnosis can be delayed until a later age. The oldest patient in the literature is a 56-year old patient with intracranial extension. There are some reports about male predominance. NDSCs are typically seen as midline masses. They usually have a sinus opening in the nasal dorsum. Intermittent secretion of sebaceous material and recurrent infections are seen frequently. The hair outgoing from the opening is pathognomonic for the NDSC but is found in less than half of the patients. NDSC is seen sporadically but familial cases have been reported in the literature.

There is no association of a syndrome with the formation of the NDSC. There are other congenital anomalies that have been reported. These include craniofacial anomalies, hypertelorism, cleft palate, hemifacial microsomia, aural atresia, pinna deformities, branchial sinus anomalies, cardiac, genital and gastrointestinal anomalies. These associations have been reported with different ratios in the literature. There is no proved described genetic transmission. There is some familial transmission reported in the literature. NDSCs make up 11-12% of all head and neck dermoids. They can be seen between glabella and columella. The ratio of the intracranial extension is controversial. Suspicion of intracranial extension is important for every patient with NDSC. Cranial CT and/or MRI are essential to determine the extension. Cranial CT is valuable to show the bone alterations and help diagnosis. Disadvantages of CCT are expectation of ionized radiation and interpretation problems because of the unseparated crista galli and perpendicular plate of the ethmoid bone in infants under one year old. MRI has high resolution in soft tissue so it clearly exhibits the intracranial extension.

The treatment of the NDSC is surgical excision. The most favorable technique is the open rhinoplasty like in our two cases.
The reasons for choosing this technique are exposure, good esthetic results and allowing the reconstruction of the nasal dorsum. The formation of the surgery depends on the lesions’ localization and extension.

With the clinical findings and imaging modalities, we can estimate the intracranial extension and plan appropriate surgical treatment. In most of the cases an early diagnosis is made in the first three years after birth. In some cases, however, the diagnosis may be prolonged. This must be kept in mind for differential diagnosis. The treatment is surgical. The investigation for the intracranial extension must be performed before the surgery. With the open septorhinoplasty technique we can achieve good esthetic and functional results.

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