

Nasal Dermoid Sinus Cyst expanding into Frontal Sinus: A Rare Presentation

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ABSTRACT

Nasal dermoid sinus cysts are uncommon congenital anomalies that require interventions only when they present as complications. They frequently present in childhood as midline nasal masses requiring excision. They may present with intracranial as well as intraorbital extensions, making it necessary to subject the patient for proper radiological diagnosis preoperatively. Here, we present a rare case report of 28-year-old male patient who presented at our tertiary health care center with nasal dermoid cyst expanding into the frontal sinus causing proptosis and blurring of vision. The cyst was excised by combined approach, i.e. external nasal dermoid sinus tract excision and endoscopic nasofrontal sinusotomy for complete removal. The embryology, clinical history, diagnosis and surgical management are discussed.

Keywords: Nasal dermoid sinus cyst, Frontal sinus, Endoscopy.

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INTRODUCTION

Dermoid cysts are benign choristomas.¹ Nasal dermoid sinus cysts (NDSCs) are an uncommon midline anomaly with an embryologic origin, quite distinct from dermoids elsewhere on the face and requiring an especially careful approach to management.² Dermoid cysts occur in three primary locations in the head and neck: The frontotemporal region, the periorbital region and the nasoglabellar region.³ They are congenital lesions lined by stratified squamous epithelium, with normal dermal appendages, including hairs and sebaceous glands. The differential diagnosis needs to be carefully considered during assessment and includes ectopic neuroglia, encephaloceles and teratomas. Its usual clinical presentation is in the form of a sinus tract on dorsum of nose, anywhere between the base of columella and glabella, distal one-third being the most common site. They are firm, nonpulsatile and noncompressible masses which do not transilluminate.⁴ On reviewing the English literature, to the best of our knowledge, this is the second NDSC extending into the frontal sinus and orbit and the first case which was managed by endoscopic frontal sinusotomy along with excision of tract externally. Proper clinical history and radiological investigations are important before planning the surgical management.

CASE REPORT

A 28-year-old male presented with a slowly progressing, painless swelling involving the nasal side of the upper left eyelid for last 1 year. It was associated with foreign body sensation and numbness of skin around left eye. The patient had history of intermittent watering of left eye and blurring of vision. He had history of small opening on dorsum of nose since childhood but patient noticed intermittent, foul smelling, cheesy material from the opening since 4 years but he did not get any treatment for the same. There was no history of headache, seizures, nasal discharge, trauma or any other general ailment. On examination, there was a diffuse swelling at the nasal side of the left upper eyelid measuring approximately 3 × 2 cm on left side, and the skin over the swelling appeared stretched. Examination revealed the swelling to be firm, noncompressible, nonreducible and nonpulsatile. Visual acuity and ocular movements were within normal limits. Systemic examination revealed no neurological deficit. There was a sinus tract opening present over the dorsum of nose about 1 cm below the root of the nose. Cheesy material with hair could be appreciated coming out of the sinus opening.

Contrast enhanced computed tomographic (CECT) scan of head, orbits and paranasal sinuses showed a large well-margined hypodense nonenhancing lesion of 4.9 × 4.0 × 1.9 cm size in the left frontal sinus eroding the posterior table, superior orbital rim medially and erosion of the roof of ethmoids (Fig. 1). Considering the clinical and radiological findings, the possibility of a benign lesion was considered with differential diagnosis of dermoid cyst, epidermoid cyst or frontoethmoidal mucocele.

Subsequently, the patient was operated upon and vertically elliptical incision was given around the sinus opening and the sinus tract was excised which was opening into the frontal sinus posteriorly. Cheesy foul smelling material with tufts of hair was present. Mucocele sac was marsupialized endoscopically by performing left frontal sinusotomy. Frontal sinus ostium was widened; cheesy material was present in whole of the widened frontal sinus which was extruded. There was a large defect in the posterior wall of the frontal sinus and superomedial wall of the orbit.

Histopathological examination of the resected mass showed stromal fragments, sebaceous glands and fibrofatty tissue with infiltration by chronic inflammatory cells spread

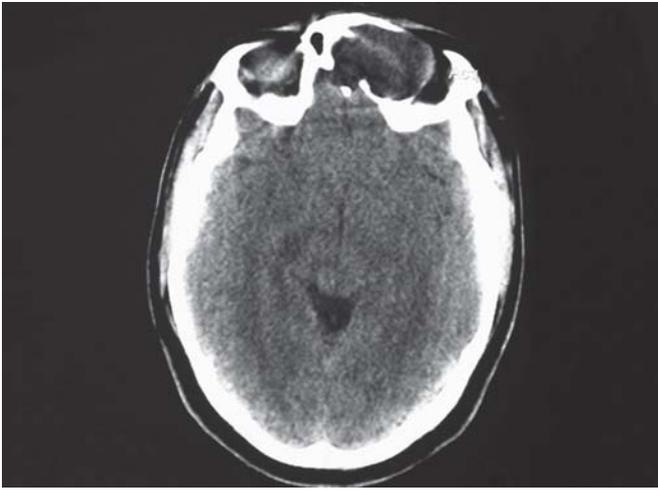


Fig. 1: Noncontrast computerized tomography of nose and paranasal sinus showing midline NDSC extending posteriorly into the left frontal sinus

around blood vessels. The section also showed keratin material consistent with dermoid cyst. Postoperative recovery was uneventful. There was no CSF leak or neurological deficit. The patient improved cosmetically, and is under regular follow-up since 8 months with no fresh complaints or signs of recurrence (Fig. 2).

DISCUSSION

Dermoid cysts are a subset of benign heterotopic neoplasms termed choristomas.¹ They are thought to arise in the early embryonic period, during closure of the anterior neuropore, during development of the frontonasal process or during closure of the fonticulus nasofrontalis (a potential defect between the developing frontal and nasal bones).^{5,6} They may present in a variety of ways: As a cystic mass or as a sinus opening onto the midline dorsum of the nose between

the glabella and the columella.⁷ Commonly, the sinus will discharge cheesy offensive material or grow abnormal hairs. Both cysts and sinuses may have a connection with an intracranial component through an abnormal foramen cecum in the anterior cranial fossa. Such connections are not usually apparent on clinical examination. Etiologically, they are thought to derive from dermal and epidermal tissues trapped in the cranial fusion lines as the neural tube closes in embryogenesis.⁸ Histologically, they have a lining of squamous epithelium with dermal elements, such as hair follicles, sebaceous and sweat glands. Within the cyst, keratin, hair, smooth muscle and lipid debris can be found. The sites involved by dermoid cyst lie at the confluence of the surgical expertise of otolaryngologists, neurosurgeons, plastic surgeons and ophthalmologists. At times, this calls for a multidisciplinary approach to resection. Our case of NDSC extending into the frontal sinus eroding the posterior table and floor causing proptosis (Fig. 3) is a finding that has been previously described in the English literature only once⁹ and which was managed externally by osteoplastic flap approach. Another case of dermoid cyst arising from the medial part of orbit extending into the frontal sinus was reported which was excised by external incision in the frontal sinus floor.³ Two intracranial nasal dermoid excised endoscopically were reported by Schuster et al¹⁰ recently but were not involving the frontal sinus. In general, these lesions are present in childhood. Pryor et al¹¹ reviewed 49 cases of pediatric dermoid cysts and found the periorbital region to be the most common. Their study showed that 61% of cases presented periorbitally. Midline nasal and forehead dermoids accounted for only 16% of cases.¹¹ A clear understanding of several features of NDSCs is critical to the diagnosis and management of this condition. The

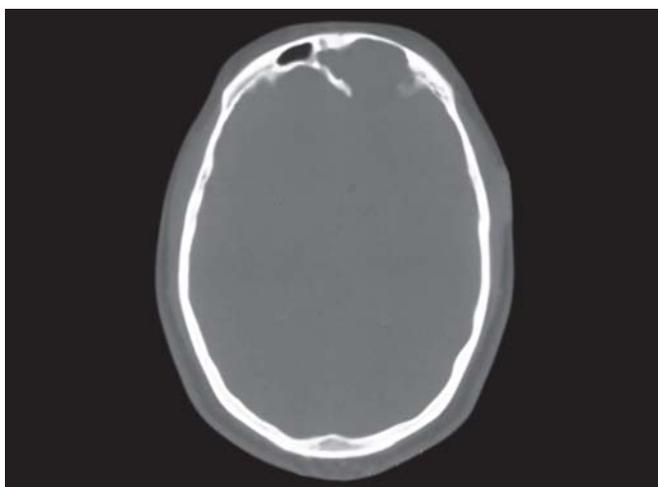


Fig. 2: Postoperative computerized tomography of nose paranasal sinus showing a disease free frontal sinus with bony remodeling



Fig. 3: Noncontrast computerized tomography of nose and paranasal sinus showing NDSC causing expansile erosion of the posterior table of left frontal sinus

objective of imaging studies is to confirm the clinical diagnosis and to delineate any intracranial involvement, if present. The complementary roles of CT and MRI appear well-established in congenital midline nasal masses. Bony detail is best defined by CT, while MRI images soft tissues more precisely.¹² Some maintain, however, that CT provides all the information required and that certain specific findings can reliably and specifically diagnose intracranial extension. Differential diagnosis of midline nasal masses includes congenital lesions, inflammatory masses and benign or malignant neoplasms. A variety of presurgical complications include osteomyelitis, cellulitis, abscess, external deformity of nose requiring rhinoplasty, CSF leak and even frontal lobe abscess.⁴

Simple biopsy could be dangerous as this may lead to CSF leak and meningitis in cases with intracranial extension. Surgical approaches to the removal of NDSC can be entirely extracranial or require a craniotomy. In one study, a variety of extracranial techniques were employed.⁷ Excision and direct primary closure has the advantage of removing abnormal skin overlying a cyst or the opening of a sinus. Incisional wounds, particularly those oriented vertically, are well tolerated. Surgical scars generally settle well, and the surgeon has an opportunity to remove dysplastic or stretched skin over a sinus (Fig. 4). Open rhinoplasty, either alone or combined with direct excision, offers the opportunity to correct the position of the alar cartilages, which are often splayed by a lesion at the nasal tip.^{2,13}

Endoscopic approaches are useful for superficial lesions with no extension that lie in the glabellar region. To avoid a big exposed scar, less time consuming surgery and to have complete surgical removal, endoscopic approach was used. An upper eyelid incision provides adequate exposure



Fig. 4: Postoperative clinical photography of the patient showing the well-healed scar of the punctum of NDSC

of most orbital lesions. Lesions invading deeply within the orbit may require a more aggressive approach. A bicoronal approach may be required for exposure, if there is extensive involvement of the frontal sinus.¹⁴ However, a combined approach, as described, was well-tolerated by the patient with excellent postoperative response. Endoscopic excision combined with sinus tract excision is adequate even with extensive frontal sinus involvement and orbital part as well.

CONCLUSION

Although NDSCs are uncommon and complex lesions, they can be managed successfully with careful clinical assessment, preoperative imaging and appropriate surgery. Tailored definitive surgery, addressing defined pathology and anatomy, allows successful treatment with a low recurrence rate and few complications. The endoscopic transnasal skull base approach combined with external elliptical incision is an excellent approach to achieve complete resection of nasal dermoid cysts expanding into the frontal sinus as well as the orbit.

REFERENCES

1. Ahuja R, Azar NF. Orbital dermoids in children. *Semin Ophthalmol* 2006;21:207-11.
2. Bilkay U, Gundogan H, Ozek C, et al. Nasal dermoid sinus cysts and the role of open rhinoplasty. *Ann Plast Surg* 2001;47:8-14.
3. Pham NS, Dublin AB, Strong EB. Dermoid cyst of the orbit and frontal sinus: A case report. *Skull Base* 2010;20(4):275-78.
4. Meher R, Singh I, Aggarwal S. Nasal dermoid with intracranial extension. *J Postgrad Med* 2005;51:39-40.
5. Charrier JB, Rouillon I, Roger G, et al. Craniofacial dermoids: An embryological theory unifying nasal dermoid sinus cysts. *Cleft Palate Craniofac J* 2005;42:51-57.
6. Pratt LW. Midline cysts of the nasal dorsum: Embryologic origin and treatment. *Laryngoscope* 1965;75:968-80.
7. Blake WE, Chow CW, Holmes AD, Meara JG. Nasal dermoid sinus cysts: A retrospective review and discussion of investigation and management. *Ann Plast Surg* 2006;57:535-40.
8. Yeola M, Johrapurkar SR, Bhole AM, Chawla M, Chopra S, Paliwal A. Orbital floor dermoid: An unusual presentation. *Indian J Ophthalmol* 2009;57:51-52.
9. Post G, McMains KC, Kountakis SE. Adult nasal dermoid sinus cyst. *Am J Otolaryngol* 2005 Nov-Dec;26(6):403-05.
10. Schuster D, Riley KO, Cure JK, Woodworth BA. Endoscopic resection of intracranial dermoid cysts. *J Laryngol Otol* 2011;125:423-27.
11. Pryor SG, Lewis JE, Weaver AL, Orvidas LJ. Pediatric dermoid cysts of the head and neck. *Otolaryngol Head Neck Surg* 2005;132:938-42.
12. Rahbar R, Shah P, Mulliken JB, et al. The presentation and management of nasal dermoid: A 30 year experience. *Arch Otolaryngol Head Neck Surg* 2003;129:464-71.

13. Rohrich RJ, Lowe JB, Schwartz MR. The role of open rhinoplasty in management of nasal dermoid cysts. *Plast Reconstr Surg* 1999;104:2163-71.
14. Van Aalst JA, Luerssen TG, Whitehead WE, et al. Keystone approach for intracranial nasofrontal dermoid sinuses. *Plast Reconstr Surg* 2005;6:13-19.

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