Midline Approach to Pediatric Nasofrontal Dermoid Cysts

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IMPORTANCE To highlight the advantages of the vertical midline incision in providing satisfactory cosmesis with complete excision of pediatric nasofrontal dermoid cysts.

OBSERVATIONS Retrospective case series of nasofrontal dermoid cysts in 4 patients treated at a single tertiary medical center from June 1, 2010, through July 31, 2012. The mean age at surgery was 2.5 years. The anatomical location of the nasofrontal dermoid cysts differed: (1) supratip extending through the upper lateral cartilages to the cartilaginous septum, (2) upper dorsum and subcutaneous tissue, (3) tip and supratip extending deep to the nasal bones with involvement of the anterior cranial fossae and dura, and (4) nasal tip extending deep to the level of the rhinion and involving the upper lateral cartilages and below the left medial canthus. Preoperative imaging was performed on all patients. There was one case of intracranial extension. All patients underwent surgical excision with the vertical midline incision. Nasal reconstruction was performed with local soft-tissue flaps (1 patient), regenerative tissue matrix (2 patients), and bone dust pate (1 patient). The patient with intracranial involvement also underwent a frontal craniotomy. All lesions were histologically confirmed as dermoid cysts. Mean follow-up was 1.5 years. There were no complications or recurrences. All patients had cosmetically acceptable scars.

CONCLUSIONS AND RELEVANCE A vertical midline incision with modifications to excise involved skin provides a satisfactory and cosmetically sound approach to congenital lesions of the nasofrontal region. It affords adequate exposure for complete excision and reconstruction. A multidisciplinary team consisting of a neurosurgeon, facial plastic surgeon, and pediatric otolaryngologist is needed to optimize outcomes.

C ongenital midline nasal lesions are rare and have been reported to occur in approximately 1 in 30 000 births.\textsuperscript{1,2} The differential diagnosis is vast and includes encephaloceles, gliomas, hemangiomas, and nasofrontal dermoid cysts. Nasofrontal dermoid cysts account for approximately 60\% of all congenital midline nasal lesions and up to 12\% of all head and neck dermoid cysts.\textsuperscript{3-4} Currently, there are no predictable patterns of occurrence. Nasofrontal dermoid cysts are true cysts that contain keratinizing squamous epithelium and adnexal structures, such as hair follicles, sebaceous glands, and sweat glands. The anomalies are thought to arise from incomplete separation of neuroectodermal and ectodermal tissue during development of the anterior skull base at approximately 8\ weeks’ gestation. They usually present at a young age as a mass along the nasal dorsum or glabella. Up to 50\% of these lesions have gross cutaneous involvement and 20\% may extend intracranially.\textsuperscript{3} They characteristically lack transillumination and fail to enlarge with the Valsalva maneuver. Nasofrontal dermoid cysts are benign in nature but are susceptible to recurrent infections and may progress to osteomyelitis, meningitis, or intracranial abscess. Moreover, they can cause significant disfigurement and warrant prompt evaluation. The recommended treatment is complete surgical excision. Many surgical approaches have been described, including midline vertical incision, open rhinoplasty, Lynch incision, gullwing incision, coronal incision, and endoscopic techniques. The objective of our study was to compare our experience with the midline vertical incision in achieving these goals with other surgical approaches.

Complete neurologic evaluation and high-resolution, multiplanar imaging modalities (computed tomography [CT] and magnetic resonance imaging [MRI]) are required for preoperative planning and to rule out intracranial extension. Suggestive findings may include defects in the anterior skull base, widening of the foramen cecum or cribriform plate, and bifid nasal septum and crista galli.

While planning the surgical strategy, surgeons must consider the location and extent of the lesion because deeper lesions may require more invasive techniques. Coronal incisions and frontal craniotomy may also be required for lesions that extend intracranially. Adequate exposure must be achieved for excision and surgical exploration to reduce the risk of recurrence, which has been reported in 50\% to 100\% of cases if resection is incomplete.\textsuperscript{3} Obtaining acceptable cosmesis, particularly in pediatric patients, adds to the complexity of treating this pathologic process.
Methods

A retrospective review of the medical records of 4 pediatric patients who presented with nasofrontal dermoid cysts to a single, tertiary institution from June 1, 2010, through July 31, 2012, was performed. University of Maryland Baltimore Institutional Review Board exemption was obtained for this report. Informed consent was obtained in writing for all surgical procedures and publication of photographs. All 4 patients had a nasofrontal dermoid cyst, 1 with intracranial extension. All patients underwent preoperative imaging. Three patients underwent both CT and MRI, whereas 1 patient underwent MRI alone. All were treated through a vertical midline approach with complete excision followed by local reconstruction. A coronal incision with frontal craniotomy was used in the patient with intracranial extension. All patients received postoperative care and follow-up by their primary surgeon.

Results

We studied 4 patients (2 boys and 2 girls) who presented with a midline nasal mass with cutaneous involvement. The mean age at presentation was 1 year. The mean age at surgery was 2.5 years. The anatomical location of the nasofrontal dermoid cysts differed in each patient: (1) supratip extending through the upper lateral cartilages to the cartilaginous septum, (2) upper dorsum and subcutaneous tissue, (3) tip and supratip extending deep to the nasal bones with involvement of the anterior cranial fossae and dura, and (4) nasal tip extending deep to the level of the rhinion and involving the upper lateral cartilages and below the left medial canthus. All patients underwent MRI, which confirmed a hyperintense lesion on T1- and hypointensity on T2-weighted imaging. In addition, CT revealed an enlarged foramen cecum in the case of intracranial extension (Figure 1A). All 4 patients underwent surgical removal of the lesions with excision of involved skin that used the vertical midline incision. Nasal reconstruction was performed with local soft-tissue advancement flaps (1 patient), regenerative tissue matrix (Alloderm) (2 patients), and bone dust pate (1 patient). The patient with intracranial involvement had an infected lesion on the nasal dorsum (Figure 2). Exposure of the entire lesion was obtained through the midline nasal incision (Figure 3) and a frontal craniotomy. All 4 lesions were histologically confirmed as dermoid cysts. Mean follow-up was 1.5 years. There were no complications or recurrences. All patients had a cosmetically acceptable incisional scar (Figure 4).

Discussion

The current standard of treatment for pediatric nasofrontal dermoid cysts is early, complete surgical resection with the primary goal of preventing complications, structural distortion, and recurrence. These masses have the potential to be very complex in nature, and approximately 20% of cases will have intracranial extension. Initial diagnostic testing should be performed with a high suspicion for intracranial involvement. High-resolution, multiplanar imaging is therefore recommended. Positive predictive values in CT and MRI are 85.7% and 100%, respectively. Negative predictive values have been
estimated at 50% for CT and MRI. The preferred first-line study of choice is MRI because of its high soft-tissue resolution, lack of radiation exposure, and cost-effectiveness.

The optimal treatment plan will ultimately depend on several factors, including size and location of the lesion, integrity of nasofrontal anatomy, and presence of intracranial extension. Pollock states that surgical techniques should adhere to 4 main principles: they should allow exposure to the mass and associated sinus tract, they should allow access to the skull base, they should provide exposure for reconstruction of the nasal dorsum, and they should result in a cosmetically acceptable scar. Failure to adhere to these principles may result in unsatisfactory cosmetic and functional outcomes. Furthermore, recurrence has been estimated at 50% to 100% if resection is incomplete.

Historically, the vertical midline approach has been widely supported in the literature and was the most commonly used approach, yielding satisfactory results with minimal recurrence. Winterton and colleagues described a retrospective study of 19 patients with pathologically confirmed nasal dermoid cysts at various locations from the glabella to the nasal tip. Eighteen patients underwent surgical excision with the vertical midline elliptical excision with or without a frontal craniotomy, whereas one patient underwent an external rhinoplasty approach. The authors noted adequate surgical scar cosmesis at a mean follow-up of 4.1 years, although this was not objectified. Five documented superficial recurrences required additional surgery.

Throughout the literature there have been scant reports of using techniques, such as the Lynch and gullwing incisions, but these incisions have almost universally resulted in suboptimal cosmetic outcomes compared with other approaches. Alternatively, the closed rhinoplasty and endoscopic approaches have been used but in small lesions without significant cutaneous involvement. Most of the data on these surgical approaches have been described only in small case studies.

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The largest study of 42 patients advocates for the open rhinoplasty approach, endorsing superior exposure and cosmesis. In this study, various approaches were used, including the vertical midline incision (18 patients), external rhinoplasty (4 patients), Lynch incision (3 patients), transverse incision (5 patients), lateral rhinotomy (2 patients), endoscopic technique (2 patients), and combined approaches (8 patients). The authors noted improved surgical cosmesis with external rhinoplasty, although there were no objective data or comment on cutaneous involvement. The mean follow-up was 7.5 years, and 5 recurrences were documented, all with the use of different surgical approaches.

Bloom et al also published a small retrospective study of 10 patients, advocating for the superior cosmesis of the open approach without recurrence. These patients presented with masses in the glabella (6 patients), dorsum (3 patients), and nasal tip (1 patient). Six of these patients were treated with external rhinoplasty, 3 with the Lynch incision, and the last with a vertical midline excision. Cutaneous involvement or follow-up was not mentioned in this study.

Finally, a retrospective study of 8 patients by Bilkay et al found satisfactory cosmesis without recurrence in all patients undergoing open rhinoplasty (4 patients), vertical midline incision (3 patients), and the Lynch incision (1 patient). All of these patients were postoperatively evaluated by the surgeon and parents at a mean follow-up of 4.5 years. Overall, the authors still advocated for the open rhinoplasty, arguing an improved aesthetic result, wider exposure, more control over osteotomies, and better visualization of the cribiform plate.

Overall, the paucity of these lesions precludes large patient populations and data sets for retrospective analysis. Previous literature has adequately documented the advantages and disadvantages of various surgical approaches relative to recurrence rates and complications, although reports of cosmesis and follow-up are inconsistent. Many reviews found a preference for external rhinoplasty because of its superior exposure and cosmesis but did not always document the extent of cutaneous involvement, which still required an elliptical excision along the dorsum.

We agree that external rhinoplasty satisfies Pollock’s 4 principles but think that this approach tends to be more invasive and does not negate an elliptical excision of involved skin along the nasal dorsum. In our experience, minor extension of this excision in the craniocaudal direction allowed for adequate exposure of the mass and sinus tract in its entirety without excess manipulation of uninvolved tissue. This held true for our patients, who presented with anatomically distinct regions of the nasoglabella, including the patient undergoing intracranial extension (Figures 1-4). Reconstruction of the nasal dorsum in one patient with a mild soft-tissue defect was achieved with soft-tissue advancement flaps from bilateral nasal sidewalls. Two other patients had moderate soft-tissue loss that required regenerative tissue matrix for augmentation in addition to local tissue advancement. One patient with a bony defect that resulted from the tract that extended between the nasal bones underwent bone dust pate formed from the frontal craniotomy drill-hole sites. In all cases, a functional nasal airway and cosmetically acceptable nasal dorsal shape and midline scar were noted up to 1.5 years postoperatively.

**Conclusions**

Many options are available for surgical management of congenital nasal lesions, each with their strengths and weaknesses. Lesions with a visible pit or violation of the overlying skin necessitate superficial excision. Intracranial extension requires a frontal craniotomy and collaboration with a neurosurgical team. In these cases, the vertical midline approach provides wide exposure for excision and reconstruction of lesions in various areas of the nasoglabellar region. This approach also results in a cosmetically acceptable scar. Surgical management should be determined on a case by case basis with a multidisciplinary team.