Teratoma Tongue : Case report and review of literature

Manish Gupta • Neena Chaudhary • A. K. Rai

Abstract

Teratomas are true neoplasms composed of tissues from all three germinal layers and may exhibit variable levels of maturity. They have an unknown origin and eccentric microscopic appearance. Teratomas arising from the oral cavity are rare in the newborn; only 22 cases¹ have been reported in the literature. We describe a female neonate with an oral teratoma originating from the tongue that was successfully treated with surgery.

Keywords Oral teratoma • Congenital.

Introduction

Teratomas are defined as true neoplasms composed of multiple tissues foreign to the site from which they originate. They are the most common extra-gonadal germ-cell tumors of childhood consisting of tissues from at least two of the three germ layers. These tumors are most often benign in their histology, but result in a high degree of morbidity and possible mortality by virtue of their size and location, causing airway obstruction and respiratory distress.

They arise most commonly in a midline or paraxial location from the brain to the sacral area and are most often located in

the sacrococcygeal region. Ninety percent of head and neck teratomas present during the neonatal and infantile period, predominantly involving the neck and nasopharynx and occurring only once in every 20,000- 40,000 births². Teratomas rarely originate from other superficial facial structures of the head.

Of the head and neck teratoma, pure oral presentation is especially rare, with most of the cases involving the tongue¹ (Table 1). Our case is 23rd case of oral teratoma and it also arose from tongue.

Case report

A 3-month-old girl presented with swelling on the undersurface of tongue. The tongue was protruding out. Her mother reported that the lesion was congenital and had remained unaltered since her birth. The baby did not had difficulty in feeding or breathing the prenatal and perinatal courses were uncomplicated and family medical histories were noncontributory. On examination (Fig.1) approx. 2.5 cm X 2.5 cm X 2.5 cm sessile, cystic mass was seen present in ventral surface of tongue. Overlying tissue was not discolored and regional lymphadenopathy was absent. Computed tomography (Fig.2) in coronal, axial and reformatted sagittal section revealed a 2.5 cm X 2.5 cm cystic anterior lingual structure primarily in midline. The wall of the cyst was thin and regular and the contents consisted of homogeneous fluid. There was no bone involvement.

With a clinical diagnosis of a dermoid cyst an excisional biopsy was performed. Under general endotracheal anaesthesia a retention suture was placed in the lingual tip, and a midline incision was made at ventral surface of tongue over the mass. Submucosal dissection allowed delivery of a well-encapsulated cyst (Fig. 3), which was separated from the genioglossus and hyoglossus muscles. The wound was closed in two layers. Blood loss was minimal and the post-operative course was uneventful. Feeding was resumed without difficulty following day.

Manish Gupta (✉)
Senior Resident

1217 Govt. Medical College Hospital Campus,
Sect. 32-B, Chandigarh-160030.
Mob: 9915025819
Email: mg1217@humlog.com

Neena Chaudhary
Specialist

A. K. Rai
Consultant & Head,

Department of Otorhinolaryngology
Vardhaman Mahavir Medical College and Safdarjang Hospital
New Delhi 110029 India

The surgical specimen measured 2.5 cm X 2.5 cm containing caseous material and hair. Histopathologic findings consisted of cyst wall lining of orthokeratinized stratified squamous epithelium with sebaceous glands, blood vessels and muscle observed in the underlying connective tissue a diagnosis of teratoid cyst was made. No evidence of malignant transformation was noted. One year after surgical removal of the lesion, there are no signs of recurrence. Alpha-fetoprotein (AFP) levels were normal.

Discussion

Teratomas occur in 1 out of 4,000 live births³. Head and Neck occurrence is generally localized to the neck and nasopharynx and comprises 1 to 10% of cases. Other extracranial presentations of teratoma are very rare and only 22 cases of pure oral teratomas have been reported in the literature. The histogenesis of teratoma formation remains debatable. The most popular theory suggests that presumably due to alterations of cellular membrane chemistry, teratomas arise from totipotential embryonic tissues that are somehow displaced during ontogeny. This leads to an assemblage of tissues often alien to the site in which they arise⁴. The synchronous presence of embryonal, fetal and adult elements is possible and identifies the level and type of differentiation. Immaturity should not be equated with malignancy and usually correlates with the immaturity of the host.

Among the classification systems describing teratomas, Arnold's system that was proposed in the late 19th century is still the most widely recognized and accepted one:

- Dermoid tumors: The most common form of teratoma, composed of ectoderm and mesoderm.
- Teratoid tumors: Poorly differentiated lesions that are composed of three germinal layers.
- True teratomas: Histologically identifiable tissue from all three germ layers. They may be solid or cystic.
- Epignathus: Also known as "fetus in fetu" or "parasitic fetus", contains fetal organs. Epignathus is a misnomer and its etymological meaning is "upon the jaw". However, it has been used for almost every teratoma of the oral cavity and pharynx tissues as well as for teratomas protruding from the mouth.

It is reported that more than 90% of oral teratoma are diagnosed during the perinatal period. Ultrasonography is useful in prenatal diagnosis⁵, particularly for oropharyngeal teratomas that interfere with fetal swallowing of amniotic fluid. Polyhydramnios is the most characteristic finding associated with fetuses with head and neck teratomas, but it is especially seen in relation to large cervical and nasopharyngeal teratomas and is not clearly described in pure oral teratomas. When a congenital oral teratoma is suspected, the child should be electively delivered via cesarean section. If an oral airway fails to ensure the airway,

Fig. 1 Cystic mass in anterior tongue ventral surface.

Fig. 2 Sagittal reformatted computed tomogram showing cystic mass in anterior tongue.

Fig. 3 Midline incision and submucosal dissection on ventral surface of tongue showing a well-encapsulated cyst.

oral or nasal endotracheal intubation or, if necessary, tracheostomy must be performed⁵. In our case no antenatal ultrasonography report was available, thus the mass was detected only after delivery. Children with pure oral teratomas seem to have less dramatic respiratory behaviour compared

with children with other head and neck presentations. When the oral teratomas grow, they tend to protrude outside of the mouth, rather than posteriorly toward the oropharynx. Because newborns are obligate nasal breathers, obstruction of the oral compartment is relatively less important and usually causes less urgent feeding problems. In addition, children with oral teratomas have been reported to associate anomalies including cleft palate, cystic hygroma and other multifocal teratomas. In our case, no associated anomalies were found.

A differential diagnosis of the mass should be done. Ultrasonography establishes the presence of solid and cystic tissue components. Plain films and Computed tomogram are useful in demonstrating virtually diagnostic calcifications. Because of their avascular character, teratomas do not enhance with administration of contrast material and thus can cause diagnostic confusion with choristomas, endodermal sinus tumors and granular cell tumors. Teratomas of the floor of mouth may imitate thyroglossal duct cysts². Also, there are number of lesions⁷ found in the floor of the mouth that closely resemble this lesion clinically. These are ranula, unilateral or bilateral blockage of wharton's duct, thyroglossal duct cyst, cystic hygroma, branchial cleft cyst, acute infection or cellulites of the floor of the mouth, infection of the submaxillary or sublingual glands, benign and malignant tumors of the floor of the mouth and adjacent

salivary glands and normal fat mass in the submental area. To these must be added the dysontogenetic cysts and aberrant thyroid tissue.

Early resection is advised, especially for airway obstructing teratomas. Failing to diagnose or treat head and neck teratomas until late adolescence or adulthood causes a risk of malignant degeneration up to 90%. Although this possibility is not clear for oral teratomas, early surgical intervention is highly advisable even if the mass is small and the patient is free of respiratory or feeding problems. Because an oral teratoma is well defined, complete excision is usually possible, as was accomplished in our case. Recurrences are rarely seen in head and neck teratomas. Our patient was free from any recurrences at the end of the 5-year follow up period. In malignant teratomas, radiotherapy and chemotherapy are used after complete extirpation of the tumor⁶. Alpha-fetoprotein (AFP) has been shown to be a reliable indicator of disease activity, and some authors advocate investigating teratoma recurrence by doing serial serum AFP levels, April et al⁶, suggested that these measurements should be continued for patients with extensive lesions or those without a capsule that were difficult to resect. Measurement of serum beta- HCG during the follow-up has been advised. In our case, all of the AFP measurements were within the normal range.

Table 1 Summary of the clinical data of latest 10 cases of oral teratomas described in the English-language literature

Case No.	Sex	Age at diagnosis	Size (cm)	Location	Treatment	Follow-up
1 Ekici	M	Prenatal	9 X 11	Hard palate	Dead 2 min after cesarean	Dead
2 April	F	Birth	-	Palate*	Surgery	1 year
3 Uchida	F	Birth	7 X 5	Tongue	Surgery	2 years
4 Wakhlu	M	Birth	-	Hard palate	Surgery	-
5 Yoon'	F	Prenatal	12 X 6	Tongue	Preg. terminated at 28 weeks	Dead
6 Selimoglu	F	Birth	13	Tongue	Surgery	2 years
7 Liang	F	Birth	4.5 X 2.5	Hard palate	Surgery	1 year
8 Cay	M	Birth	6 X 4	Hard palate	Surgery	1 year
9 Lopes	F	9 years	3.0 X 1.5	Buccal mucosa	Surgery	1 year
10 Gupta	F	3 months	2.5X 2.5	Tongue	Surgery	5 year

* parapharyngeal and skull base extension.

References

1. Lopes MA, Pereira CM, Perez DE, Vargas PA, de Almeida OP. Benign teratoma of the buccal mucosa in a 9-year-old girl: Report of case and review of literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2005 Nov; 100(5): 598-602.
2. Azizkhan RG, Haase GM, Applebaum H. Diagnosis, management and outcome of cervicofacial teratomas in neonates: a children's cancer group study. *J. Pediatr Surg* 1995; 30: 312-316.
3. Cay A, Bektas D, Imamoglu M, Bahadir O, Cobamoglu U, Sarihan H. Oral teratoma: a case report and literature review. *Pediatr Surg Int*. 2004 Apr; 20(4): 304-8.
4. Batsakis JG. Pathology consultation. Nomenclature of developmental tumors. *Ann Otol Rhinol Laryngol* 1984; 93: 98-99.
5. Raveh E, Papsin BC, Farine D. The outcome after perinatal management of infants with potential airway obstruction. *Int J Pediatr Otorhinolaryngol* 1998; 46: 207-214.
6. April MM, Ward RF, Garelick JM. Diagnosis, management and follow-up of congenital head and neck teratomas. *Laryngoscope* 1998; 108: 1398-1401.