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CASE REPORT

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Oral teratoma: a case report and literature review

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Abstract Teratomas are true neoplasms composed of tissues from all three germinal layers. They have an unknown origin and eccentric microscopic appearance. Teratomas arising from the oral cavity are rare in the newborn; only 13 cases have been reported in the literature. We describe a male neonate with an oral teratoma originating from the anterior hard palate that was successfully treated with surgery.

Keywords Oral teratoma · Congenital · Children · Review

Introduction

Teratomas are defined as true neoplasms composed of multiple tissues foreign to the site from which they originate. 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 births [4]. Teratomas rarely originate from other superficial facial structures of the head.

Of the head and neck teratomas, pure oral presentation is especially rare. We report a neonate with a teratoma that originated from the hard palate and that was causing feeding problems. We also review oral teratomas reported in the literature, along with current diagnosis and management approaches (Table 1). We have excluded teratomas arising from the oropharynx or from both the oral cavity and other anatomical structures, as they have different clinical behaviors.

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Case report

A 3200-g male infant born of a 24-year-old mother by vaginal delivery after 38 weeks gestation presented with a pedunculated mass protruding from his mouth (Fig. 1). The mother had not had ultrasonography (USG) examinations during her pregnancy. The mass was 6×4 cm in diameter and originated from the anterior hard palate near the midline. The mass was attached to the oral cavity (hard palate), with a peduncle 1 cm in diameter. It hampered feeding without causing respiratory distress. Because there was no associated anomaly and the mass was localized to the hard palate, we did not obtain fine needle aspiration, computerized tomography (CT), or magnetic resonance imaging (MRI). An excisional biopsy was planned. The mass was totally excised, and a profuse bleeding from the attachment site was observed and controlled. The mucosal defect was repaired by suturing two locoregional flaps with 4–0 Vicryl. Histopathologic examination revealed a mature teratoma composed of mature keratinizing squamous epithelium, skin adnexae, adipose tissue, neuroglial tissue, and bone formation (Fig. 2). The patient showed no signs of recurrence in the 1st year of follow-up. Alpha-fetoprotein (AFP) levels were normal.

Discussion

Teratomas occur in 1 out of 4,000 live births. Head and neck occurrence is generally localized to the neck and nasopharynx and comprises 1–10% of cases. Other extracranial presentations of teratomas are very rare,

Table 1 Oral teratomas (*EE* extraoral extension, *RD* respiratory distress, *FP* feeding problems)

Case (ref.)	Sex	Age at diagnosis	Size (cm)	EE	RD/FP	Treatment	Histology[a]	Associated pathologies	Location	Follow-up
1 (Strachan)	F	Premature exitus after 2 h	9.5×8	+	+?/?	NA	Cartilage, striped and unstriped muscle, lymphoid, muscular, glandular		Hard palate (mass was filling the oropharynx and nasopharynx and in close junction to sella turcica without connection to the sphenoid bone, and after running forward in left nasal cavity projected from left nostril	NA
2 (Miller)	F	Birth	7×3.5×3	+	−/+	Early resection	Bone, cartilage, ducts, glands (sebaceous, eccrine), hair follicles, cornified stratified squamous epithelium		Tongue (superior aspect of the right anterolateral tongue)	5 years
3 (Greir)	M	Birth (tongue base), 5 months (tongue)	6.5×4.5×2	–	−/+	Early resection	Mature neural, skin, muscle, respiratory, cartilage, bone	Multifocal/ recurrence? (tongue base-first diagnosed)	Tongue (in the substance of the tongue)	?
4 (Bras)	M	Birth	4.5×3x1	–	−/+	Early resection	Immature neural, cartilage, muscle, squamous		Tongue (left side)	?
5 (Dudgeon)	F	Birth (anterior cervical), 5 months tongue	2×1.3	–	−/−	Early resection	Gastric, intestinal, respiratory, smooth muscle, neural, pancreatic, salivary, Brunner's glands	Multifocal (tongue + anterior cervical/ submandibular)	Tongue (right lateral)	Uneventful in short term
6 (Ashley)	M	Birth	10×12	+	+ (when crying)/+	Early resection (after aspiration of the cystic content)	Immature neural, intestine, papillary, squamous, cartilage	Bil. inguinal hernias, hydrocele, umbilical hernia	Tongue (in the substance)	2 years
7 (Zakaria)	F	Birth	> 8×8[b]	+	−/+	Early resection	Nerve, cartilage, hair follicles, epidermoid structures, pseudoglandular formation		Hard palate (midline)	?
8 (Lalwani)	F	Birth	4×5	–	−/−	Laser excision	Nonkeratizing squamous, muscle, respiratory, gastrointestinal, well differentiated		Tongue (right anterior dorsal)	1.5 year
9 (Smith)	F	Prenatal	4	+	NA	Pregnancy terminated in 29th gestation week	Cartilage, squamous, bone, respiratory, mature and immature neural		Hard palate (intracranial spread to third and lateral ventricles)	NA
10 (Ekici)	M	Prenatal (USG) in 26th fetal week	9×11×13	+	+/?	(exitus 2 min after cesarean)	Benign teratoma		Hard palate	NA

Table 1 (cond.)

Case (ref.)	Sex	Age at diagnosis	Size (cm)	EE	RD/FP	Treatment	Histology[a]	Associated pathologies	Location	Follow-up
11 (April)	F	Birth	>8×8[b]	–	(+)?/+	Lateral pharyngotomy + oral approach, skull base extension of tm, defect repair with locoregional flaps	three germinal layers		Junction of the hard and soft palate (right), parapharyngeal and skull base extension	Uneventful in 1 year
12 (Uchida)	F	Birth	7×5	+	–/+	Early resection	Respiratory, gastrointestinal, mature neural, fat, connective tissue	Multifocal (left upper pharyngeal wall), cleft palate	Tongue (left side)	Uneventful in 2 years
13 (Wakhlu)	M	Birth	2×2[b]	–	–/–	Early resection	Mature elements from all germinal layers	Cystic hygroma	Hard palate (midline)	?
14 (Cay)	M	Birth	6×4	–	–/+	Early resection	Mature squamous epithelium, adipose tissue, neuroglial tissue, bone		Hard palate (midline)	Uneventful in 1 year

[a] All cases are reported as benign
[b] Sizes are not mentioned in the corresponding articles; these are estimated dimensions from case photographs

Fig. 1 A pedunculated mass protruding from the child's mouth

and only 13 cases of pure oral teratomas (seven from the tongue and six from the hard palate) have been reported in the literature (Table 1) [2, 3, 7, 9, 10, 12, 14, 16, 20, 22, 23, 24,25].

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 arise [5]. 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 [11,18].

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 teratomas are diagnosed during the perinatal period. USG is useful

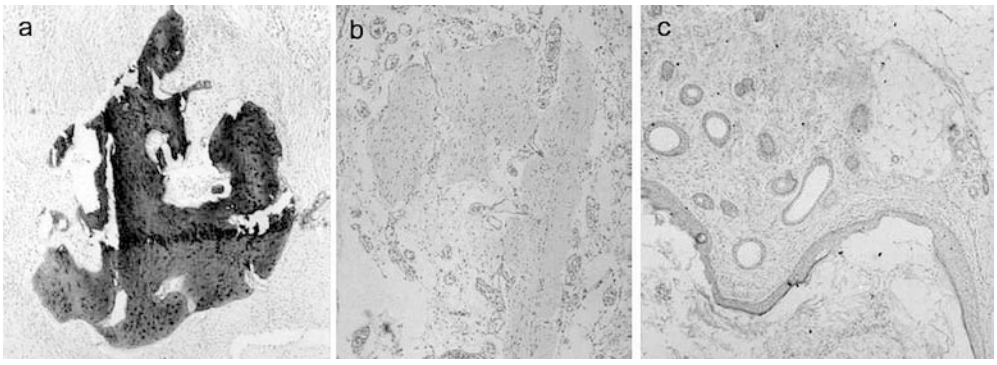

Fig. 2 a Irregular compact bone formation and fibroadipose tissue (HEX40). **b** Islands of neuroglial tissue and vascular proliferation (HEX40). **c** Mature squamous epithelium, skin adnexae, and adipose tissue (HEX40)

in prenatal diagnosis [17], 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. Eleven out of 14 oral teratomas that we reviewed had not been diagnosed until birth, even when the mothers had regular USG exams and their infants had considerably large teratomas. In the present case report, no prenatal USG was obtained, and the diagnosis was established postnatally.

Children with pure oral teratomas seem to have less dramatic respiratory behavior 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 [23]. In addition, children with oral teratomas have been reported to have associated anomalies including cleft palate [23], cystic hygroma [24], and other multifocal teratomas [9,23]. In our case, no associated anomalies were found.

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, oral or nasal endotracheal intubation or, if necessary, tracheotomy must be performed [17]. After securing the airway, a differential diagnosis of the mass should be done. USG establishes the presence of solid and cystic tissue components. Plain films and CT are useful in demonstrating virtually diagnostic calcifications [1,23]. In our case we did not need to obtain a CT scan because we did not suspect teratoma and the mass was occupying only the hard palate. 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 [13,19]. Teratomas of the floor of the mouth may imitate thyroglossal duct cysts [13]. The diagnosis is sometimes possible only after histopathologic examination. In our case the preoperative diagnosis was epulis, but histopathology revealed a mature teratoma.

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% [8]. 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 [24]. Recurrences are rarely seen in head and neck teratomas [23]. Our patient was free from any recurrences at the end of the 1 year follow-up period. In malignant teratomas, radiotherapy and chemotherapy are used after complete extirpation of the tumor [13,21].

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 [4, 6,15]. April et al. suggested that these measurements should be continued for patients with extensive lesions or those without a capsule [2]. Measurement of serum beta-HCG during the follow-up has also been advised . In our case, all of the AFP measurements were within the normal range.

Oral teratomas are extranodal germ cell tumors that occur very rarely during infancy and childhood. Children with oral teratomas have less severe symptoms when compared with those with oropharyngeal, nasopharyngeal, and cervical teratomas. They cause fewer problems during pregnancy and are less likely to be detected antenatally. If a child is diagnosed with an oral teratoma, although all reported pure oral teratomas have been benign, the risk of malignant change is evident, and long-term follow-up is mandatory, even if the tumor is totally excised with free margins.

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