Case Report

Endoscopic Treatment for Nasal Dermoid Sinus Cyst: A Report of 6 Pediatric Cases

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Abstract

Nasal dermoid sinus cysts are the most common congenital midline nasal lesion. The frequency of intracranial extensions varies from 5% to 45%. Complete surgical excision of nasal dermoid cyst and any associated sinus tract is essential for cure and any residual ectodermal elements result in a high rate of recurrence and complicated infections.

Many different approaches have been described for the removal of nasal dermoids, ranging from a simple extracranial excision to complex procedures in which a combined extracranial-intracranial approach is required.

We hereby report six cases of nasal dermoid sinus cysts in children (5 with intracranial extension) which were managed with an endonasal endoscopic procedure. We describe the technique we implemented for this procedure and for the reconstruction of the skull base defect.

ABBREVIATIONS

NDSC: Nasal Dermoid Sinus Cyst; CSF: Cerebrospinal Fluid

INTRODUCTION

Nasal dermoid sinus cysts (NDSCs) are the most common congenital midline nasal lesions [1,2], accounting for over 60% of all the midline nasal lesions in children [3], for 1–3% of all dermoid cysts, and for 4–12% of head and neck dermoids [4–7].

Midline congenital lesions of the nose, consisting of nasal dermoid sinus cysts, glioma and encephaloceles are rare congenital anomalies. Their incidence is estimated at 1:20.000 to 1:40.000 births [8,9]. Inflammatory lesions, post-traumatic deformities, benign neoplasms, malignant neoplasms, congenital and vascular masses have to be excluded [7].

Sessions [10] was the first to coin the expression nasal dermoid sinus cyst to include all the lesions in the nose containing ectoderm (stratified squamous epithelium) and mesoderm (adnexal structure).

Its pathogenesis involves the incomplete obliteration of neuroectoderm in the developing frontonasal region [9].

Nasal dermoid is usually visible at birth or in early infancy and the mean age of presentation varies from 14 to 34 months, with a slight male predominance [2,10–12]. A familial basis has been suggested [3,13]. Nasal dermoid typically presents as a midline mass, most commonly along the dorsum from the naso-glabellar region to the base of the columella, and may be associated with a sinus opening. Intermittent discharge of sebaceous material and recurrent infection are also common. Even if it is present in less than half of patients, hair protruding through a punctum is pathognomonic for nasal dermoid, [14].

The frequency of associated multiple anomalies (such as aural atresia, deformity of the pinna, mental retardation, craniosynostosis, lacrimal duct cysts, hydrocephalus, pharyngeal arch sinus, hypertelorism, hemifacial microsomia and cleft/lip palate) greatly varies in literature, ranging from 6% to 41%, [2,4,10,11,15].

In literature, the frequency of intracranial extensions varies from 5% to 45% [4,10,15–18]. Several degrees of extensions are distinguished. The great majority remain extradural. Intradural extensions with involvement of brain parenchyma has also been reported [1,19–23]. Computed tomography (CT) and magnetic resonance imaging (MRI) scans are the gold standards in the radiographic evaluation of nasal dermoid, and each modality offers complementary information for the diagnosis and the determination of the extent of the lesion, as well as allows pre-surgical planning.

Progressive enlargement of a nasal dermoid can cause soft tissue and skeletal deformity, local infection, meningitis, and brain abscess. Timely diagnosis is, therefore, essential, and surgical excision is the only therapeutic modality.
Many different approaches have been described for the removal of nasal dermoids in the past two decades, ranging from a simple extracranial excision to complex procedures in which a combined extracranial–intracranial approach is required. In this paper, we report six cases of NDSCs in children that were managed with an endonasal endoscopic procedure. We describe the technique we implemented for this procedure and for the reconstruction of the skull base defect.

**CASE PRESENTATION**

From September 2007 until January 2015 we performed 6 endonasal endoscopic procedures in pediatric patients. The mean age was 5.8 years (range from 2 to 16 years), two patients were female and four male.

All the cases came to our attention with a history of nasal midline recurrent infections often with intermittent discharge. Physical examination revealed in all patients a draining sinus opening on the nasal dorsum with a protruding hair. 2 patients presented a cutaneous fistula Figure (1a) and a cutaneous abscess Figure (1d) in the right and left paranasal area respectively. 3 children had an intense swelling glabellar extended to the frontal region Figure (1b–e).

The other head and neck findings were within normal limits.

All the patients underwent CT scan and MRI with gadolinium that shown in 5 of 6 cases intracranial extradural extension. No intradural extention was reported. A fistulous tract extending from the cyst out to the nasal dorsum was detected in all the cases. Details of patients and their presentation are included in (Table 1). Our surgical technique is described below. The patient is positioned, under general anesthesia, in a semi recumbent position with head extended. A nasal decongestion with 5% Xylocaine (R) is performed. We proceed, after subperichondrium dissection, to endonasal endoscopic resection of the antero-superior portion of the nasal septum which also includes the upper portion of the fistula in order to increase the surgical maneuverability through a bilateral nasal access.

![Figure 1 Pre-operative images showing the typical midline or paramedian mass with sinus opening in nasal dorsum in our 6 cases.](image)

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<td>Sinus Infection, Discharge</td>
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**Table 1: Patient Demographics, Presentation and Investigation.**

**Abbreviations:** Pt: Patient; F: Female; M: Male; CT: Computer Tomography; MRI: Magnetic Resonance Imaging

Considering the limited anatomical space, normally we proceed by emptying the cyst contents, trying to preserve the integrity of the walls. The next step concerns the subperiosteal dissection of the cyst walls from the surrounding bony structures, generally within the crista galli, until the superior insertion of the cyst (intracranial extradural) in the falx cerebri is identified. The cyst may be so toughly adherent to the dural plan, that, is required the use of cutting instruments with unavoidable CSF leak.

Once the cyst is radically removed, it is necessary to proceed to the second part of the surgery, the removal of the fistulous portion on the nasal dorsum.

To do that, a perilesional elliptic incision is carried out by deeply following the fistulous tract penetrating the nasal bones. Under endoscopic control occurs the actual removal of the fistulous intranasal tract.

In order to reduce granulation and, when necessary, to perform dural plasty, after regularization of perilesional bone margins, mucoperiosteum from the resected portion of nasal septum is taken and positioned on the exposed bone and dural surface. Cartilage from the removed portion of the septum can also be used to close the skull base defect (Figure 2). Absorbable nasal packing kept in place for 1 day in the patient without CSF leak and 2–3 days in the other cases.

We achieved the radical remove of cyst and fistulous portion in all the cases. All the lesions were extradural, but in 2 cases the lesions were adherent to the dura and we had to remove part of the dura itself; the defect was then successfully repaired with a free, nasal septal mucoperichondrial flap alone or with septal cartilage. A post operative CT scan was performed in all the cases to control the presence of any residual and the closure of the skull base defect (Figure 3).

No complications occurred during and after surgery and, particularly, no post operative CSF leak occurred.

Nowadays (minimum follow up of 18 months) no recurrence of NDSC is reported. In only one case (case no.6) we had to perform 6 months after the first surgery, in local anesthesia, an external removal of a second fistulous tract that wasn’t evident at the time of the previous surgery (Table 2).
DISCUSSION

Using a transnasal endoscopic approach the morbidity associated with frontal craniotomy is minimized or eliminated when open excision is avoided. The transnasal approach does not require frontal lobe retraction and the non-invasive nature of dermoid cysts makes the plane of dissection easy to manipulate, therefore, a careful piecemeal resection of the tumor while preserving the underlying neural and vascular structures should be the main strategy for the operation. The patients had excellent postoperative recovery and a shorter hospital stay. All patients with NDSCs must be considered as potentially having an intracranial extension and, therefore, undergo a thorough preoperative radiological evaluation [25].

Workup has to include fine-cut CT, and complementary MRI should be considered if there is concern for intracranial extension.

Some authors considered that MRI alone is the most cost-effective and accurate means of evaluating nasal dermoids and it is essential for preoperative planning [24].

In conclusion we can state that, in expert hands, the endoscopic endonasal approach for dermoids that extend to the anterior cranial fossa is an effective technique for the resection of nasal dermoids with intracranial extension. The endoscopic transnasal skull base approach is an excellent alternative to traditional frontal craniotomy in order to achieve the complete resection of intracranial dermoid cysts.

The use of a team-based approach, including an endoscopic otolaryngologist and a neurosurgeon in case of intradural extension, is crucial to the success of the transnasal endoscopic technique. The aim of the endoscopic removal is to eliminate the increased morbidity associated with frontal craniotomy. Such endoscopic removal has minimal morbidity and mortality, and better aesthetic results compared with open craniotomy, but it still achieves the complete resection of the tumour.

REFERENCES