Nasal Dermoid: A Segmental Approach

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Background/Purpose: Nasal Dermoid is a rare congenital anomaly. It represents persistent contact between the ectoderm and neuro-ectoderm at the prenasal space. The purpose of this study is to present the behavior of nasal dermoids, details of operative findings and results of management. An approach to the problem based on segmental divisions of the lesion has been also proposed.

Patients& Methods: Six cases of nasal dermoids were diagnosed and successfully treated at the Children’s Hospital, Cairo University during the period from June 2004 to October 2008.

Results: The average age at presentation was 2.4 years. The clinical presentations were diverse. The most common was ill-defined facial midline swelling (n=3). Others included paramedian swelling, midline and paramedian sinuses. Operative findings revealed almost identical behavior of the lesion in every case. Based on these operative findings, the lesion can be divided into four distinct segments: the proximal nasal dimple, the main tract, the central attachment and the accessory tract. Complete removal implies resection of all segments. Open facial approach was used in all cases in the form of either midline or modified lateral rhinotomy incision. One case showed minimal cerebrospinal fluid (CSF) leak that spontaneously stopped. There were no other medical or surgical complications. The mean follow up period was 15 months. No evidence of recurrence was noted in any patient. The final aesthetic outcome was satisfactory.

Conclusion: Nasal dermoid is a rare and often clinically underestimated congenital anomaly. Applying the concept of “track segments” is helpful in surgical planning and complete excision. The open facial approach provides generous exposure and allows complete resection with satisfactory esthetic outcome.

Index Word: nasal dermoid, midline facial swelling.

INTRODUCTION

Nasal dermoid is a rare anomaly. Its incidence has been reported to be 1 out of 20,000 live births. It constitutes 12% of all head and neck dermoids.1 The first reported case of nasal dermoid was back in the 1890s.2 Since then, only few reports with considerable number of cases were published. To the best of our knowledge, the largest three series in literature included 42, 36 and 25 cases over a period of 30, 20 and 8 years respectively.3,5 Plenty of other reports included a number of less than five patients.6-8

The mechanism of development of a nasal dermoid cyst is based upon the relation of the neuro-ectoderm and the ectoderm. The frontal and nasal bones are derived from mesoderm and are actually a zone of separation between the neuro-ectoderm and the surface ectoderm. At the bony-cartilaginous junction between the nasal bones and the cartilaginous dorsum, a space used to be found early in the embryonic life, a space that can be the contact site between the two elements. This is referred to as the “pre-nasal” space.1

Normally the dura extends through this “pre-nasal” space to touch the surface ectoderm then recedes centrally towards the foramen cecum totally free off its previous ectodermal contact. During this recession, persistence of neuro-ectodermal - ectodermal...
attachment pulls on the surface skin to form a deep tract that can extend up to the foramen cecum. It is the strict persistent contact of both the neuro-ectodermal and ectodermal elements that can result in the formation of nasal dermoid.9

Six cases of nasal dermoids that were diagnosed and successfully treated are reviewed in this article. The operative findings, details of surgical exposure and resection and results of surgery are presented.

PATIENTS AND METHODS

This series included six cases of nasal dermoids. Patients presented and were treated by the authors at the Children’s Hospital, Cairo University during the period from June 2004 to October 2008. History was taken including family history and history of adverse intrauterine effects. Clinical examination was done with emphasis on auxology and presence of other congenital anomalies (Figure 1).

All patients underwent both CT and MRI imaging. No intracranial extensions were found. The exact course of the tract and the bony defect were outlined. After radiological assessment, patients underwent surgical resection via an external facial approach. Preoperative antibiotics were administered in all cases.

Surgical Technique:

Two types of external facial incisions were used: the midline nasal dorsal incision for midline lesions and the modified lateral rhinotomy incision for extended ones. Incisions were planned in natural skin creases, respecting the esthetic subunits of the face. All incisions were designed to include elliptical skin incisions to include dimples and sinus openings. (Figure 2). Resection was planned to include all components of the nasal dermoid. This included the dorsal nasal sinus, the tract, any extensions either in the form of an intact swelling or a discharging sinus and finally the attachment of the tract to the skull base. The excised specimens were examined grossly and microscopically.

The outcome included assessment of surgical complications, recurrence, and final esthetic appearance. Assessment of the final esthetic outcome included two aspects: skeletal element and the final scar. Scars were classified according to a three-point score (score 1: totally noticeable, score 2 partly noticeable and score 3: totally concealed). The follow up period ranged from 8-24 months, with a mean of 15 months.

RESULTS

Four patients were males and two were females. Their ages ranged from 1-16 years with a median age of 2.4 years.

Four types of clinical presentations were encountered: midline swelling (n=3), paramedian swelling at the region of the medial canthus and upper eye lid (n=1), midline sinus (n=1) and paramedian sinus at the medial canthal region (n=1). A dimple on the dorsal nasal skin was noted in all cases. This was dormant in all but one case, who had repeated suppurations and discharge. Hair tuft was noted inside the nasal dimple in two patients. (Figure 1)

No associated congenital anomalies were noted except in one patient who had associated unilateral microtia and external auditory meatal atresia. Previous surgical attempts were documented in two patients. These were the two patients who presented with chronic discharging facial sinuses. The average number of previous surgeries was two attempts. Previous surgical attempts were simple sinus excision and were followed by immediate recurrence.

According to the operative findings, the tracts almost followed the same course. For descriptive purpose, the anomalous tract was found to be divided into four segments from distal to proximal: the distal nasal dimple, the main tract, the proximal cranial end and accessory tracts.

In all cases, the distal-most end of the lesion is represented by a skin pit located exactly in the midline at the supra-tip dorsum. There was no fixed site of this end along the length of the supra-tip area, which extends from level of the bony dorsum to the level of the nasal tip. This end was dry and asymptomatic in five cases, while in one case it was repeatedly discharging.

The main tract extended from the distal to the proximal ends. Its relation to the dorsal nasal skeleton was identical in all cases. It passed superficial to the cartilaginous dorsum then passed underneath the nasal bones to end at the base of the anterior cranial fossa. A classic deformity was noted in the form of cartilaginous dorsal saddling. During the passage of the main tract over the cartilaginous dorsum it caused this deformity by compression against the vault. (Figure 3)
Fig 1. Different Presentations of Nasal Dermoid: Midline swelling (A), Midline sinus (B), Paramedian swelling at the left medial canthal region and upper eye lid (C) and Paramedian sinus at the right medial canthal region (D).

Fig 2. Facial Incisions used. Midline incision for midline lesions (A), modified lateral rhinotomy extending to the upper eye lid (B) and modified lateral rhinotomy extending to the medial canthal region (C). Note that nasal pits as well as sinuses are included in the skin ellipse with the specimen.
Fig 3(A&B). Skeletal Deformity seen after dermoid resection. Note the concavity of the roof of the cartilaginous vault secondary to tract compression. This deformity was constant in all cases.

Fig 4. Different types of attachment between the central part of the dermoid and the dura: narrow (A) and wide tubular (B).

Fig 5. Esthetic outcome after open facial approach: midline (A) and modified lateral rhinotomy extending to the upper eye lid (B)
The distal end of the tract represented its central attachment to the skull base. This was located at the area of the foramen cecum. Access to this central bony attachment was achieved by drilling the bone at the root of the nose after out-fracturing the nasal bones. The attachment was dense fibrous in three cases, narrow tubular in two and wide tubular in one case. (Figure 4) There was no intracranial extension in any of the cases.

Accessory tracts were noted in three cases with different patterns. Extension to the orbit was noted in two cases, the first caused chronic discharging sinus at the medial canthal area and the second caused repeated upper eye lid cellulitis without skin breakdown. The third case in which an accessory tract developed had extension of infection through the frontal bone to open at the skin as a chronic discharging sinus at the glabellar area.

Examination of the main tract specimen after resection revealed skin lining with all forms of skin appendages including hair follicles.

Minimal intra-operative CSF leak was encountered in one case which was conservatively managed. The patient was kept under antibiotic cover for one week until the leak spontaneously stopped. No other anesthetic or surgical complications were encountered.

A median follow up period of 15 months showed no evidence of recurrence. Regarding the esthetic outcome, skeletal deformity in the form of obvious saddling was noted in one case. This patient underwent augmentation rhinoplasty 12 months after resection. Scars were excellent (score 3) in five patients, while was partly noticeable (score 2) in one patient. None of the patients had bad scars (score 1). (Figure 5)

**DISCUSSION**

Nasal dermoid is a rare congenital anomaly that is often clinically underestimated. Understanding the embryological background and its potential for central invasion is crucial for adequate management.

The most common presentation is a naso-glabbellar swelling. However, it can present in other diverse forms. In this study, 3 patients (50%) did not present with the classic nasal swelling. Other presentations include chronic discharging sinuses around the nose, and ill-defined canthal swellings. (Figure 1) Repeated infection either contained in the facial swelling or along the discharging sinus is a known presenting feature. Blake et al. reported infection as the presenting feature in 25% of cases.5

This article approaches the problem of nasal dermoid on basis of similar operative findings noted. Since the behavior of the tract was identical in the six cases, a segmental description may be useful in classification and management of such lesions. The abnormal tract can be clearly divided into four segments. The distal end is usually represented by a skin pit. This pit is important in two different aspects. The first is being useful in reaching the clinical diagnosis, although it does not cause any symptoms. The second aspect is the need for including this pit in resection to avoid recurrence. While this distal skin pit was a constant feature in all cases included in this series, others reported its absence in some. Denoyelle et al. reported 26 out of 36 patients with nasal dermoids had nasal pits, either as an only finding or combined with a midline mass. However, they reported 10 patients with masses who did not show this nasal pit.4

The main tract starts from the distal end and establishes a complex relation with the nasal skeleton. First, it runs above the cartilaginous vault. Pressure on the cartilaginous dorsum caused a classic “boat-shaped” deformity at the vault that was almost identical in all cases included in this study. (Figure 3) Then, it passes underneath the nasal bones towards the area of the foramen cecum. Attachment to the nasal bones may be dense enough to dictate osteotomies.10

A single case included in this series suffered obvious saddle nasal deformity after surgery. It is believed that the combination of the aforementioned “boat-shaped deformity” of the cartilaginous dorsum and the need for osteotomies to achieve resection is the underlying cause of the saddle deformity. Also, it has to be mentioned that this patient was 16 years old. Accordingly, presentation at older ages can increase the risk of post resection saddling. Holzmann et al. stressed on the importance of preserving the bony cartilaginous junction of the dorsum to avoid later growth impairment.10

The third segment is the central attachment, which is always at the region of the foramen cecum. The attachment to the dura was noted to be either fibrous or tubular adhesion. (Figure 4) The former can be
easily dissected off the dura, while the later needed careful dissection through a wide exposure to avoid leaving any epidermal remnants at the dural base. Denoyelle et al. reported only 2 among 36 cases who had dense meningeal adhesions. Holzmann et al. stressed on the need for osteotomy to release the tract from nasal bones and obtain adequate exposure.

The last segment of the lesion is the “accessory tract” that may or not exist. This tract represents the extension of infection through a facial plane to present at a site that is relatively away from the main tract and its proximal and distal attachments. The orbital region can be affected by spreading infection. Blake et al. reported 6 patients among 25, presenting with infection at the site of the main tract, with deep extension causing osteomyelitis only in one patient. In this series, orbital accessory tracts were noted in two patients. These were either in the form of discharging inner canthal sinus or cellulitis of the upper eye lid.

The only way to treat a nasal dermoid is complete resection leaving no remnants behind. The extensions have to be thoroughly assessed before surgery to provide the best chances for radical resection. CT scan can reveal bony defects. However, MRI has lately been recommended as the most effective and accurate means of evaluating a nasal dermoid. Regarding the approach, it seems the open rhinoplasty approach is gaining popularity. Open rhinoplasty approach can be also supported by nasal endoscopy.

Two concerns about the external rhinoplasty approach are worthy mention: exposure of extensions and associated obligatory external facial scars. While this approach may be satisfactory in dealing with the main tract and its proximal attachment, it may not be so in dealing with any lateral or superior extensions. Reviewing the three cases included in this series that showed medial canthal, upper eye lid and glabellar extensions, dissection of the diseased tissue needs proper exposure, especially that there was no clear line of demarcation between healthy and diseased tissues. The external facial incision is a modification of the lateral rhinotomy incision that can be tailored as needed. The need for tailoring arises from the direction of spread of the accessory tract.

The second concern is the obligatory facial scars. In cases presenting with multiple sinuses, elliptical excision of these sinuses is obligatory. This will end up with multiple facial scars. Even if multiple scars do not compromise the dorsal nasal skin with a columellar incision, the aesthetic outcome of multiple scars may not be quite different from a single one, especially if the intervening segments are hidden in junctional areas between facial esthetic units. The final esthetic outcome of external facial incisions in this series was satisfactory. Denoyelle et al. reported excellent scars in most cases, with widened scar in 4 out of 36 patients.

Finally, nasal dermoid is a clinically deceiving entity that is often more complicated than how it looks. Understanding the embryological background and behavior of the tract are essential to obtain complete resection. Based on constant operative findings, a “segmental approach” to the problem is proposed. Dividing the tract into different segments helps in preoperative surgical planning and in obtaining complete resection. Wide open exposure can be achieved with open facial incisions with satisfactory esthetic outcome.

REFERENCES


