Naso-Orbital Fistula in a Young Child

Abstract
Nasal dermoids are the most common midline congenital masses of nose presenting in early childhood. Most of them present before the age of 3 years. They present as midline nasal mass or a sinus opening anywhere from glabella to columella. They are lined by keratinized squamous epithelium with adnexal structures. Some of them may secrete sebaceous material and are prone to recurrent infection. They may be associated with intracranial extension. Here, we present a case of infected nasal dermoid with intracranial extension and orbital fistula.

Keywords: Dermoid; Ectropion; Nasofrontal

Introduction
Nasal dermoid was first described by Cruvlier in 1817. The most accepted theory of nasal dermoid is Grunwald prenasal space theory. The prenasal space is located between the nasal bones and the nasal capsule (precursor of nasal septum and nasal cartilages). During foetal development these spaces are closed by fusing and ossifying. Abnormal development of these structures is thought to be the cause of formation of nasal dermoids. We present a case of nasal dermoid with recurrent infection presenting as orbital fistula in a 3 year old child managed by complete surgical excision.

Case Report
A 3 year old male child presented to otolaryngology OPD with complaints of swelling over dorsum of nose since the age of 1 year. The swelling was approximately the size of pea, not associated with pain and there was hair follicle over the swelling. There was no increase in the size of swelling on crying or coughing. Since the swelling was asymptomatic, parents took no treatment for it, however 6 months later, parents noticed another swelling near the medial canthus of right eye. It was associated with pain and redness of overlying skin. Parents went to a private hospital where child was admitted and started on intravenous antibiotics. The swelling did not decrease in size and incision and drainage of the swelling was done. Approx. 2ml pus was drained and dressings were done daily. Child improved and pus stopped with scarring of incision site. Child was discharged after 7 days of i.v antibiotics. Few days later, parents noticed pus discharge from the scarred incision site through a small opening. Parents went to private hospital and the child was referred to higher centre. On examination, there was nothing of medial part of right upper eyelid with a small scar near the medial can thus with a fistulous opening and pus discharge from it. On examination of nose, a 2x2cm swelling was seen on the dorsum at the root of nose.

It was not tender, soft, with a pit and a hair follicle over it. Cough impulse was absent. Anterior rhinoscopic examination was normal. Ophthalmic examination revealed notch of right upper eyelid medially. The cornea, vision and eye movements were normal. There were no other associated congenital anomalies. A provisional diagnosis of infected nasal dermoid was made and child was admitted and i.v antibiotics were started. MRI PNS and brain was done which revealed a well defined peripherally enhancing lesion in fronto-ethmoid region with bony defect in the right side of the frontal bone with contiguous spread in preseptal, extracranial space of right orbit suggestive of infected dermoid cyst (Figure 1). CECT face was done which revealed small well defined round hypodense lesion measuring 1.5x1.45x1cm in the fronto-ethmoidal region at the root of nose with associated bony defect in the right frontal bone. The foramen caecum appeared widened suggesting intracranial extension. Excision of the dermoid under general anaesthesia was planned. Methylene blue dye was injected through the pit on the dorsum of nose and it came out through the fistula at medial canthus confirming communication between the two sites (Figure 2). A midline vertical incision was marked incorporating the opening on the nose and tract was traced. Tract was found going medially between the nasal bones. A part of nasal bones was removed and tract was found going intracranially through a fibrous strand in the region of cribiform plate. The fibrous strand was cauterized with bipolar cautery and tract along with sac was removed in toto. The margins of orbital fistula were freshened and sutured. 6 months follow up revealed healed fistula. Histopathological report of the tract confirmed it to be dermoid.
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Discussion

Nasal dermoids comprise 1 to 3% of all dermoid cysts. Hair follicle with punctum is pathognomonic of nasal dermoid. They may be associated with other congenital anomalies. These include craniofacial anomalies, hyper-telorism, cleft palate, hemifacial microsomia, aural atresia, pinna deformities, branchial sinus anomalies, cardiac, genital and gastrointestinal anomalies [1]. Therefore, these anomalies must be looked for. Most of the patients present at birth or in early childhood, but cases of adult onset nasal dermoids have been reported [1]. The oldest patient in the literature is a 56-year-old man [2]. In our case, the child presented at 3 years of age without any associated anomalies.

Clinically, they may present as asymptomatic inconspicuous mass on dorsum of nose. They may secrete sebaceous material and are prone to recurrent infection. Infected nasal dermoids may present as abscess and if untreated, infection can lead to osteomyelitis of underlying bone resulting in deformity. Increase in the size of swelling on crying or coughing is pathognomonic of intracranial extension. Intracranial extension is seen in 4-45% cases [3]. In our case, patient presented with an infected nasal dermoid which was drained considering it to be a simple abscess, ultimately leading to an orbital fistula. Therefore, nasal dermoid should be considered when young child presents with midline nasal abscess.

The definitive treatment for nasal dermoid is complete surgical excision. Imaging forms an important part of preoperative evaluation for nasal dermoids. Imaging is required to know the size and extent of the swelling and to rule out intracranial extension. Both CT scan and MRI are complimentary for planning surgical excision for nasal dermoids. CT scan is essential to identify bony anatomy and intracranial extension. The reported findings in case of nasal dermoids with intracranial extension on CT scan are: bifid crista galli, widened nasal septum, defects in cribiform plate, widened perpendicular plate and interorbital widening [4]. MRI depicts soft tissue anatomy well and therefore, helps us to know the intracranial extension. Intracranial extension of nasal dermoid is most frequently extra-axial, attached to the dura or confined within the leaves of the anterior falx. Intra-axial extension into brain parenchyma is exceptional [5]. Multplanar thin section MRI with T1 weighted images must be obtained. Gadolinium enhanced, fat suppressed T1 weighted images are used to depict the anatomy of enhancing cartilage at skull base in infants. The use of contrast helps in differentiating between non-enhancing dermoid and other enhancing lesions such as hemangiomas [6].

In a study on ten patients of nasal dermoids, it was concluded that MRI alone is the most accurate investigation of choice for nasal dermoids and CT scan is associated with false positive and false negative results [7]. In our case, both, CT and MRI were done. We recommend both, CT and MRI must be done before planning surgery. Various surgical approaches have been described for nasal dermoid. When intracranial extension is suspected, combined approach is essential to ensure complete surgical excision. The best surgical approach is the one which ensures complete removal of the swelling with the tract, is able to deal with intracranial extension and repair of CSF leak if it occurs.

The various surgical approaches are external rhinoplasty, midline vertical incision, craniofacial, subcranial, horizontal nasofrontal incision and endoscopic approach. We used midline vertical incision in our case. The advantages of external rhinoplasty approach are: wide exposure, cosmetically acceptable scar and is able to deal with intracranial extension. Midline vertical incision allows wide exposure and can be extended to skull base horizontal nasofrontal incision can be used for cysts limited to nasofrontal region. Endoscopic approach is useful for nasal dermoid limited to nasal cavity without cutaneous or intracranial extension. The recurrence rate of nasal dermoid after surgical excision is 12%. With craniofacial approach it is reported as 30% and with subcranial approach it is reduced to 16% [8]. Postoperative complications are: tension pneumocephalus, CSF leak and subdural haematoma.

Conclusion

Nasal dermoid, though, a rare congenital anomaly is the most common cause of midline nasal mass in children. High index of suspicion is required to diagnose it early and avoid delay in treatment. Early surgical intervention is required to prevent recurrent infection and associated complications and prevent distortion of nasal bone anatomy.

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