Fetal Oral & Neck Lesions

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No conflict of interest
Fetal Oral & Neck Lesions - Overview

- Background
- Approach to the differential diagnosis

Incidence

- Congenital tumors are rare!
- Incidence if 1.7–13.5 per 100,000 live births.

Severino M, Neuroradiology 2010
Milani HJ, World J Radiol 2015
Pharyngo-epiglottic folds

Lips 21%
Toungue 13%
Cheek 5%
Floor of mouth 4%
Salivary glands 3%
Alveolar ridge 13%

95 patients
30-year period
Age at presentation: 1 day – 16 years
83 (87%) - Benign
12 (13%) – Malignant
- 5 Rhabdomyosarcomas
- 2 Fibrosarcoma
- 2 Carcinomas of the parotid
- 1 Osteosarcoma
- 2 Metastases
<table>
<thead>
<tr>
<th>Category</th>
<th>Types</th>
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<tbody>
<tr>
<td>Vascular</td>
<td>Oral vascular neoplasms:</td>
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<tr>
<td></td>
<td>Congenital hemangioma</td>
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<td></td>
<td>Infantile hemangioma</td>
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<td></td>
<td>Others, mostly outside oral cavity (eg, Kaposiform hemangioendothelioma)</td>
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<td>Oral vascular malformations:</td>
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<td>VM</td>
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<td>LM</td>
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<td>Arteriovenous malformation</td>
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<td>Arteriovenous malformation</td>
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<td>Mixed vascular malformations</td>
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<tr>
<td>Nonvascular</td>
<td>Epignathus</td>
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<td>Epulis</td>
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<td>Developmental oral thyroid anoma:</td>
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<td>Lingual thyroidal duct cyst</td>
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<td>Lingual ectopic thyroid gland</td>
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<td>Oral duplication cyst</td>
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<td>Oral hamartoma</td>
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<td>Oral lipoma</td>
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<td>Lingual dermoid or epidermoid cyst</td>
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<td>Many others</td>
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<td>Acquired neoplasms</td>
<td>Recurrent</td>
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<td>Oral teratoma</td>
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<td>Oral epithelial lesions</td>
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<td>Papilloma</td>
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<td>Fibromyxoidoma</td>
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<td>Cystic hygroma</td>
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<td>Chondromyxoidoma</td>
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<td>Odontoma</td>
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<td>Arteriovenous</td>
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<td>Kaposiform stromal nevus</td>
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<td>Malignant lesions</td>
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<td>Oral submucous carcinoma</td>
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<td>Oral lymphoma</td>
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<td>Oral mesenchymal sarcoma</td>
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<td>Oral myofibromatosus</td>
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<td>Conditions that mimic oral neoplasms</td>
<td>Asymmetric normal salivary gland</td>
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<td></td>
<td>Macroglossia</td>
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<td>Forehead encephalocele</td>
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<td>Cleft lip or palate</td>
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</tbody>
</table>

Sources: References 2-4. Next: LM = lymphatic malformation, VM = venous malformation.
Congenital lesions according to anatomic location

Site: Midline palate (a)
**DD:** Epignathus

Site: Soft tissues of the maxilla or mandible (b)
**DD:** Epulis or epidermal cyst or ranula

Site: Anterior two-thirds of the tongue (c)
**DD:** Oral foregut duplication cyst

Site: Posterior one-third of the tongue (d)
**DD:** Congenital thyroid anomaly or lingual hamartoma

Site: Span multiple neck spaces (e)
**DD:** Vascular anomalies (neuroblastoma, sarcoma – rare)
Oral teratoma

- Incidence of all teratomas is 1:4000.
- Head and neck are the second and third most common sites involved, accounting for 10-15% of all fetal teratomas.
- May contain cystic and solid components.
- Grows rapidly and may be up to 30 cm in diameter.
- Calcifications are observed in 50% of cases.
- Color Doppler US may show a prominent feeding vessel that arises from the palate and extends into the mass.
Oral teratoma

• Associated with cleft palate and other oral malformations in 6-20% of patients.

• Can be isolated or in association with:
  • Trisomy 13
  • Ring X-chromosome mosaicism
  • Aicardi syndrome
  • Pierre-Robin syndrome

The advantage of prenatal MRI is its ability to define the mass origin in the hard or soft palate.

MR image confirms the origin of the mass (arrow) in the midline palate
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**Congenital epulis**

- AKA
  - Congenital granular cell tumor
  - Congenital granular cell lesion
  - Gingival granular cell tumor of the newborn
  - Neumann’s tumor
- Rare! 6:1,000,000
- F > M (8-9:1)
- Predilection for the maxillary alveolar process.
- 90% presents as a single pedunculated gingival mass.

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Kumar P. The Laryngoscope. 2002
Kim SK. Prenat Diagn. 2006
Yuwanat M. J Neonatal Surg. 2015
Congenital epulis

- Usually isolated
- In up to 10% can be associated with:
  - Polydactyly
  - Goiter
  - Maxillary hypoplasia
  - Neurofibromatosis
  - Triple X syndrome
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Salivary glands

Three paired major salivary glands as well as hundreds of minor salivary glands drain directly into the mouth, providing moisture to the oral cavity.
Ranula
Ranula

- Rare
- Cyst arising from the sublingual or minor salivary glands.

Simple ranula

Confined to the sublingual space.

A well circumscribed fluid attenuation lesion within the left sublingual space (*).
Plunging/complex/diving ranula

Extends into the submandibular space.

A well-circumscribed cystic lesion predominantly occupying the left submandibular space causing a mass effect on the left submandibular gland.

- Intra-oral cystic lesion measuring 20 × 21 mm
- No solid component
- No calcifications.
• Planned Cesarean section at 38 + 3 weeks’ gestation
• Ex utero intrapartum treatment (EXIT) was performed and 40 mL of clear fluid was aspirated from the lesion before the cord was clamped.
• Histology revealed a mucous cyst lined by salivary duct epithelium.

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Foregut

Foregut duplication cyst

- Benign developmental anomalies along the foregut-derived alimentary tract.
Foregut duplication cyst

- Benign developmental anomalies along the foregut-derived alimentary tract.

- Rare – 1:4500
- Most commonly arise in the thorax and abdomen.
- 0.3% present in the head and neck.
- M > F (3:2)

Lee AD. World J Clin Cases. 2020
• This cyst may interfere with anterior fusion of the vertebral mesoderm
• Vertebral anomalies in approximately 50% of cases, mainly:
  • Scoliosis
  • Hemivertebra
  • Spina bifida

Fetus at 28wks shows a markedly bright, circumscribed lesion (white arrowhead) within the anterior aspect of the fetal tongue.

Gantwerker EA, JAMA Otolaryngology. 2014
• Cesarean delivery at term
• Apgar scores were 8/9.
• Intubation few minutes after birth

A. Ventral incision has been made along the anterior tongue. The surrounding musculature has been gently peeled away anteriorly.
B. The anterior aspect has been opened and contents drained.
C. The cavity closed down with a Penrose drain left in place.
Pseudostratified ciliated columnar epithelium

Gastric epithelium

Gantwerker EA. JAMA Otolaryngology, 2014

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Thyroid gland embryology

- The thyroid primordium originates at the foramen caecum at 2-3 weeks’ gestation.

Nakayama S. Oral Maxillofac Surg 2009
Zander DA. Radiographics 2014
Congenital thyroid anomalies

Ectopic thyroid gland

Thyroglossal duct cyst

Failure of migration, in addition to functional development.

Thyroid ectopia

Failure of migration, in addition to functional development.

Tucker D. J Clin Endocrinol Metab, 2016
Highly vascular ectopic thyroid gland (arrowed) lying at the level of the base of the tongue (arrowed).

Thyroid ectopia

Thyroglossal duct cyst

<table>
<thead>
<tr>
<th>Location</th>
<th>Percentage</th>
</tr>
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<tbody>
<tr>
<td>Lingual</td>
<td>2%</td>
</tr>
<tr>
<td>Suprahyoid</td>
<td>24%</td>
</tr>
<tr>
<td>Thyrohyoid</td>
<td>61%</td>
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<tr>
<td>Suprasternal</td>
<td>13%</td>
</tr>
<tr>
<td>Mediastinal</td>
<td>rare</td>
</tr>
</tbody>
</table>

Nakayama S. Oral Maxillofac Surg 2009
Zander DA. Radiographics 2014
Thyroglossal duct cyst

- Incidence is 7% according to a postmortem study of 200 adults.
- Many of cystic remnants never detected clinically.
- Usually within 2cm of the midline.
- Cysts do not typically invade to laryngeal structures.

*Ellis PD. Laryngoscope, 1977
Chou J. Surg Radiol Anat, 2013
Tárrega ER. Case Reports in Obstetrics and Gynecology 2016*

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**Case Report**

Prenatal Ultrasound Diagnosis of a Cyst of the Oral Cavity: An Unusual Case of Thyroglossal Duct Cyst Located on the Tongue Base

Male fetus @ 28 weeks

- Cyst lesion located under the tongue, homogeneous, without solid components, with well-defined limits.
- Histology confirmed thyroglossal duct cyst.

*Tárrega ER. Case Reports in Obstetrics and Gynecology 2016*
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Lingual hamartoma

- Hamartoma - proliferation of normal tissue endogenous to the anatomic site of occurrence.
- The tissues appear disorganized and ill-defined, merging with the normal surrounding tissues.
- Tongue hamartoma may be composed of vascular, nerve, skeletal or smooth muscle, fat, or salivary gland tissue.
- It usually manifests as a solitary polypoid lesion.

Oral-Facial-Digital (OFD) syndrome
Rare genetic disorders characterized by the association of abnormalities of
- Oral cavity: lingual hamartoma, abnormal frenula and lobulated tongue
- Face: hypertelorism and low-set ears
- Extremities: brachydactyly and polydactyly

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Teratoma  Vascular lesion
Fetal Teratoma

- Incidence 0.07-2.8:1,000
- Accounts for the majority of solid fetal tumors.
- Predominantly benign tumors.
- Located in the midline of the body.

- Sonographically, they appear as:
  - Mixed echogenicity with cystic and solid parts
  - Calcifications
  - Blood perfusion of varying degree
- Typically, not associated with abnormal genetic findings.
Lymphatic malformation

- 50-75% located in the cervicofacial region.
- Appearance can vary!!
- Typically no or only minimal solid portions.
- Typically no increased vascularity.
- Typically no calcification.

73 cases of prenatally detected lymphatic malformations (1997-2015)

Genetic associations in 7/73 (9.6%):
- 4 - CLOVES syndrome (congenital lipomatous overgrowth, vascular malformations, epidermal nevi, and scoliosis/skeletal/spinal anomalies)
- 1 - Klippel-Tranaunay syndrome
- 1 - Trisomy 21
- 1 - Chromosomal microdeletion
- **Type I** - Trans-spatial cystic lesions with multiple internal septations.
- **Type II** - Predominantly cystic with no more than three septations.

Type III - purely cystic
Type IV - a combination of cystic and solid components (solid tissue should comprise at least 30% of lesion's volume).

Large complex appearance with solid portions and calcified phleboliths

Vascular malformation with solid component ? Teratoma

Invasive facial cervical veno-lymphatic malformation with cystic and solid components

Teratoma with sharp margin between the lesion and surrounding tissue

Infiltration

Distortion
Even a large venolymphatic malformations usually produces a rather mild mass effect on surrounding structures.
Thank You!

Watch Your Mouth!

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