CASE REPORT

Median philtrum sinus: a rare presentation and review of management options

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SUMMARY

Nasal dermoids are rare congenital abnormalities, and there is only one previously reported case of a philtrum sinus tract extending to the skull base. A 2-month-old boy was presented with an incidental finding of a median philtrum sinus with no infective features and no other abnormal clinical findings. MRI demonstrated a sinus tract extending from the philtrum to the crista galli. On multidisciplinary review, a conservative approach was taken due to the asymptomatic nature of the patient. Given the rarity of nasal dermoids, diagnosis requires precise clinical examination and MRI to identify the extent of the tract. Management options require a multidisciplinary case-specific approach and include non-surgical as well as surgical approaches.

BACKGROUND

Congenital midline nasal lesions are rare, with nasal dermoids being the most common type. These are present in 1 in 20 000–40 000 live births.^{1,2} Nasal dermoids are theorised to have an embryological origin where ectodermal tissue becomes trapped and adherent to the nasal capsule, forming a tract that can extend from any midline point of the nose to the anterior cranial fossa.¹ They usually present at birth as a pit at any point on the midline of the nose, which may be associated with discharge.³ If infected, nasal dermoids extending to the anterior cranial fossa may result in serious sequelae, such as meningitis, frontal bone osteomyelitis or intracranial abscesses.⁴ Nasal dermoids with an opening in the philtrum are much less common, with only one previously reported case in the literature.⁵ Rarer still, these may be associated with medial upper lip fistulae (MULFs), which are less common, occurring in only 1 in 100 000 live births.⁶ However, unlike nasal dermoids, MULFs tend to extend to the upper lip mucosa or into the oral cavity.⁷ This is the second reported case in the literature of a paediatric nasal dermoid sinus extending from the philtrum to the skull base. Theories of pathogenesis and management strategies will be explored.

CASE PRESENTATION

A 2-month-old boy was referred by the paediatric medical unit to the otolaryngology clinic with an incidental finding of a median philtrum sinus at birth. The infant was otherwise well and had a normal delivery without any antenatal complication. There was no family history of birth defects or parental exposure to environmental factors. No

other congenital abnormalities were found at birth. There was no history of infection or discharge, and the infant had never developed any other symptoms or signs suggestive of meningitis, frontal bone osteomyelitis or intracranial infection. Clinical examination showed a small midline pit in the philtrum, which was dry (figure 1). No other congenital abnormality was found on examination.

INVESTIGATIONS

MRI of the head showed a sinus tract extending from the philtrum to the crista galli with possible extension into the anterior cranial fossa. No dural enhancement or intracranial lesion was seen (figure 2).

TREATMENT

The patient was reviewed at a multidisciplinary meeting, with a conservative approach taken due to the asymptomatic nature of the patient compared with the significant risks and potential complications associated with any curative procedure, including cosmetic outcomes.

OUTCOME AND FOLLOW-UP

Serial follow-up every 3 months in the first year has not shown any infective features. Further reviews have been planned at 24 and 36 months of age.

DISCUSSION

Nasal dermoids are rare congenital abnormalities that usually present as pits at any point in the midline of the nose. Given the rarity of this condition, a high level of clinical suspicion is required for any infant presented with a pit or swelling in the midline of the nose or philtrum. Postulated theories for the pathogenesis of both nasal dermoid cysts and MULFs include failure of ectodermal invagination and fusion, as well as failure of separation between skin and dura, resulting in the formation of a cyst along an anatomical line from the midface to the anterior cranial fossa, where ectodermal inclusion is likely to have occurred.^{8,9} If such a cyst opens to the external nasal skin, a sinus can form.^{1,5} Therefore, a potential tract may exist from the nasal skin into the anterior cranial fossa. There are currently no evidence to suggest their association with antenatal complications, other birth defects or parental environmental exposures.

Diagnosis of nasal dermoids requires precise clinical examination at birth, including the identification of other congenital abnormalities. Previous

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Figure 1 Midline philtrum sinus in a 2-month-old infant.

episodes of meningitis, frontal bone osteomyelitis or intracranial abscesses are important aspects of clinical history that will need to be considered. The previously reported case by Locke and Kubba⁵ was presented at the age of 24 months with a discharging midline philtrum pit.

The radiological modality of choice is MRI, which identifies the extent of the nasal dermoid as well as any intracranial involvement with low false-positive and false-negative rates.⁴ Due to the radiation associated with CT imaging in infants, CT scans would only be indicated if there are any contraindications to MRI or if further details regarding the bony anatomy is required for preoperative assessment.⁵

Management options include both conservative and surgical approaches. Non-surgical approaches involve the prevention of meningeal or intracranial infections with immunisations as per local guidelines. Moreover, it is important to educate the parents with regard to clinical features suggestive of meningitis or intracranial abscesses in order to expedite early medical treatment. The previously reported case by Locke and Kubba underwent surgical excision of the entire tract via an external rhinoplasty approach.^{5 10} Sinuses lower in the philtrum and closer to the upper lip may be excised via an intraoral approach as

for MULFs.⁷ Charrier *et al*⁷ reported a case of an MULF that extended to beneath the upper lip mucosa and was completely excised via an intraoral approach.

We would recommend that surgical indications include persistent discharge or previous episodes of meningitis, frontal bone osteomyelitis or intracranial abscess, with the aim of surgery to eliminate these risks. However, surgery may be associated with significant risks, such as skull base injury, cerebrospinal fluid leak, nasal and septal deformities, as well as incomplete removal of the entire tract resulting in recurrent symptoms.^{2 10 11} Moreover, recent studies have shown the negative effects that general anaesthetic can have on childhood development.^{12 13} Therefore, careful consideration of clinical history, examination and radiological findings should be made in a multidisciplinary setting to determine whether the infant may benefit from surgery.

Learning points

- ▶ Nasal dermoids are rare congenital abnormalities.
- ▶ A high level of clinical suspicion is required for any infant presented with a pit or swelling in the midline of the nose or philtrum.
- ▶ The radiological investigation of choice is MRI.
- ▶ Management requires a multidisciplinary approach and surgery is recommended for patients with persistent discharge or previous episodes of meningitis, frontal bone osteomyelitis or intracranial abscess.

Contributors Guardian consent and data were obtained by MMKK. Both MMKK and PP were involved with the preparation and editing of the manuscript.

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REFERENCES

- 1 Pratt LW. Midline Cysts of the Nasal Dorsum: Embryologic Origin and Treatment. *The Laryngoscope* 1965;75:968–80.
- 2 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.
- 3 Pryor SG, Lewis JE, Weaver AL, *et al*. Pediatric dermoid cysts of the head and neck. *Otolaryngol Head Neck Surg* 2005;132:938–42.
- 4 Blake WE, Chow CW, Holmes AD, *et al*. Nasal dermoid sinus cysts: a retrospective review and discussion of investigation and management. *Ann Plast Surg* 2006;57:535–40.
- 5 Locke R, Kubba H. A case of a nasal dermoid presenting as a median upper lip sinus. *Int J Oral Maxillofac Surg* 2011;40:985–7.
- 6 Coccia CT, Bixler D. Cleft lip, cleft palate, and congenital fistulas of the lower lip. Report of a familial occurrence. *Oral Surg Oral Med Oral Pathol* 1967;24:246–9.
- 7 Charrier JB, Rouillon I, Roger G, *et al*. Congenital isolated midline sinus of the upper lip: clinical and embryological approaches. *Cleft Palate Craniofac J* 2006;43:488–91.
- 8 Sancho MA, Albert A, Cusi V, *et al*. Upper lip fistulas: three new cases. *Cleft Palate Craniofac J* 2002;39:457–60.
- 9 Charrier JB, Rouillon I, Roger G, *et al*. Craniofacial dermoids: an embryological theory unifying nasal dermoid sinus cysts. *Cleft Palate Craniofac J* 2005;42:51–7.
- 10 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.
- 11 McCaffrey TV, McDonald TJ, Gorenstein A. Dermoid cysts of the nose: review of 21 cases. *Otolaryngol Head Neck Surg* 1979;87:52–9.
- 12 Backeljauw B, Holland SK, Altaye M, *et al*. Cognition and Brain Structure Following Early Childhood Surgery With Anesthesia. *Pediatrics* 2015;136:e1–e12.
- 13 Bakri MH, Ismail EA, Ali MS, *et al*. Behavioral and emotional effects of repeated general anesthesia in young children. *Saudi J Anaesth* 2015;9:161–6.

Figure 2 (A,B) MRI scan showing a sinus tract extending from the philtrum to the crista galli.

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