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Surgical management of nasal dermoid lesions

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Nasal dermoids are the most common congenital midline nasal lesions. They often present as a midline pit or nasal mass and arise from incomplete or failed regression of the dural diverticulum during embryogenesis. Management requires complete resection of the tract and an appropriate surgical approach is based on location, extent, and presence and/or degree of intracranial extension. Classically requiring an open craniotomy for resection, newer strategies employ a minimally disruptive approach to fully excise the lesion and the tract to prevent recurrence. This article presents details on embryology, imaging work up, and surgical technique for resection of these lesions either via an open approach or with an endoscopic-assisted method.

Introduction

The differential diagnosis for midline nasal masses includes congenital midline embryologic remnants such as glioma, encephalocele, and nasal dermoid, as well as other less common benign masses including epidermoid cyst, hemangioma, teratoma, neurofibroma, arteriovenous malformation, lipoma, and lymphatic or venous malformations. Congenital midline masses are rare occurring in 1:20,000-1:40,000 live births.[1,2] Nasal dermoid lesions are the most common, accounting for the majority of midline congenital lesions in the midface,[3] and are embryologically derived frontonasal inclusion cysts or tracts. They were first described by Cruvelier in 1817.[4] The degree of symptoms may vary from a small non-bothersome mass to a draining or infected mass. In all cases, surgical excision is indicated through a variety of surgical techniques in order to prevent local infection.

Epidemiology

Nasal dermoid lesions comprise 1% of dermoid cysts found throughout the body.[5] Dermoid cysts are present at birth, but only 70% are diagnosed before the age of 5 years.[6] They tend to grow slowly for the first few years of life. The majority occur sporadically with a reported male preponderance.[7] Although they typically occur as isolated anomalies, 5%-41% occur with associated anomalies, such as aural atresia, developmental delay, hydrocephalus, branchial arch anomalies, cleft lip and palate, or hemifacial microsomia.[7]

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Embryology and presentation

Nasal dermoid lesions usually present as a midline pit, in the majority of cases, or a nasal mass. They derive from embryologic structures that persist in the setting of trapped rests of cells or incomplete fusion planes at the anterior neuropore. Embryogenesis of the midface occurs between the 4th and 8th week of gestation. At the end of this process, the foramen cecum forms a defect in the anterior skull base at the apex of what becomes the prenasal space, which is a potential space located between the nasal bones and the nasal and septal cartilages. The prenasal space fuses with the fonticulus frontalis, which is a fontanelle between the inferior aspect of the frontal bones and the developing nasal bones. During embryogenesis, a protrusion of dura extends from the anterior cranial fossa through the foramen cecum, traversing the prenasal space, and apposes ectoderm at the site of the future rhinion. This dural diverticulum extends toward (and contacts) the skin of the nasal tip.[8] The frontal and nasal bones fuse and the dural diverticulum involutes.

Incomplete or failed regression of the dural diverticulum at any point from the nasal tip to the cranium results in a nasal dermoid cyst, sinus, or tract. The apposition and irrevocable interdigitation of the dura with the skin may pull the ectoderm posterosuperiorly, toward the foramen cecum. In some cases, the ectoderm can extend through the foramen cecum, resulting in a tract from nasal tip to dura. Proliferation of the trapped elements produces the findings typically seen in the lesions, including glands and hair. Although the nasal dermoid can be extranasal, intranasal, intracranial, or a combination, based on the site of embryologic arrest or incomplete involution, the most common location of the lesions is near the superficial nasal tip. When present, the intracranial communication is classically through the foramen cecum to the anterior skull base with extradural adherence to the falx cerebri.

Diagnosis

Clinical findings

Nasal dermoid lesions are present at birth or in early childhood as a midline nasal mass. Sixty percent are located on the lower nasal dorsum resulting in protrusion and/or widening of the nose.[9] The lower lateral nasal cartilages can be splayed laterally by the mass. Nasal dermoid lesions are typically midline and may be found at any point from the columella to the glabella.

Nasal dermoid lesions are firm, noncompressible masses that do not transilluminate, differentiating them from encephaloceles or gliomas. Additionally, unlike encephaloceles, nasal dermoid lesions do not enlarge with crying, straining, or occlusion of the internal jugular vein. Occlusion of the internal jugular veins resulting in enlargement of the mass is typical of an encephalocele (positive Furstenberg test). Dermoid lesions typically end in a subcutaneous hair tract with a pit or single hair at the skin opening. Hair arising from the pit is pathognomic for a nasal dermoid lesion. If more distal on the nasal dorsum, such as on the tip or columella, there is typically a single pit. Lesions more cranial or those with a long tract, may demonstrate multiple pits along the dorsum or a combination of a midline pit and small pits at the bilateral medial canthi (Figure 1). Presentation of nasal dermoids associated with an infected cyst may lead to purulent discharge. Resection of the lesions is important, as infectious complications can lead to recurrent abscesses, osteomyelitis, and potentially meningitis or brain abscess if intracranial extension is present.

Figure 1 Photograph of patient presenting with nasal dermoid with a midline pit as well as smaller pits near bilateral medial canthi. Recent purulent drainage was present from the right sided medial canthus skin opening. (Color version of figure is available online.)

The clinical presentation of nasal dermoids drives contemporary surgical decision making, although imaging is essential for any suspected dermoid prior to operative intervention. Our clinical experience has identified 4 variations in dermoid presentation:

(1) An isolated pit, with no associated mass, located at the bony-cartilaginous junction. This is often associated with a tuft a hair emanating from the pit.
(2) A protrusion at the nasal tip with pit present along nasal dorsum between the bony cartilaginous junction and nasal tip.
(3) A midline pit with or without a mass, as well as pits overlying the lateral nasal bones at the level of the medial canthi.
(4) Any of the above with large intracranial component ("Dumbbell lesion").

Imaging

Preoperative imaging is essential to characterize the lesion, and more importantly, the presence and degree of intracranial extension. Computed tomography (CT) and

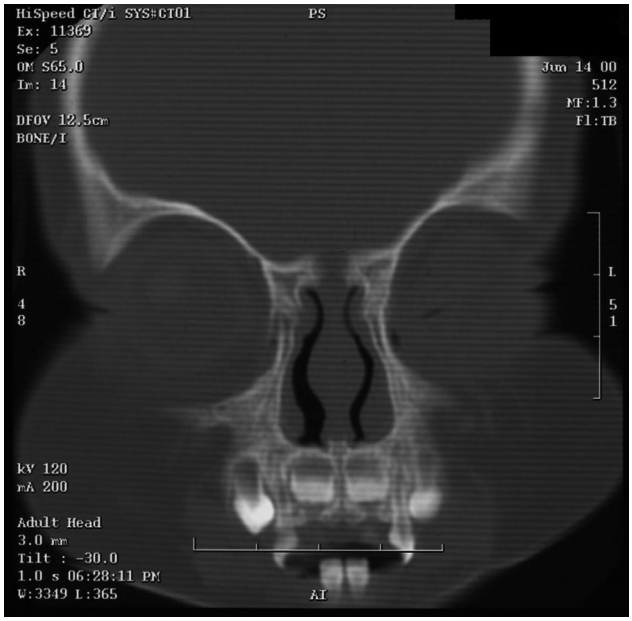

Figure 2 Coronal cross sectional CT image demonstrating midline anterior skull base defect in a patient with nasal dermoid cyst with intracranial extension. (Color version of figure is available online.)

magnetic resonance imaging (MRI) provide complementary information. A thin-slice CT scan of the facial skeleton with contrast is important to differentiate the dermoid from the surrounding nasal mucosa and identify bony defects in the anterior skull base (Figure 2). An enlarged foramen cecum or bifid crista galli are suggestive—but not specific—for intracranial involvement. Pensler and Bauer suggested that presence of fibrous tissue connecting the dermoid cyst to the dura may lead to an enlarged foramen cecum or bifid crista galli without true intracranial involvement.[10] If a normal appearing crista galli and foramen cecum is identified, intracranial extension can be ruled out.

Multiplanar, fine-section MRI with gadolinium is helpful to depict the soft tissue anatomy and to differentiate the nonenhancing dermoid from enhancing lesions such as teratoma and hemangioma. High signal attenuation on T1-weighted images is suggestive of intracranial extension (Figure 3). As the age of presentation of patients with nasal dermoids is quite young, sedation is typically required to obtain adequate MRI imaging. In our experience, surgical decision making is presented to the family prior to obtaining an MRI due to the low probability of significant intracranial extension. Operative time is reserved and the patient is sedated for the imaging study, if no significant intracranial component is present, surgical resection is carried out while under the same anesthetic after a brief discussion with the family, detailing the results of the findings. Preoperative, as well as preimaging, discussions on possible surgical intervention based on MRI findings, as well as a presentation of the risk and benefits allows for more efficient patient care and improved use of resources.

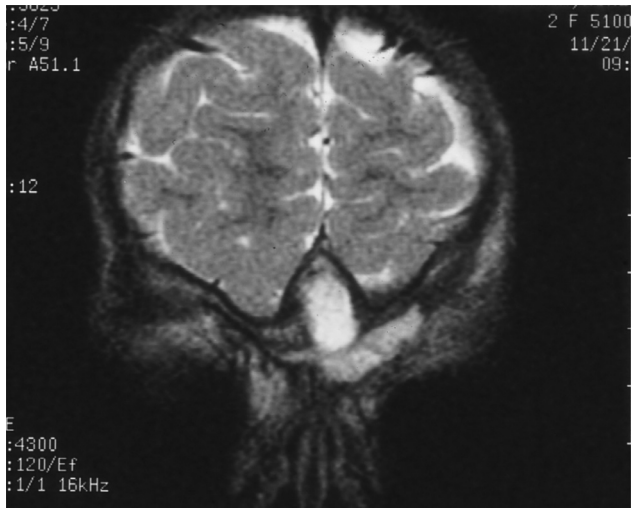

Figure 3 Coronal cross sectional T1-weighted MRI with high signal attenuation intracranially of a nasal dermoid cyst. (Color version of figure is available online.)

Further discussion of various surgical techniques and options is presented below.

In summary, CT and MR are complementary imaging modalities in the workup and surgical planning for nasal dermoid lesions.

Pathologic findings

Histologically, nasal dermoids contain ectodermal and mesodermal, but not endodermal, embryonic elements, such as hair follicles, sebaceous glands, and apocrine glands. When present in the cell wall of the cyst, these findings differentiate it from a simple epidermoid cyst. Keratin debris is prominent on histopathological analysis with an absence of glial features that are present in encephaloceles and gliomas.

Surgical approaches and techniques

Open frontal craniotomy

Surgical options for resection of nasal dermoids are based on location, extent, and presence and/or degree of intracranial extension. The goal of surgery, regardless of technique employed, is for complete resection of the cyst and its tract. If incomplete resection is performed, recurrence is nearly certain.[11] Traditionally, nasal dermoids were resected via an open frontal craniotomy using a coronal incision (Figure 4). This technique is still necessary when a large intracranial portion or dural attachment is present, although this is rare. Craniotomy adds morbidity compared to endoscopic approaches due to the required brain retraction and high risk of olfactory nerve damage. If a pit or prenasal portion is present, a combined approach is necessary to fully resect the lesion and provide the optical aesthetic outcome. Nonetheless, this technique is usually

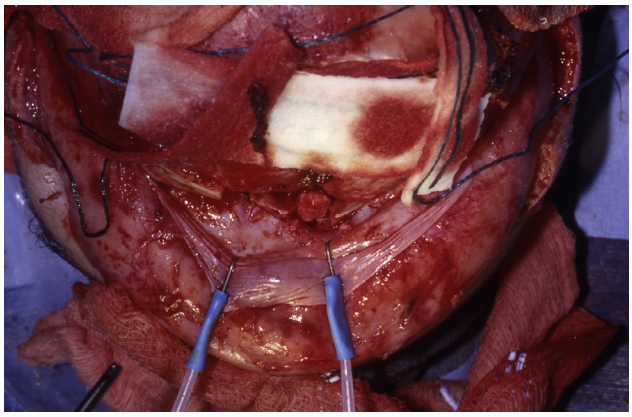

Figure 4 Intraoperative photograph of frontal craniotomy in removal of nasal dermoid cyst with significant intracranial extension. (Color version of figure is available online.)

sufficient to guarantee adequate resection of extranasal, intransal, and intracranial lesions. Disadvantages of the technique include a large skin incision, alopecia around the incision line, and risks associated with brain retraction and olfactory nerve damage.

Rhinoplasty and direct excision through nasal dorsum incision

When a lesion is isolated to the nasal dorsum without a significant tract or obvious pit, an open rhinoplasty technique alone can be employed to resect the mass. When a pit is present, it is typically found at or just below the bony cartilaginous junction (Figure 5). Since the pit is relatively high and difficult to access with an isolated open rhinoplasty approach, and since one must excise the pit anyway, multiple approaches are usually necessary to completely resect the lesion. For removal of a nasal tip mass, a mid-columellar incision is made and the skin is elevated off the lower lateral cartilages and up until dissection is carried above the lesion. After ensuring there is no tract leading up toward the foramen cecum, the mass is resected after circumferential dissection is completed (Figure 5). When a pit is present, the mass can be removed either via a mid-columellar incision, as well as an elliptical incision surrounding the pit and dissection can be carried out to circumferentially outside the epithelial matrix to remove the mass and the pit en bloc. This dissection is performed using operative loupes or a microscope, as well as fine instruments, (ie, otomicroscopic ear instruments), such as duckbills and a McCabe flap knife. The incision for the elliptical incision should be oriented in a craniocaudal vector to allow most acceptable aesthetic outcomes.

In situations in which multiple pits are present, typically in the region of the lateral nasal bones, elliptical incisions around the pits are used to resect this component (Figure 6). Closure of the incision should be performed in a horizontal fashion using a broken line technique to prevent tethering of the medial canthi and lid deformity. If a tract is present extending toward the skull base

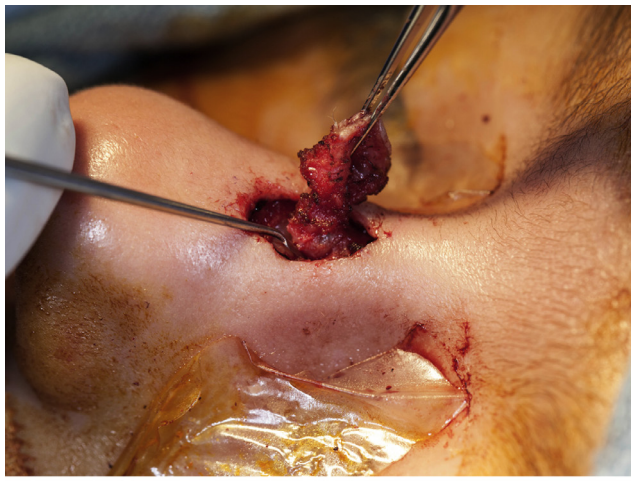

Figure 5 and 6 Intraoperative photographs of nasal dermoid cyst being excised through a direct elliptical incision in which the involved skin and cyst are removed en bloc. (Color version of figure is available online.)

requiring open dissection, osteotomy, or lateral retraction of the nasal bones is performed to allow for optimal exposure. Patients undergoing resection of nasal dermoids are typically very young, and thus the nasal bones are incompletely fused, making separation straightforward. After resection, the separated nasal bones can be medialized to their proper position. Splinting is useful to ensure appropriate healing.

Endoscopic-assisted rhinoplasty

In situations where a tract extends up to the foramen cecum, an open rhinoplasty technique is used to resect the nasal portion (Figure 7). Once the prenasal space is exposed, a narrow rigid endoscope is employed to further follow the tract up to the foramen cecum with magnification (Figure 8). Dissection is carried out until the cyst and its tract are completely removed. When minimal intracranial extension without intradural involvement is present, the dermoid can still be removed in this fashion. Often, a

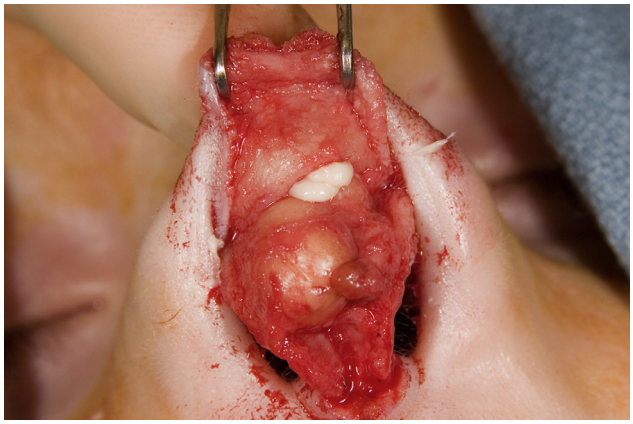

Figure 7 Intraoperative photograph of rhinoplasty approach to remove nasal dermoid cyst without significant intracranial involvement. The cyst is located just anterior to lower lateral cartilages in the midline and there is a small amount of cyst material draining from the cyst cavity's anterior wall. (Color version of figure is available online.)

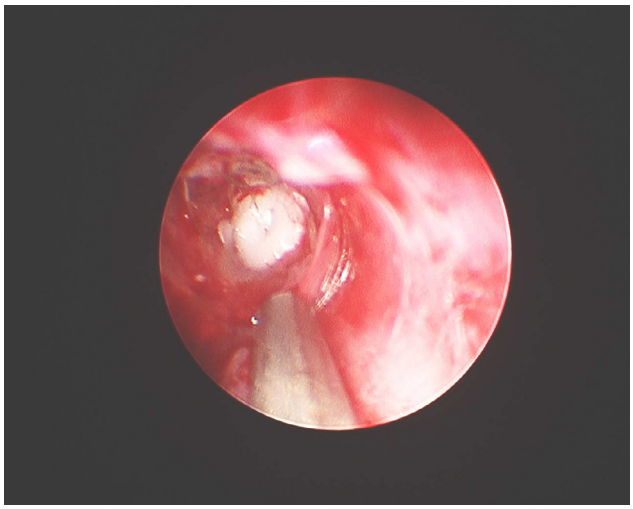

Figure 8 Rigid endoscopic image of cyst resection at the skull base through a rhinoplasty (columellar incision) approach. (Color version of figure is available online.)

small fibrous stalk extends to the dura. If a cerebrospinal fluid (CSF) leak is encountered, a small piece of temporalis fascia can be used to plug the skull base defect. This alone is typically sufficient to prevent further CSF leakage, especially with closure and healing of the potential prenasal space.

Transnasal

For nasal dermoids with intranasal components without skin involvement, endoscopic techniques can be utilized.[7,12,13] Preoperative evaluation of imaging is essential for identifying the site of skull base involvement. If a CSF leak is encountered, repair is carried out based on the size and rate of flow. Options for repair consist of use of autologous or allografts. Blood supply and mucosa for a nasoseptal flap should be diligently preserved and protected for use as necessary.

Postoperative management and care

When noncraniotomy techniques are used and when there is no significant intracranial extension, patients are able to discharge home with routine wound care. Overnight admission for appropriate pain control and monitoring is reasonable for the very young patient. If CSF leak was encountered, appropriate monitoring is necessary depending on the size of the skull base defect. We typically do not use intranasal splints or packing, but an external nasal splint can be used to protect the nasal dorsum if the nasal boned required significant mobilization. In the very young patient, we typically use absorbable sutures to prevent need for removal. For patients with a significant mass effect from the dermoid (ie, a cystic component), it is important to counsel the family that the protrusion will become a depression postoperatively. In our experience, we typically have not placed alloderm or fat to fill the paucity, as the contour will usually normalize with growth and development. Moisturizing ointment can be used on all external incisions to aid in healing. Patients are typically seen at 1 week postoperation to evaluate healing and remove any external splints or wound dressings. If complete resection is accomplished and the patient remains asymptomatic without signs or symptoms of recurrence, no routine follow-up is necessary in long term.

Conclusion

Nasal dermoids are rare congenital lesions. The goal of management is complete resection to prevent further infections or growth is the most cosmetically appealing way possible. We describe various options for surgical management based on location and intracranial extension. The majority of lesions can be addressed without the need for an open craniotomy using a variety of rhinoplasty and endoscopic-assisted rhinoplasty surgical approaches.

Disclosures

The authors reported no proprietary or commercial interest in any product mentioned or concept discussed in this article.

References

1. Pratt LW: Midline cysts of the nasal dorsum: Embryologic origin and treatment. Laryngoscope 75:968–980, 1965.
2. Hughes GB, Sharpino G, Hunt W, et al: Management of the congenital midline nasal mass: A review. Head Neck Surg 2:222–233, 1980.

3. Rohrich RJ, Lowe JB, Schwartz MR: The role of open rhinoplasty in the management of nasal dermoid cysts. Plast Reconstr Surg 104:1459–1466, 1999 quiz 1467; discussion 1468.
4. Sessions RB: Nasal dermal sinuses–new concepts and explanations. Laryngoscope 92:1–28, 1982.
5. Crawford JK, Webster JP: Congenital dermoid cysts of the nose. Plast Reconstr Surg (1946) 9:235–260, 1952.
6. Pollard ZF, Harley RD, Calhoun J: Dermoid cysts in children. Pediatrics 57:379–382, 1976.
7. Rahbar R, Shah P, Mulliken JB, et al: The presentation and management of nasal dermoid: A 30-year experience. Arch Otolaryngol Head Neck Surg 129:464–471, 2003.
8. Pinheiro-Neto CD, Snyderman CH, Fernandez-Miranda J, et al: Endoscopic endonasal surgery for nasal dermoids. Otolaryngol Clin North Am 44:981–987, 2011 ix.
9. Szeremeta W, Parikh TD, Widelitz JS: Congenital nasal malformations. Otolaryngol Clin North Am 40:97–112, 2007 vi-vii.
10. Pensler JM, Bauer BS, Naidich TP: Craniofacial dermoids. Plast Reconstr Surg 82:953–958, 1988.
11. Posnick JC, Bortoluzzi P, Armstrong DC, et al: Intracranial nasal dermoid sinus cysts: Computed tomographic scan findings and surgical results. Plast Reconstr Surg 93:745–754, 1994 discussion 755-746.
12. Weiss DD, Robson CD, Mulliken JB: Transnasal endoscopic excision of midline nasal dermoid from the anterior cranial base. Plast Reconstr Surg 102:2119–2123, 1998.
13. Schuster D, Riley KO, Cure JK, et al: Endoscopic resection of intracranial dermoid cysts. J Laryngol Otol 125:423–427, 2011.