



Management of Nasal Dermoid Cyst with Sinus in a Secondary Healthcare Setting - A Case Report

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Abstract

Nasal dermoid sinus cysts (NDSC) are rare congenital midline lesions that have a distinct embryological origin and require careful approach to management. NDSCs can present in various forms such as cystic swelling, sinuses or intracranially extending tracts. Because of its frequent association with tracts or sinuses connecting to the skull base or the anterior cranial fossa, a careful and tailored approach is a must. Here we present a case of 4 year old child with nasal dermoid cyst with sinus tract who underwent complete surgical excision. The postoperative follow-up showed no complication. This case underscores the significance of early diagnosis of NDSCs and need for its complete excision to prevent recurrence.

Keywords: Nasal Dermoid Sinus Cyst; Congenital; Vertical Incision; Intracranial

Introduction

A dermoid cyst is a developmental anomaly that arises from the entrapment of ectodermal elements along the lines of embryonic closure [1,2]. Head and neck dermoid cysts constitute about 7%

of all dermoid cases [3]. Nasal dermoid cysts are a rare entity representing 3% of all dermoids and 12% of head and neck dermoids [4]. The incidence of dermoid cysts and fistulas in the midline of the

nose is 1/20,000 to 1/40,000 [5]. Nasal dermoids can present as cysts, sinus, fistula and/or with intracranial extension.

Nasal dermoid sinus cysts are rare congenital midline lesions that have a distinct embryological origin and require careful approach to management. Patients with NDSCs usually present as midline lesions either as cystic swelling or as sinus tract usually opening in the midline of nasal dorsum between the glabella and the columella. Commonly, the sinus may discharge cheesy material or purulent discharge with secondary bacterial infection. A tuft of hairs emerging from a midline nasal swelling or punctum is nasal dermoid sinus cyst until proved otherwise [6]. They are usually present from birth, although symptoms may not manifest until later in life [7].

Several theories have been proposed regarding the pathogenesis of nasal dermoid sinus cyst. The most popular and accepted theory was proposed by Grunwald in 1910 [8], which was later coined the 'prenasal theory' by Pratt in 1965 [9] and later Bradley [10] referred it as 'cranial theory'. This theory suggests that NDSCs develop due to failure of separation of neuroectodermal tissue from ectodermal tissue during 8th and 9th week of embryonic development. When the neuroectodermal tissue recedes from the prenasal space, its dermal attachments the nasal ectoderm can be pulled upwards and inwards creating an epithelium lined sinus or cyst.

Hartley, et al. have suggested a staging system depending on deep extension of the lesion [11]

- **Superficial:** Remain within the soft tissue.
- **Intraosseous:** Extend to frontal and nasal bones.
- **Intracranial extradural:** Breaches the anterior cranial vault
- **Intracranial intradural:** Most complex, extends intracranially to involve the dura

If recurrent infections are present, it may lead to underlying bony and cartilage resorption leading to nasal deformity. They can experience severe complications such as periorbital cellulitis, facial cellulitis and focal abscess.

It is very important to assess and rule out intracranial extension of NDSCs. Intracranial extension should be assumed until proven otherwise. They may have an abnormal communication with in-

tracranial space through an abnormal foramen cecum in the anterior cranial fossa. Such connections are not usually apparent on clinical examination. When intracranial extension is present, complications like meningitis, meningoencephalitis and/or frontal lobe abscess may occur. Reported rates of intracranial extension vary considerably, ranging from 4 to 57%, although one meta-analysis placed the incidence at 19.6% [12]. Also other congenital midline tumours like ectopic neuroglia, encephalocele and teratoma need to be carefully considered during assessment.

Imaging studies such as CT scan or MRI scan are important to confirm the diagnosis, assess the extent, structures involved and rule out intracranial extent. They also help in planning for surgery. Dermoid cysts appear as non-enhancing low-density masses on CT scans and are hyper intense on T1-weighted MRI sequences [13].

Early surgical excision is recommended to prevent complications or further expansion. Depending on its site and extent, different approaches are curated to ensure complete surgical clearance of the disease. The surgical approach depends on the site, size and extent as well as surgeon's expertise. NDSCs without intracranial extension are approached by external rhinoplasty approach or by direct midline vertical incision. Other external incisions include transverse linear, curvilinear, lateral rhinotomy, glabellar incision [14,15]. Because of its frequent association with tracts or sinuses connecting to the skull base or the anterior cranial fossa, a careful and tailored approach is a must. Here we present a case of 4 year old child with nasal dermoid cyst with sinus tract, a rare congenital anomaly.

Case Presentation

A 4 year old boy was brought to our hospital by his parents with complaints of purulent discharge from wound over nose since 5 days. He had similar episodes in the past since birth. They also reported that the child had tufts of hair protruding from a punctum present on the bridge of the nose since birth. No previous episodes of headache or dizziness, no documented fever or neck stiffness or seizures was seen. Antenatal, natal and postnatal histories were uneventful. No similar conditions was seen in his siblings or relatives.

The examination revealed a pit in the glabellar region with purulent discharge coming from it. The surrounding skin was erythe-

matous and tender. Another pit was seen in the middle part of nasal dorsum with tuft of hair coming out of it. Rest of the examination was normal.



Figure 1: A midline NDSC with a pit in the middle of nasal dorsum with tuft of hair protruding from it. Another pit at the glabellar region is seen.

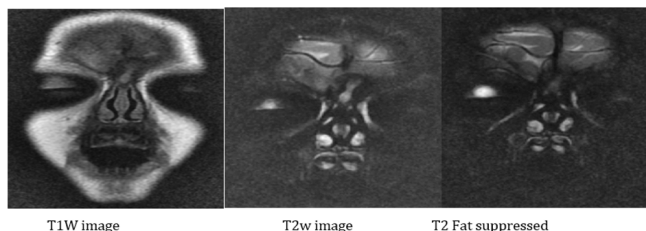


Figure 2: Magnetic Resonance Imaging (MRI) of the face revealed curvilinear sinus tract appearing hyper intense on T1w images and T2w images and hypointense on T2 fat suppressed coronal images extending from skin over glabella through nasal process of frontal bone running inferiorly in midline and opening to skin over the nose in the midline. No ramification or underlying bony erosion or intracranial extension was seen. These findings were suggestive of nasal dermoid cyst with sinus.



Figure 3: The sinus tract being dissected from surrounding tissue. A midline vertical incision over nasal dorsum was made and methylene blue dye was instilled into the sinus tract for better delineation.

After routine evaluation and pre-operative workup, the child underwent complete excision of the nasal dermoid cyst with sinus under general anaesthesia. A direct midline vertical incision with elliptical incision to include the puncta was made after instilling methylene blue dye in the sinus tract. The sinus was separated from surrounding tissue and excised intoto. After ensuring haemostasis, the incision was closed in layers. Post operatively the child had developed bilateral periorbital swelling which subsided with conservative management. The wound healed well. The child was followed up and no recurrence was seen. Histopathological evaluation came as dermoid cyst with sinus.

Discussion

Nasal dermoid sinus cysts are rare congenital lesions that usually present with the sinus opening commonly located along the midline of the nasal bridge. Hair protruding from the sinus or punctum is considered pathognomonic for a nasal dermoid sinus cyst. However, the cyst may remain asymptomatic and be discovered incidentally during routine medical examination also. In our case,

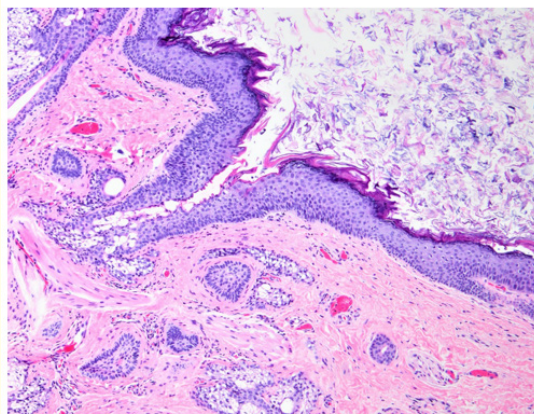


Figure 4: Photograph of histopathology of the nasal dermoid sinus cyst lined by stratified squamous epithelium with cutaneous adnexal structures.

the child presented with repeated episodes of purulent discharge from the sinus opening associated with pain, fever. The child had tufts of hair protruding from a punctum present on the bridge of the nose since birth.

It is important to rule out intracranial extension of the NDSC. Imaging studies like CT and MRI help to rule out the intracranial extension and to assess the extent of the cyst, sinus tract and any associated abnormalities. They also help in planning of surgery. In our case, MRI of face was done which ruled out the intra cranial extension.

The treatment of choice for intracranial NDSC involves an external nasal incision combined with a bicoronal flap and frontal craniotomy [16,17]. However this approach is associated with greater morbidity. It requires blood transfusion, longer operative time, and longer hospital stay. To obviate these drawbacks, *minimal access keyhole-type craniotomies have been proposed*. Endoscopic removal of nasal dermoid cysts that extend intracranially has gained popularity as it has a lower morbidity rate than that of a frontal craniotomy [18,19]. In our case, midline vertical incision was used.

Post operatively, the patient should be monitored for any signs of infection, delayed wound healing. Regular follow-ups to be done to look for any signs of recurrence or complications. Our patient's

post-operative period was uneventful and his wound has healed well.

Conclusion

NDSCs are rare congenital midline anomalies with varied presentation. Early evaluation and definitive treatment are paramount for positive outcome and to prevent complications. Clinical examination and radiological investigations are important for diagnosis and to rule out intracranial extension. The surgical approach is curated accordingly. The definitive treatment for NSDCs is complete surgical excision. Regular post operative follow-ups are essential to ensure long term resolution of the disease. This case emphasizes the importance of early diagnosis by clinical and radiological evaluation and complete surgical excision of NSDC to prevent complications and recurrence.

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