International Journal of Pediatric Otorhinolaryngology 79 (2015) 18–22

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: www.elsevier.com/locate/ijporl

Nasal dermoids in children: A proposal for a new classification based on 103 cases at Great Ormond Street Hospital

B.E.J. Hartley, N. Eze *, M. Trozzi, S. Toma, R. Hewitt, C. Jephson, L. Cochrane, M. Wyatt, D. Albert

Great Ormond Street Hospital, United Kingdom

ARTICLE INFO

Article history:
Received 24 June 2014
Received in revised form 15 October 2014
Accepted 19 October 2014
Available online 24 October 2014

Keywords:
Nasal dermoid
Classification
Paediatric

ABSTRACT

Objectives: Nasal dermoids are rare developmental anomalies seen in children. This study reports the largest case series of 103 patients seen in a quaternary specialist unit over a 10-year period. We report the surgical and radiological findings and propose a new classification system, which clearly describes the extent of the lesions, thus allowing better surgical planning.

Methods: A retrospective review of case notes was conducted. Data collection included demographics, initial presentation, site of lesion, pre-operative CT and MRI imaging, surgical procedure, intraoperative findings (including depth of lesion), complications and recurrence. Surgical findings were correlated with radiological findings.

Results: A total of 103 patients were included in the study. The mean age at presentation was 29 months. 89% of children presented with a naso-glabellar or columellar lesion and 11% had a medial canthal lesion. All the patients underwent preoperative imaging and were treated with surgical excision. 58 children had superficial lesions, 45 had subcutaneous tracts extending to varying depths. Of these, 38 had intraosseous extension into the frontonasal bones, eight extended intracranially but remained extradural and two had intradural extension. There was good correlation between radiological and surgical findings. The superficial lesions were locally excised. The lesions with intraosseous tracts were removed via open rhinoplasty and the frontonasal bones drilled for access. Intracranial extension was approached either via a bicoronal flap and frontal craniotomy or the less invasive anterior small window craniotomy.

Conclusions: This report describes the largest published cases series of nasal dermoids. The cases demonstrate the presenting features and the variable extent of the lesions. The new proposed classification; superficial, intraosseous, intracranial extradural and intracranial intradural, allows precise surgical planning. In the presence of intracranial extension, the low morbidity technique of using a brow incision and small window anterior craniotomy avoids the more invasive and commonly used bicoronal flap and frontal craniotomy.

1. Introduction

Nasal dermoids are rare developmental anomalies occurring in 1 in 20,000 births and are the most common congenital midline lesion [1,2]. They represent 3% of all dermoids and 12% of head and neck dermoids [3,4]. The term, nasal dermoid cyst was first described by Cruvelier in 1817 [5]. The child presented with a hair-containing sinus on the nasal dorsum. The terminology has been somewhat confusing with many descriptions including dermoid, dermal cyst and dermoid cyst. Sessions [5] coined the term 'nasal dermal sinus cyst' which incorporated all lesions with mesoderm (adnexae) and ectoderm (stratified squamous epithelium).

Three layers are used during the formation of the nose during embryogenesis; ectoderm, mesoderm and a deep cartilaginous capsule. Intramembranous ossification occurs in the mesoderm to create the nasal and frontal bones in the eighth and ninth week. They are separated by the fonticulus nasofrontalis [6]. The prenasal space forms between the nasal bone superficially and the deeper cartilaginous capsule. A small tract of dura extends to the skin and becomes separated and encircled by the foramen caecum as the nasal process of the frontal bone grows. The dura will usually obliterate thus removing the neuroectodermal connection. After

* Corresponding author at: ENT Dept, Great Ormond Street Hospital, Great Ormond Street, WC1N 3JH, United Kingdom. Tel.: +44 207 405 9200.
E-mail address: nnekaeze@yahoo.co.uk (N. Eze).

http://dx.doi.org/10.1016/j.ijporl.2014.10.020
0165-5876/

this, the fonticulus nasofrontalis and foramen caecum fuse and the cribiform plate forms.

Embryologically, dermoids are derived from ectodermal and mesodermal layers [7]. They are lined by stratified squamous epithelium and may contain adnexal structures such as hair follicles and sebaceous glands. They arise early in embryonic development during closure of either the anterior neuropore or the fonticulus nasofrontalis or during development of the frontonasal process [8].

Several theories relating to origin have been published but several do not account for the intracranial element of the disease [5,9]. The final and most popular theory was by Grunwald described in 1910 [10], which was later coined the 'prenasal theory' by Pratt [11] and the 'cranial theory' by Bradley [12]. The theory suggests that as the neuroectodermal tract recedes, dermal attachments may follow, such that as the dura recedes from the prenasal space the nasal ectoderm is pulled upwards and inwards creating an epithelium lined sinus or cyst.

Dermoids in the nasofrontal region may present as a mass, a sinus or a fistula tract and need to be differentiated from other congenital nasal masses including gliomas, encephalocoeles and polyps [13]. They appear in the midline on the dorsum of the nose between the glabella and columella. The lesions can discharge and progressively enlarge with time leading to local soft tissue or skeletal deformity. The cysts and sinuses may connect with an intracranial component via an abnormal foramen caecum in the anterior cranial fossa. Appropriate management includes accurate diagnosis and complete surgical excision in order to avoid recurrence and development of complications.

Given the variability in the extent of the sinus or fistula tract, much consideration has been given in this report to the description of the lesion using a new classification system such that the surgical approach can be better tailored to each patient. In addition the surgical findings are correlated to the radiological findings.

2. Methods

A retrospective analysis was performed in all patients admitted for treatment of nasal dermoid between 2001 and 2012 at Great Ormond Street Hospital. 112 patients were identified of which 103 were suitable for inclusion in this series. Nine patients were excluded due to unavailability of complete medical records. The data recorded included patient demographics, initial presentation, site of lesion, pre-operative CT and/or MRI findings, surgical procedure, intraoperative findings, complications and recurrence. Operative findings were correlated with preoperative imaging.

3. Results

103 patients are presented in this series, there were 44 (43%) female and 59 (57%) male. The age at presentation ranged from birth to 90 months with a mean of 29 months. 91 children presented with a nasoglabellar mass of which 66 had a visible sinus. Other presentations included a mass in the inner canthum (10), nasal tip and columella (2). 27 (28%) patients had a history of infection resulting in erythema, pain and discharge. A summary of the clinical presentations is summarised in Table 1. No patient had a history of intracranial infection or meningitis. Eight patients had an associated craniofacial anomaly and three patients had a positive family history. Follow up ranged from 3 months to 8 years with a mean of 3.2 years.

All the children underwent preoperative imaging in the form of magnetic resonance imaging (MRI) (103) or combined MRI and computed tomography (CT) (11). The presence of a tract was noted in the preoperative images of 42 patients. The preoperative imaging revealed intracranial extradural extension in eight patients thus necessitating anterior craniotomy for access. In all the cases, the tract was adherent to the dura which was carefully separated from the lesion without leakage of cerebrospinal fluid. Two patients had intracranial intradural extension, which was identified on preoperative imaging.

Table 1
Summary of clinical presentation.

	Mass	Sinus	Mass and sinus	Total
Glabella	4	1	8	13
Nasal dorsum	9	3	66	78
Nasal tip/columella	–	2	–	2
Inner canthum	1	1	8	10
Total	14	7	82	103

The 103 patients all underwent surgical excision, which included excision and direct closure (56), open rhinoplasty (37), bicoronal excision and craniotomy (7) and small window craniotomy (3). The extent of all tracts was noted and is summarised in Table 2.

Surgical and radiological findings were concordant in 90 (87%) cases. In four cases the intracranial extension was not noted but was evident intraoperatively as the tract extended up to but did not breech the dura. The two cases with intradural extension were both identified preoperatively.

Histopathological analysis of all the specimens confirmed the diagnosis of dermoid cyst with dermoid elements identifiable.

Complications in this group were few and included wound infection in two cases and recurrence in three cases which presented as subcutaneous masses which were readily re-excised. There were few concerns regarding the cosmetic results and there were no cases of altered nasal bone development and growth during follow-up.

4. Discussion

Nasal dermoids cysts commonly present at birth or soon after and have a male predominance, which is seen in this report (57%) and is comparable to figures published by Denoyelle [1] and Rahbar [14]. The lesions are occasionally known to be familial [5,15,16]. In this series, three patients had a clear positive family history.

These lesions usually present as a midline mass on the nasal dorsum and may be associated with a sinus opening and intermittent discharge of sebaceous material and recurrent infection (Fig. 1). Hair can commonly emerge from the sinus opening and is deemed pathopneumonic for nasal dermoid [17]. In this series 66 patients had a dorsal sinus and five had hair emanating from the sinus. As in the report by Rahbar [14], the position of the superficial cyst or sinus did not aid in the prediction of intracranial extension.

Intracranial extension has been well described in the literature (Wardinsky 45%, Ghestem 32%, Bartlett 44%, Posnick 36%, Sessions 31%, Denoyelle 17%, Bradley 4%) [1,5,6,12,17–19]. The intracranial tract traverses the foramen caecum or cribiform plate to reach the anterior skull base and adhere to the falx cerebri extradurally.

Table 2
Classification compared to series published by Bradley et al. (1983).

Classification	Bradley (1983) $N=74$	This series $N=103$
Superficial	(45) 61%	(56) 54%
Intraosseous	(17) 23%	(37) 36%
Intracranial extradural	(12) 16%	(8) 8%
Intracranial intradural	0	(2) 2%