CASE REPORT

Epidermoid cyst of the soft palate in an infant

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ABSTRACT

Epidermoid cysts are benign malformations that can be encountered anywhere in the body and are rarely observed in the oral cavity accounting for <0.01% of all cysts of the oral cavity. They can be classified as either congenital or acquired without any clinical or histologic differences. Our literature search did not find any report of a congenital epidermoid cyst located in the soft palate associated with a complete palatal cleft in an infant. This is a case report of a 9-month-old female patient who had a cleft palate with an associated soft tissue mass at the junction of soft palate and uvula.

Key words: Cleft palate, epidermoid cyst, uvula, soft palate

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.[3] 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.[1,4-6] 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

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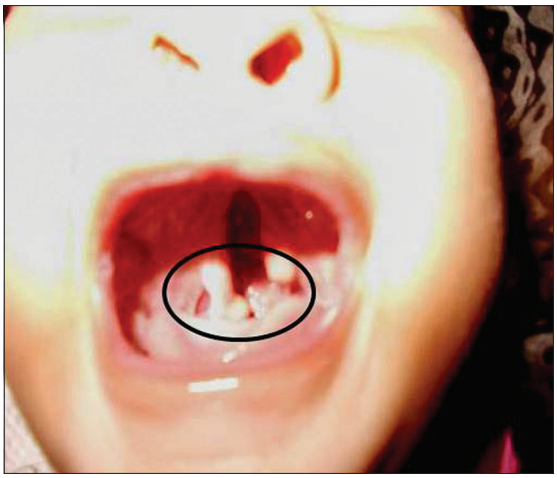

Figure 1: Clinical image revealing cleft palate with a whitish, oval shaped, solitary solid mass with pale overlying mucosa located behind the uvula and right side of the soft palate

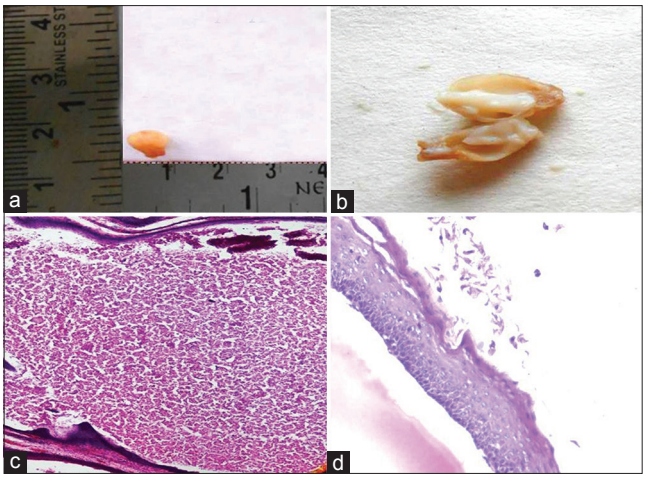

Figure 2: (a) Gross specimen creamish white in color and soft in consistency; (b) Cut section of the gross specimen showing cystic space; (c) Photomicrograph demonstrating the cystic cavity containing keratin (H&E stain, ×40) (d) Photomicrograph demonstrating parakeratinized stratified squamous epithelium with flattened rete ridges and the cystic space with keratin flecks (H&E stain, ×100

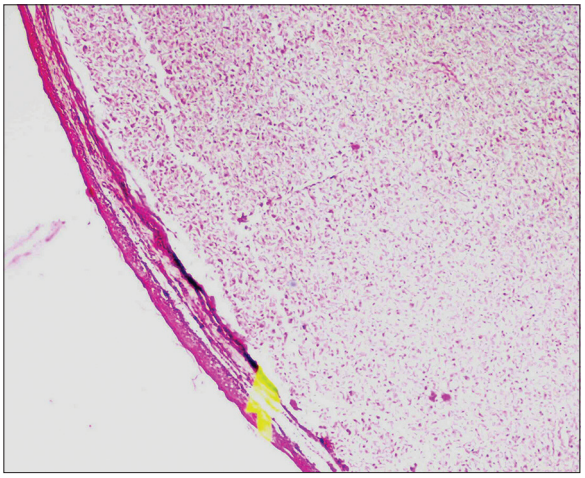

Figure 3: Histopathological image demonstrating epithelium and keratin (H&E stain, x40)

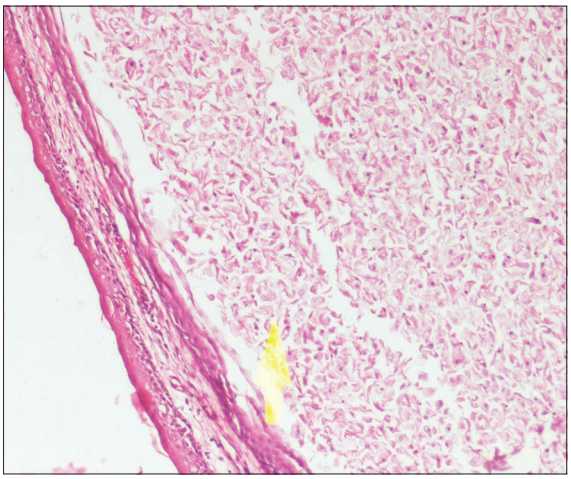

Figure 4: High power view of the epithelium and keratin in the lumen (H&E stain, x100)

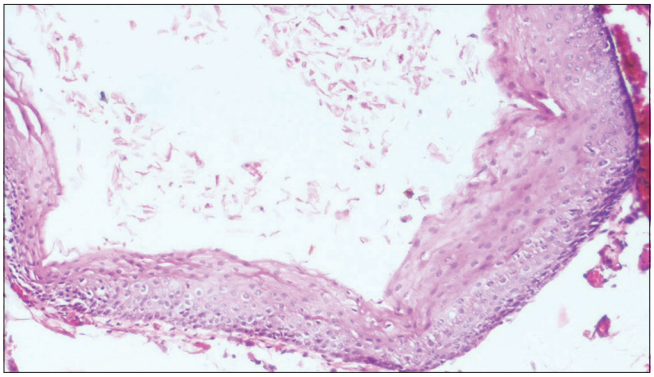

Figure 5: Photomicrograph showing keratinizing epithelium and keratin flecks in the lumen (H&E stain, 100) Histopathological figure at ×10

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.

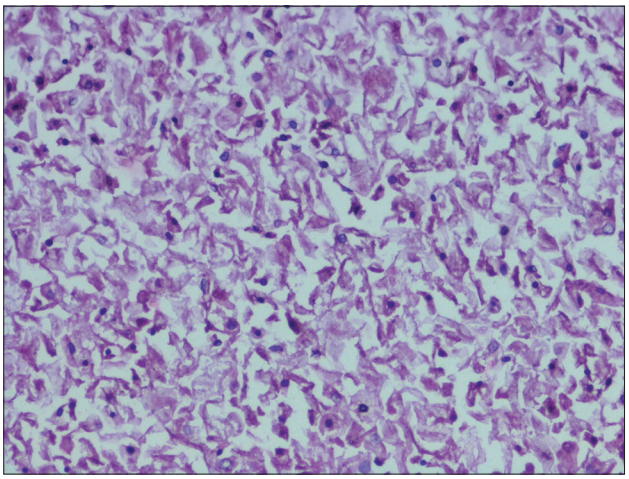

Figure 6: Photomicrgraph showing keratin flecks (H&E stain, x200)

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 "ëpidermoid 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 soft palate	Dermoid 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 2.12 months/male 3.3 months/male	1998	Uvula Uvula Uvula	Epidermoid cyst
Muramatsu *et al*.[1]	Neonate/male 1 month/female	2002	Uvula Uvula	Epidermoid cyst
Caylakli *et al*.[6]	6 years/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.[10,11] 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 confluenced 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.[1,15] 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.

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Conflicts of interest

There are no conflicts of interest.

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