

Original Article

Management of midline nasal dermoid lesions in children by external rhinoplasty



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المخلص

أهداف البحث: الأكياس الجلدية الأنفية هي عيوب خلقية نادرة تنشأ من الأدمة والأرومية المتوسطة. ويتم علاجها بالاستئصال الجراحي الكامل وهناك دعوات للعديد من الشقوق الجراحية. نعرض خبرتنا في استخدام نهج تجميل الأنف الخارجي لسبعة أطفال لديهم الأكياس الجلدية الأنفية والنواسير.

طرق البحث: تمت الدراسة بأسلوب استشرافي لـ 7 حالات لديهم أكياس جلدية أنفية ونواسير، تم تشخيصها سريريا وإشعاعيا، تم علاجهم بنجاح بواسطة نهج تجميل الأنف الخارجي في اثنين من المستشفيات التعليمية خلال الفترة من أبريل 2011م إلى نوفمبر 2013م. بعد الجراحة، كان متوسط متابعتهم سريريا لمدة 12 شهرا وملاحظتهم بالفحص السريري وتم توثيق النتائج وتحليلها.

النتائج: تم علاج سبعة أطفال لديهم أكياس جلدية أنفية و/أو ناسور بالاستئصال الجراحي بنجاح من خلال نهج تجميل الأنف الخارجي؛ 5 فتيات (71%) وولدين (29%) من الفئة العمرية 1.5 عاما إلى 16 عاما. وشمل الكشف السريري تورم خط الوسط في الوجه في خمس حالات، وناسور في خط الوسط في حالة واحدة وهوة ظاهرة في خط الوسط الأنفي عند مريض واحد. لم يكن هناك امتدادات تابعة للأكياس في هذه السبع حالات. تم تحقيق الاستئصال الكامل في 6 حالات، بينما كان هناك ظهور جديد بعد 6 أشهر في حالة واحدة مع استئصال كامل ناجح ومراجعة نهج تجميل الأنف الخارجي. كما تم متابعة جميع المرضى تقريبا لمدة عام بعد الجراحة.

الاستنتاجات: يعتبر نهج تجميل الأنف الخارجي نهجا جراحيا فاعلا لعلاج الأكياس الجلدية الأنفية والناسور للأطفال مع معدل منخفض جدا للظهور مجددا.

الكلمات المفتاحية: أطفال؛ كيس؛ جلدي؛ خط الوسط؛ أنفي

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Abstract

Objectives: Nasal dermoid cysts are rare congenital lesions of ectodermal and mesodermal origin. Treatment of these cysts entails complete surgical excision, and several surgical incisions have been advocated. We present our experience utilizing an external rhinoplasty approach in seven children with nasal dermoid cysts and fistulae.

Methods: A prospective study of 7 cases of nasal dermoid cysts and sinuses, diagnosed clinically and radiologically, were successfully treated by an external rhinoplasty approach in two teaching hospitals during the period of Apr 2011 to Nov 2013. Post operatively, an average follow-up of 12 months was observed by clinical examination, and the outcomes were documented and analysed.

Results: Seven children, 5 girls (71%) and 2 boys (29%), with ages ranging from 1.5 years to 16 years, who had nasal dermoid cysts and/or sinuses were successfully excised via an external rhinoplasty approach. The clinical presentation included midline facial swelling in five patients, a midline sinus in one patient and a dorsal midline nasal pit in one patient. No accessory tracts were found in these seven cases. Complete excision was achieved in 6 patients. There was one recurrence after 6 months with a successful complete resection with revision external rhinoplasty. All patients were followed up for an average of one year post operatively.

Conclusion: The external rhinoplasty approach is an effective surgical approach in treating paediatric nasal dermoid cysts and sinuses with a very low incidence of recurrence.

Keywords: Children; Cyst; Dermoid; Midline; Nasal

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Introduction

Nasal dermoid cysts are rare congenital lesions of ectodermal and mesodermal origin.¹ The majority of nasal dermoid cysts are reported in children. Most of the lesions present as dermal sinuses or cysts in the midline of the nasal dorsum.² Many other cases have presented with skin defects located anywhere from the glabella to the base of the columella.² A tract can exist from the nasal dorsum to the anterior cranial fossa.³

Histologically, dermoid cysts or sinuses are composed of a fibrous capsule of squamous epithelium and contain adnexal structures as hair follicles, sebaceous glands and sweat glands.⁴

These lesions are most frequently present in children but have also been reported in adults and sometimes extend intracranially.⁵ The incidence of nasal dermoid cysts in children is 1 in 20,000–40,000 live births.⁶ Many authors have reported that up to 12% of midline nasal dermoid cysts extend to the skull base and cribriform plate.⁶ The presentation of midline nasal cysts varies in complexity, from a short, blind dermal sac to a complex, multi-loculated lesion with a potential intracranial extension.³

The accepted treatment is complete surgical excision. Meticulous surgical planning is a must to avoid unexpected surgical situations. The nasal location of the lesions and probable involvement of deeper structures together with the possibility of an intracranial extension necessitate full clinical and radiological assessments before excision to prevent complications and local recurrence. A simple subcutaneous lesion, such as an epidermoid cyst, can present a challenge when located at the nasal tip or dorsum as regards aesthetic surgical management. When performed parallel to relaxed skin tension lines, a direct transcutaneous incision (commonly used for epidermoid cyst removal anywhere else) can distort the nasal tip subunit, resulting in a disfiguring scar. This should be avoided, especially in children. Careful pre-operative planning is essential to achieve optimal aesthetics.³ If radiology shows an intracranial extension, then a combined (intracranial and transnasal) approach is mandatory.^{7–9}

Many different surgical incisions have been advocated for the transnasal approach to completely excise a nasal dermoid. Pollock⁷ advised the fulfilment of four criteria in any surgical approach to such lesions: it allows access to the cyst and permits osteotomies, enables repair of cribriform plate defects and other complications, aids in reconstruction of the nasal dorsum, and ensures an acceptable scar. This prospective study of seven paediatric cases (less than 16 years of age) with nasal dermoid sinuses or cysts included their radiological investigations, surgical approach and outcome. We present our experience in utilizing an external rhinoplasty approach in seven children

with nasal dermoid cysts and fistulae, which allowed wide access with little visible scarring.

Materials and Methods

A prospective intervention study was performed by collecting data from seven patients aged 1.5 years to 16 years, with a mean of 6.3 years, who had congenital nasal dermoid cysts and underwent surgical excision using an open rhinoplasty approach during the period from April 2011 to November 2013 in two teaching hospitals. The study was conducted at Ain Shams University Hospitals Cairo, Egypt, and Ohud teaching hospital Almadinah Almunawwarah, KSA. Following institutional review board approval, the nature of the procedure was explained to the legal representatives of the patients, and informed consent was signed before the operations. All cases were successfully managed by complete excision via an external rhinoplasty approach.

The seven cases were all child patients diagnosed and surgically intervened by the authors during the research study. The diagnosis of congenital nasal dermoid cyst depended on clinical presentations (*Figure 1*) matching with congenital nasal desmoids and CT scanning. MRI was performed for the patient with bifid crista galli. Diagnosis was confirmed according to a tissue pathology examination for all cases postoperatively. The patients were presented to our O. P. clinics with a history of slowly progressive nasal swelling dating since early childhood or a discharging sinus on the dorsum of the nose for months or years. The discharge was always scanty and odourless.

A full otorhinolaryngological examination together with ophthalmological and neurological examinations were performed for each case. Office endoscopy (1.9°) using a nasal endoscope was conducted without local anaesthesia to exclude any associated intranasal lesions. This clinical diagnosis was confirmed by high resolution thin-cut spiral CT scans (*Figure 2*) of their facial bones and paranasal sinuses, which showed the characteristic soft tissue swelling over the nasal dorsum, occasionally separating nasal bones and, in one patient (female, 16 years), extending up to the anterior cranial fossa separating the crista galli into two.

MRI was performed for the patient with bifid crista galli to exclude intracranial extension and revealed no involvement of the anterior cranial fossa by the lesion with intact dura. After confirming the diagnosis radiologically, all of our patients were scheduled for elective open rhinoplasty to excise the lesion and reconstruct the resulting defect in one session.

Operative technique

An external rhinoplasty incision (*Figure 3*) is used with an inverted (V) columellar incision. The elevation of the skin and subcutaneous tissues covers the lower lateral and upper lateral cartilages and nasal bones. Gentle dissection of the dorsal nasal skin is carried out, especially over a distended cyst, to not break the skin integrity, which would cause an unpleasant scar after healing. A small transverse ellipse of the skin around the sinus opening is made, leaving the fibrous tract attached to the cyst cavity, while dissecting the tract without opening it until the nasal

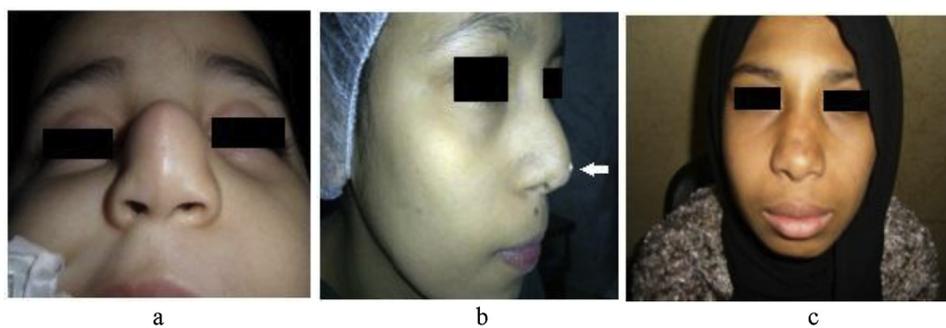


Figure 1: Different presentations. (a) Midline nasal dermoid in a 2-year-old child causing disfigurement and a visual field compromise. (b) A discharging sinus on the nasal dorsum in a 16-year-old female patient. (c) A blind pit over the nasal dorsum of a 14-year-old female child.

bones. There, the operating microscope is used to ensure the removal of any soft tissue inside the tract using a small bony curette or a burr with frequent irrigation to remove hairs and substances until the blind fundus of the tract is clearly visible and clean. An open roof resulted after clearing the bony cavity in 3 patients, which necessitated lateral and intermediate osteotomies to close it. Simple rasping of the dorsal bone was enough in the other 4 cases before realignment of the dorsal nasal skin without any irregularities or visible depressions.

After surgery, follow-ups were conducted for all patients with variable durations according to patient compliance. The follow-ups were typically conducted by clinical presentation. When recurrence occurred, follow-ups were performed by CT scanning.

Results

All patients underwent excisional surgery using the external rhinoplasty approach and were tabulated and

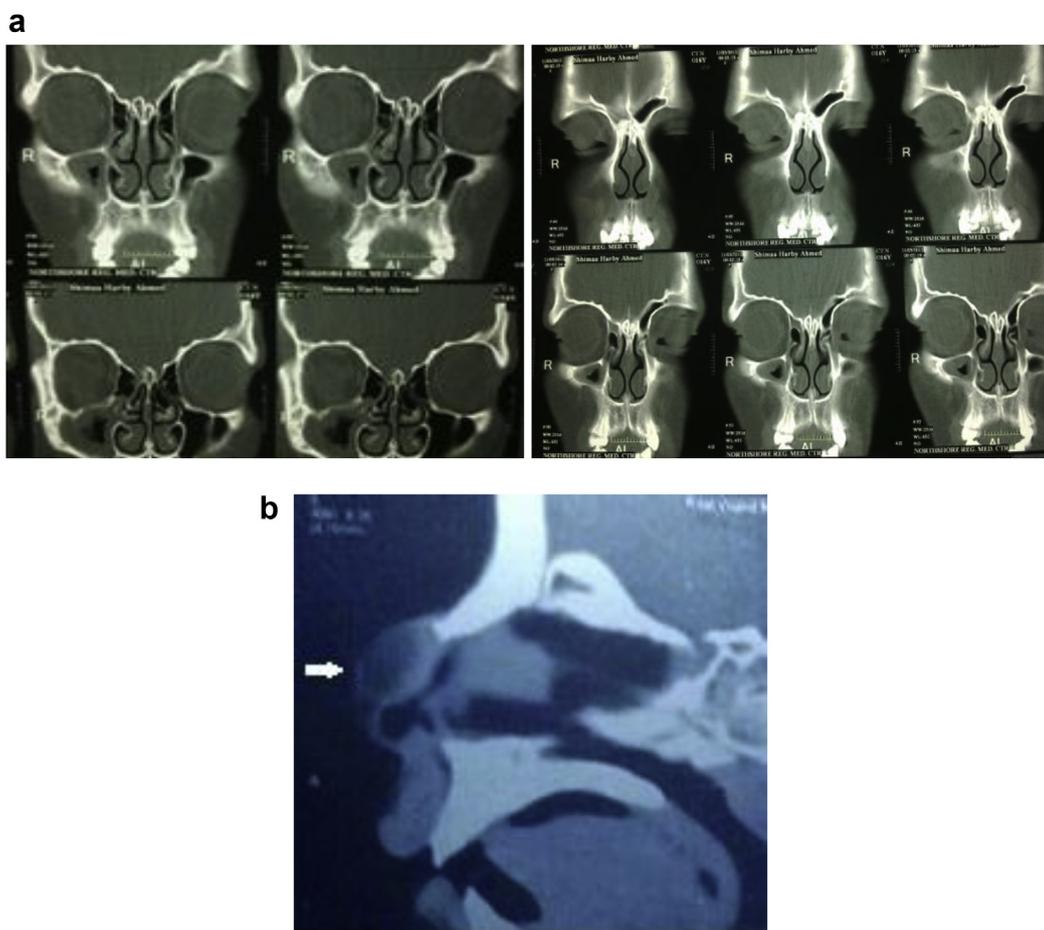


Figure 2: (a) Coronal CT scans showing a bifid crista galli in a 16-year-old female suspected have having an intracranial extension. (b) Sagittal CT Scan showing the cyst on the dorsum.



Figure 3: Open rhinoplasty approach exposing the nasal bones with a sinus tract containing hair in a 2-year-old female child.

Table 1: Patient demographics, presentations and outcome.

Age	Gender	Presentation	Complications
1.5 year	Male	Nasal swelling	None
2 years	Female	Nasal swelling	Recurrence
2 years	Female	Nasal swelling	None
14 years	Female	Discharging sinus	None
16 years	Female	Nasal dorsal pit	None
8 years	Male	Nasal swelling	None
1 year	Female	Nasal swelling	None

reviewed (Table 1). For five females (71%) and two males (29%), the youngest of whom was 1.5 years and the oldest of whom was 16 years, clinical presentation included (n: 5) midline facial swelling, (n: 1) a midline sinus and (n: 1) a dorsal midline nasal pit. No accessory side tracts were found in any of our seven cases. No intracranial extension was found in any of our seven cases. Complete excision was achieved in six patients.

Recurrence occurred in one case during the first 6 months of follow-up with slowly accumulating swelling over the nasal tip. Follow-ups ranged from 3 months to 2 years (with an average of one year), Figure 4.

Discussion

Nasal dermoid cysts or sinuses are generally rare congenital lesions in children with delayed onset of presentation. Diagnosis of such lesions can be made easily on clinical bases. The combination of a small pit, often in the midline of the nasal bridge, and the presence of emerging hairs is diagnostic of a median nasal dermoid.¹

Our study showed a female predominance of 5:2 in paediatric cases, including five females (71%) and two males (29%). The ages ranged from 1.5 years to 16 years at the age of presentation. The most common presentation was slowly enlarging dorsal nasal swelling, which causes disfigurement and psychological embarrassment to the patient or parents. This corresponds well with previously published results describing the impact of recurrent discharge and cosmetic disfigurement on children and their families.¹⁰

It is well established that nasal dermoid cysts or nasal sinuses must be treated as soon as they present with a primary complete single staged excision. Leaving these lesions or delaying treatment carries the risk of cyst infection or intracranial complications.^{1,3,11}

The role of radiological investigations is well recognized in the verification of such lesions and, more importantly, to exclude any intracranial extension of the lesion, which may change the treatment plan. A CT scan is the primary technique used, which is usually confirmative for diagnosing these lesions, but cannot exclude intracranial extension.

There is a radiologic sign, a bifid crista galli (Figure 2), which raises suspicion regarding the intracranial extension of the lesion. For such cases, MRI is strongly recommended before surgery because the treatment plan and incision may be changed according to the size and extent of the lesion.

In our case series, only one patient showed this radiologic sign. An MRI with Gadolinium was conducted, which excluded any intra cranial extension of the lesion. It is generally accepted that an inadequate excision invariably leads to recurrence³; therefore, after a full pre-operative assessment, careful surgical dissection under general anaesthesia is the recommended treatment of choice. In all of our cases, the cosmetic outcome was the most important factor in



Figure 4: Photographic views for follow-up of a case after more than two years.

determining the incision used because the reason for excision was purely cosmetic.

The external rhinoplasty approach, especially in young children with thin stretched skin over a distended cyst, is surgically demanding. Other approaches include a vertical midline skin incision over the cyst, transnasal endoscopic removal, transverse nasal incision, paramedical incision, medial canthal incision, eyebrow incision, gull-wing incision and bi-coronal flap, all of which have been advocated along with external rhinoplasty for nasal dermoids.^{1,3,10–12}

Our informed consent clearly mentioned that a further rhinoplasty could be performed after puberty. Of particular concern is the possibility of a supra-tip depression following surgery. For such a complication, the nasal cartilage and bone removed during excision should be preserved and used to carry out any reconstructive surgery as required.⁹

Out of our 7 patients, only one case (case no. 2, a 2-year-old female patient) had recurrent swelling over the nasal dorsum observed shortly after the initial surgery; this swelling was cystic and slow growing. A CT scan was performed and revealed a recurrent cyst at the area of nasal tip. A second surgery was conducted 6 months from the primary surgery using the same technique. The cyst was removed. The nasal bones showed a pit filled with hairs and desquamated epithelium, which was carefully curettaged and cleaned under a microscope. The patient was followed up for 6 months post operatively and exhibited no evidence of recurrence. Now, we believe that even a small remnant of epithelium can result in recurrence. The other cases had satisfactory cosmetic results during the follow-up period, with no further radiological studies conducted for them to avoid unnecessary exposure.

Strength and limitations

As a prospective intervention study, our study is more powerful with less bias than retrospective (review) studies, but the follow-up was for a variable duration according to the patient response, which is the main limitation of our study.

Conclusion

For aesthetic reasons, open rhinoplasty should be considered as a first treatment option in young patients with such lesions in the nasal tip. The results of surgery are acceptable in the majority of patients. A pre-operative radiological imaging evaluation is required for all patients to investigate the presence or absence of any deeper nasal or intracranial involvement. A long period of follow up is required to assess the long-term sequelae of performing open rhinoplasty in young children, although the short-term follow-up results are satisfactory.

Authors' contributions

All of the authors certify that they have participated sufficiently in the work to take public responsibility for the content, including participation in the concept, design, data

collection, analysis, writing, or revision of the manuscript. Furthermore, each author certifies that this material or similar material has not been and will not be submitted to or published in any other publication before its appearance in the journal of Taibah university medical sciences – Elsevier.

Ethical approval

The authors declare that the manuscript is original, ethically reviewed and approved.

Conflict of interest

The authors have no conflict of interest to declare.

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