CASE REPORT

Epidermoid cyst of the soft palate in an infant

Divya Uppala, Sumit Majumdar, Kameswara Rao, Sivanagendra Reddy
Department of Oral Pathology, GITAM Dental College, Department of Oral Surgery, Anil Neerukonda Institute of Dental Sciences, Visakhapatnam, Andhra Pradesh, India

Address for correspondence:
Dr. Divya Uppala,
S5, Meher Apartment, CBM Compound, VIP Road, Visakhapatnam - 530 003, Andhra Pradesh, India.
E-mail: uppala.divya@gmail.com

Received: 14-11-2014
Accepted: 22-12-2015

INTRODUCTION

Epidermoid cysts are benign developmental malformations arising from abnormal epithelial constituents of ectodermal tissue formed during the fetal period.[1,2] These lesions can be seen anywhere in the body, with the occurrence of approximately 7% in the head and neck region[1,2] and their incidence in the oral cavity makes up for 1.6% of the total occurrences[1,2] and they constitute <0.01% of all the cystic lesions of the oral cavity.[1] Epidermoid cysts of the oral cavity, in adults, are mostly seen on the floor of the mouth and in other locations including the labial,[2] palatine tonsil[3] and in the soft palate.[1] In infants incidence is highest in the floor of the mouth and lowest in the soft palate. Only four cases have been reported in literature of epidermoid cysts in infants involving the soft palate.[4-6] We present here a report of a 9-month-old female patient who underwent surgery for cleft palate and growth on the uvula of soft palate clinically diagnosed as a benign fibrous tumor which was confirmed by histology as epidermoid inclusion cyst.

CASE REPORT

A 9-month-old Indian infant was brought to the cleft care center for a surgery of cleft palate and associated growth in the soft palate. The swelling was noted 3 months earlier by the parents. The infant had difficulty in swallowing milk and breathing. Family history was not significant and her mother had an uncomplicated delivery. On physical examination, the patient was alert and intraoral examination revealed a cleft palate involving the soft palate and a whitish, oval shaped, solitary solid mass with pale overlying mucosa located behind the uvula and right side of the soft palate measuring approximately about 1 cm × 1 cm [Figure 1]. No other external facial or neck cysts, sinuses or lesions were noted and the rest of the intraoral space was unremarkable. A provisional diagnosis of benign fibrous tumor was considered and differentials included salivary gland neoplasms and benign tumors of muscular origin. The patient was admitted to the hospital for the surgery of cleft palate as the medical history was irrelevant. Surgery was performed under general anesthesia, surgery was uneventful, and the patient recovery was good. The soft tissue was excised completely and sent for histopathological examination. Macroscopically, the specimen appeared encapsulated and was creamish white with a soft consistency. Histopathological examination revealed a parakeratinized stratified squamous epithelium with flattened rete ridges and a cystic space with keratin flecks. Skin appendages were absent [Figures 2a-d and 3-6]. Based on the above findings, epidermoid cyst was diagnosed. The infant made...
a full recovery and was discharged after 1 week. The patient is under follow-up and there is no evidence of recurrence even after 1.5 years.
DISCUSSION

A thorough search using the keywords such as “soft palate,” “pediatric/congenital,” and “epidermoid cyst/dermoid cyst” in various combinations was made in PubMed. The ages of the patients described were between neonate and 62 years old. One patient had a dermoid cyst while the remaining patients had epidermoid cysts.

The most common location of the cysts was on the uvula. A keyword search using “epidermoid cyst, palate, cleft” in the PubMed literature revealed 10 cases, but none of them were associated with a cleft soft palate Table 1.

Dermoid cysts were classified by New and Erich in 1937[7] as acquired implantation, congenital teratoma and congenital inclusion dermoid cysts and by Meyer in 1955 as true dermoid cysts, epidermoid cysts and teratoid cysts.[8] A true dermoid cyst is lined with keratinized epithelium and has skin appendages like hair follicles or sebaceous glands. An epidermoid cyst is lined with simple squamous epithelium and does not contain skin appendages. A teratoid cyst in addition to skin appendages contains other tissues such as muscle, bone and cartilage.[2] According to New and Erich,[7] who conducted a study on 1495 cases, the common location was anal region (44.5%) followed by ovarian (42.1%) region. The overall incidence of pediatric head and neck cysts was 7%, with the periorbital region being the most common site. This is equally consistent with the study by Pryor et al.[9] which concentrated on pediatric dermoid cysts of the head and neck, in which 61% of head and neck dermoid cysts were periorbital in location. In neither of these series, were cysts of the soft palate mentioned.

Dermoid cysts and epidermoid cysts can be acquired or congenital,[7] but in infants, they are usually congenital.

Table 1: Reported cases of epidermoid cysts of soft palate and uvula

| References     | Age/sex  | Year | Site          | Diagnosis        |
|----------------|----------|------|---------------|------------------|
| Green, Neal[1] | 18 years/female | 1982 | Midline of Dermoid cyst soft palate | Epidermoid cyst |
| Pruszewicz et al.[1] | Neonate | 1984 | Uvula         | Epidermoid cyst  |
| Yoshinari et al.[1] | 12 months/female | 1986 | Uvula         | Epidermoid cyst  |
| Zappia et al.[1] | 62 years/male | 1991 | Soft palate   | Epidermoid cyst  |
| Sechul et al.[1] | 1.12 months/male | 1998 | Uvula         | Epidermoid cyst  |
| Muramatsu et al.[1] | 3.3 months/male | 2002 | Uvula         | Epidermoid cyst  |
| Caylakli et al.[9] | 1 month/male | 2005 | Soft palate   | Epidermoid cyst  |
| Suga et al.[1] | 1 month/male | 2010 | Uvula         | Epidermoid cyst  |
| Tsai et al.[7] | 7 months/male | 2013 | Uvula         | Epidermoid cyst  |
| Alcorn KM[15] | 2 years/female | 2014 | Uvula         | Epidermoid cyst  |

During the 6th week of gestation, the secondary palate begins to develop.[10‑15] It starts initially as two outgrowths from the maxillary prominences which are oriented in a vertical direction and layout on either side of the tongue. During 8th week, these palatine shelves orient themselves in a horizontal direction and the tongue descends downward as mandible elongates and the fetal head tilts upward.[10‑14] The palatal shelves continue to grow toward each other and contact at the medial edge epithelia. A transient midline epithelium or midline seam is formed at this time and as the growth of head increases the seam thins into a single layer of cells and undergoes disintegration completely resulting in the merging of the mesenchymal portion of two palatal shelves by the process of fusion.[11‑12] Lack of breakdown of the epithelium leads to clefts.[12] This process of shelf elevation and fusion takes place about a week later in girls than in boys which explain why girls are more prone than boys for cleft palate formation.[12] In contrast, after the fusion of palatal shelves the soft palate uvula forms through migration and proliferation of two confluent sub epithelial mesenchymal growth centers at the posterior edge of the newly formed palate with a groove between them filled by merging.[12‑14] Failure of the merging process during soft palate and uvula development can result in complete or partial clefts of the soft palate and uvula,[13] and the remnants in the midline may become cystic.[1,18] Hence, in distinction to the previous epidermoid cysts that were reported on the soft palate, in the present case cleft of secondary palate might have developed due to lack of fusion and merging; and the epidermoid cyst would have developed due to aberrant ectodermal entrapment in the uvular area followed by reactivation of these cleaved cells. Treatment for these lesions is surgical excision of the cyst. It should be excised completely and recurrence after surgery is rare.

CONCLUSION

Identification of epidermoid cyst is essential in neonates as they may cause difficulty and obstruction in breathing and swallowing which might turn out to be fatal.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Suga K, Muramatsu K, Uchiyama T, Takano N, Shibahara T. Congenital epidermoid cyst arising in soft palate near uvula: A case report. Bull Tokyo Dent Coll 2010;51:207-11.
2. Levi B, Brugman S, Wong VW, Grova M, Longaker MT, Wan DC. Palatogenesis: Engineering, pathways and pathologies. Organogenesis 2011;7:242-54.
3. Erol K, Erkan KM, Tolga D, Bengu C. Epidermoid cyst localized in the palatine tonsil. J Oral Maxillofac Pathol 2013;17:148.
4. Caylakli F, Yavuz H, Bolat F, Ozer F, Ozgirgin N. Epithelial cyst of the soft palate. Int J Pediatr Otorhinolaryngol 2005;69:545-7.
5. Tsai WC, Kuo CY, Wang CH. Epidermal inclusion cyst of the soft palate and uvula in an infant. Eur J Pediatr 2013;172:1563-4.
6. Alcorn KM, Pollock AN, Kazahaya K, Solot CB, Jackson O. Epidermoid cyst of the soft palate in a 2-year-old: A case report and review of literature of pediatric soft palate lesions. Clin Pediatr (Phila) 2014;53:1399-402.
7. New G, Erich J. Dermoid cysts of the head and neck. Surg Gynecol Obstet 1937;65:48-55.
8. Meyer I. Dermoid cysts (dermoids) of the floor of the mouth. Oral Surg Oral Med Oral Pathol 1955;8:1149-64.
9. Pryor SG, Lewis JE, Weaver AL, Orvidas LJ. Pediatric dermoid cysts of the head and neck. Otolaryngol Head Neck Surg 2005;132:938-42.
10. Nawshad A. Palatal seam disintegration: To die or not to die? That is no longer the question. Dev Dyn 2008;237:2643-56.
11. Peterson-Falzone SJ. Cleft Palate Speech. 4th ed. Missouri, U.S.A.; MOSBY Elsevier; 2001. p. 29-34.
12. Achallii S, Bhat S, Ram Shetty S, Babu SG, Suvarna R. Deformities of the uvula in the oral cavity – A case series. Iran Red Crescent Med J 2012;14:676-9.
13. Burdi AR, Faist K. Morphogenesis of the palate in normal human embryos with special emphasis on the mechanisms involved. Am J Anat 1967;120:149-60.
14. Moxham BJ. The development of the palate – A brief review. Eur J Anat 2003;7 Suppl 1:53-74.
15. Alcorn KM, Pollock AN, Kazahaya K, Solot CB, Jackson O. Epidermoid cyst of the soft palate in a 2-year-old: A case report and review of literature of pediatric soft palate lesions. Clin Pediatr (Phila) 2014;53(14):1399-402. doi: 10.1177/0009922814543325.