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Case Report

Palatal teratoma associated with cleft palate- A rare case report

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ABSTRACT

Teratomas are benign tumours that are commonly seen in the sacrococcygeal region and in ovaries. It consists of cells from the layers of ectoderm, endoderm and mesoderm. Localised teratomas in the head and neck region are rare in origin. It can be in the size of a small polyp or it may grow as an extension to the skull base. This rare case report highlights the presence of congenital teratoma in the palatine region of a child. The palatine processes and the median nasal septum have failed to fuse, thus leading to a development of cleft palate in the child. This failure of fusion may be related to the presence of the epignathus obstructing the oropharyngeal connection. The diagnosis of teratoma was confirmed radiologically and histopathologically. The lesion was surgically excised and the cleft palate defect was corrected in a single stage surgical procedure.

1. Introduction

Epignathus is a rare form of teratoma. It originates from the base of the skull which usually attaches to the hard palate or the mandible.[1] The word teratoma initially originated from the Greek word teraton which means 'monster'. This term was coined by Virchow in 1863.[2,3] It is described as a neoplasm that originates from three germ layers namely the ectoderm, mesoderm and the endoderm. Teratoma is rarely seen in the head and neck region and it is even more uncommon to spot one in infants.[4] Among head and neck teratomas the most comments site is there cervical region and second to it; is in the oropharynx region which is termed as epignathus.[1]

The epignathus, subjects to high mortality rate due to its pathologic characteristics of size and site which leads to obstructive airway in an infant.[5] Abnormal growths of size 9cm and more are usually detected in the ultrasound fetal scans in the 15th - 17th week. Prenatal scanning and diagnosis of masses in the oropharyngeal region helps to understand the threats posed in airway and aids in surgical planning well ahead of child birth.[6] This case report highlights the presence of a palatal teratoma associated with unilateral right cleft lip and complete primary cleft palate in an 1-year-old child since the time of birth.

2. Case Report

A 1-year-old male child weighing 10 kg was brought to our Plastic and reconstructive Surgery department for repair of his cleft palate. The mother underwent an anomaly scan during the fifth intrauterine life and the presence of a growth in the palate of the foetus was notified to the parents. The child was born through caesarean section and was intubated to combat the respiratory distress due to the presence of a mass in the palate. The child also presented

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with a right unilateral complete cleft lip and cleft palate with the presence of an epignathus. The tumour showed a minimal increase in size post-birth period. The child underwent cleft lip repair at a private institution. The child was examined and worked up for cleft palate repair. We studied the presence of the palatal growth through 3D Facial bone Computed Tomography (Figure 2). The mass was identified in coronal and axial sections, where the mass was located beneath the cleft palate (Figure 1a,b). The surgical plan was to excise the palatal growth and repair the cleft palate in layers. Bardach's Two flap palatoplasty was performed to repair the cleft palate. The palatal growth was excised as well and sent for histopathology. The child was extubated post-surgery and recovered well.

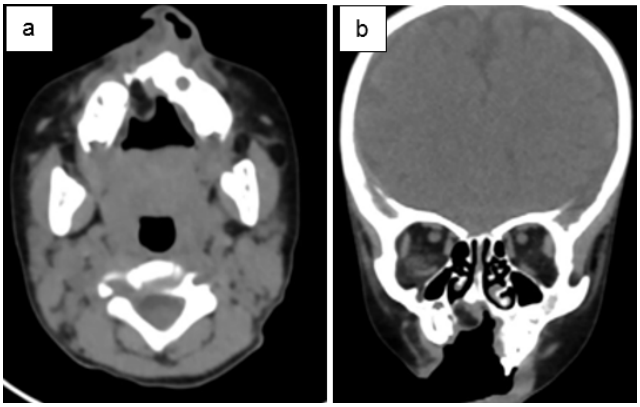

Fig. 1: a: Axial section showing a mass approximately 4×3×2cm in the right palatal vault; **b:** Palatalteratoma abutting the right nasal floor.

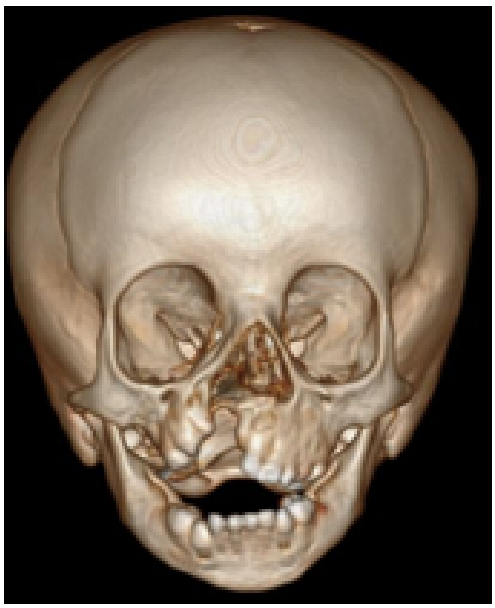

Fig. 2: 3D reconstructive image of the facial bone.

3. Discussion

The given specimen was reported with the presence of squamous epithelium and islands of fibro-muscular composite cells leading to an impression as Teratoma of the palate. These uncommon malformations in relation to oral cavity arises from three germ layers- ectoderm, endoderm and mesoderm which are embryonic in origin.[7] The most understood pathophysiology of this neoplasm maybe due to the failure of fusion during embryogenesis or incomplete migration and or implantation of pluripotent cells during the fourth-fifth intra-gestational life.[8,9] The classification of Teratomas are into four types namely, Dermoids (hairy polyps), Teratoid, True teratoma and Epignathus. Epignathus (fetus en fetu) are highly differentiated teratomas and have a high mortality rate.[10,11] Teratomas occur at the ratio of 1 in every 4000 births with a slight female predilection. These are mostly benign but malignant cases have also been reported.[12] In the presence of a teratoma it is common to be associated with an anomaly such as cleft palate, cleft lip, bifid nose or tongue due to the interference with fusion of palatine shelves in the midline. It also leads to other maxillofacial abnormalities.[13] In our case, the child had right unilateral cleft lip, right cleft alveolus, right complete cleft palate along with abnormalities in his nasal framework.

Almost every pregnant woman undergoes prenatal ultrasound screening from their 12th week of pregnancy. Ultrasounds usually detect mass oral abnormalities in a foetus from its 15th -17th week. The need to identify such mass growing tumours in the oral cavity, is because they pose life threatening complications in terms of respiration in a neonate. A case report by Kontopoulos had performed an intrauterine treatment of an oral teratoma using YAG laser. The child was born and did not face any respiratory complications and after surgery, the baby was born without any complications.[6] Other conditions that have to be ruled out as differential diagnosis are encephalocele, cystic hygroma, rhabdomyosarcoma and haemangioma. The rate of recurrence is quite low but there is evidence of literature in terms of recurrence of the tumour. Cases reported by April show recurrence in terms of inadequate resection, which is detected by asymmetric elevation of serum AFP levels. In malignant cases there is evidence of administration of radiotherapy and chemotherapy after complete surgical resection.[8]

Complete surgical resection is the key to improved prognosis in a child, born with oral teratoma. The major concern for surgery in a neonate, is to remove the obstructive tumour from its naso-oropharyngeal airway and to establish adequate respiration. Another important feature that is not to be neglected in a patient with oral teratoma, is the extensions of the tumour. A thorough radiological examination is to be carried out to delineate any intracranial extensions. Cases of intracranial extension of epignathus

with or without brain invasion have been put forth in literature by Y.Hosoda and Kirishima et al.[14] In our case the child did not have any intracranial extensions. The epignathus was completely removed before addressing the cleft palate. The excised mass was sent to histopathological evaluation. The cleft palate was repaired in the same surgery using Bardach's two flap palatoplasty technique. The buccal pad of fat was harvested from the left side to cover the palatal raw area on the same side. The child was extubated after the completion of the surgery and he recovered asymptomatically.

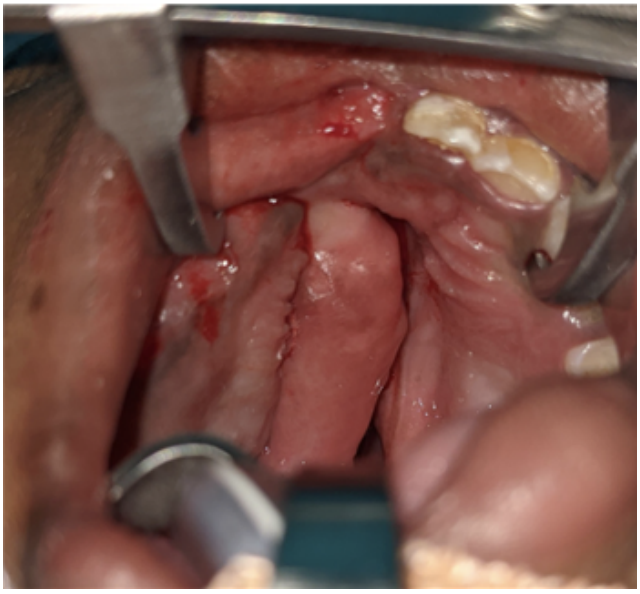

Fig. 3: Complete cleft of primary palate with palatal growth in the mid-palatine region along thevomer.

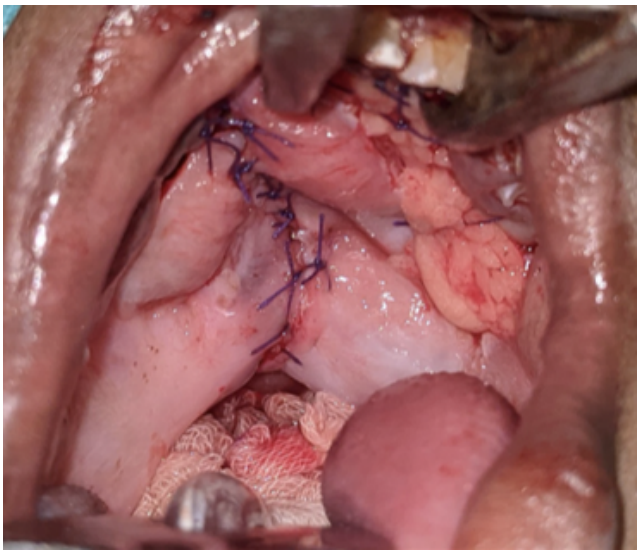

Fig. 4: Palatal repair by Bardach's two flap palatoplasty technique.

4. Conclusion

This case is reported to the literature to implement the importance of epignathus in infants due to the threat it poses to the airway. Intrauterine diagnosis is the key to eliminate the risk of airway obstruction. Oropharyngeal teratoma are usually accompanied with cleft lip and or palate. Appropriate counselling to the parents help in advance planning and benefits the rate of survival of the child. This child is currently 8 years old and is planned for lip-nose revision (LNR) surgery.

5. Source of Funding

None.

6. Conflict of Interest

None.

References

1. Jadhav SS, Korday CS, Malik S. Epignathus Leading to Fatal Airway Obstruction in a Neonate. *J Clin Diagn Res*. 2017;11(1):SD04–5. doi:10.7860/JCDR/2017/24956.9283.
2. Shah A, Latoo S, Ahmed I, Malik A. Head and neck teratomas. *J Maxillofac Oral Surg*. 2009;8(1):60–3. doi:10.1007/s12663-009-0015-x.
3. Chauhan DS, Guruprasad Y, Inderchand S. Congenital nasopharyngeal teratoma with a cleft palate:case report and a 7 year follow up. *J Maxillofac Oral Surg*. 2011;10(3):253–6. doi:10.1007/s12663-010-0140-6.
4. Halterman S, Igulada K, Steinicki E. Epignathus: Large Obstructive Teratoma Arising From the Palate. *Cleft Palate Craniofac J*. 2006;43(2):244–6.
5. Akhtar R, Riffat M, Mehmood Z. Epignathus Teratoma. *J Coll Physicians Surg Pak*. 2016;26(5):438–40.
6. Kishi Y, Soh S, Tanaka S, Nitta A, Matsushima A. A Case of Congenital Teratoma with Cleft Palate in A Neonate. *Arch Clin Med Case Rep*. 2020;4:746–53.
7. Azizkhan RG, Haase GM, Applebaum H, Dillon PW, Coran AG, King PA, et al. Diagnosis, management, and outcome of cervicofacial teratomas in neonates: a children's Cancer Group study. *J Pediatr Surg*. 1995;30(2):312–6. doi:10.1016/0022-3468(95)90580-4.
8. Becker S, Shön R, Gutwald R, Otten JE, Maier W, Hentschef R, et al. A congenital teratoma with a cleft palate: report of a case. *Br J Oral Maxillofac Surg*. 2007;45(4):326–7. doi:10.1016/j.bjoms.2005.11.007.
9. Veyssière A, Streit L, Traoré H, Bénateau H. Cleft palate caused by congenital teratoma. *Paediatr Int Child Health*. 2017;37(1):66–9. doi:10.1179/2046905515Y.0000000057.
10. Arnold J. Ein Fall von Congenitalem Zusammengesetztem Lipom der Zunge und des Pharynx. *Virchows Arch*. 1870;50:482–512. doi:10.1007/BF01948784.
11. Mutombo FL, Kikuhe JN, Mwanamolo NK, Erickson MH, Wetzig N, Kakule MK, et al. Congenital epignathus associated with a cleft palate: a case report. *J Med Case Rep*. 2021;15:422. doi:10.1186/s13256-021-03007-w.
12. Al-Mahdi A, Al-Khurrhi L, Atto GZ, Dhaher A. Giant epignathus teratoma involving the palate, tongue, and foor of the mouth. *J Craniofac Surg*. 2013;24(1):e97–9.
13. Benouaiche L, Couly G, Michel B, Devauchelle B. Diagnostic et prise en charge des tératomes cervico-faciaux congénitaux: à propos

de quatre cas, revue de la littérature et mise au point. *Ann Chir Plast Esth*. 2007;52(2):114–23. doi:10.1016/j.anplas.2006.08.007.
14. Kirishima M, Yamada S, Shinya M, Onishi S, Goto Y, Kitazono I, et al. An autopsy case of epignathus (immature teratoma of the soft palate) with intracranial extension but without brain invasion: case report and literature review. *Diagn Pathol*. 2018;13(1):99. doi:10.1186/s13000-018-0776-y.

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