

Congenital epulis and differential diagnoses of a neonatal oral mass

Oral mass in a newborn is rare but often quite daunting, leading to anxiety in parents. Very few hospitals have dental or maxillofacial facilities on site and this may lead to delayed diagnosis and intervention. A thorough clinical examination and knowledge of the oral mass is essential for precise diagnosis, management and parental reassurance. In this review article, we present a newborn baby with congenital epulis – an oral mass arising from the alveolar ridge. We also summarise the differential diagnoses based on location and consistency of the oral masses in this age group.

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Key points

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1. Congenital epulis is an uncommon benign swelling present on the gingivae of newborn infants.
2. Epulis can be confused with many other diseases that present as oral lesions in the newborn period.
3. Distinguishing and diagnosing oral masses in the newborn period is discussed.

The case

A baby girl was born at term with a birth weight of 3.3kg. She was born to a primigravida with an uneventful pregnancy, normal serology and normal antenatal anomaly scans. The last scan was performed at 20 weeks of pregnancy. The mother had a history of Hodgkin lymphoma and had received radiotherapy and chemotherapy almost a decade previously. The baby was born by assisted vaginal delivery with Apgar scores of 9 and 9 at one and five minutes, respectively. The baby was breastfed and she was latching well and sucking without any difficulty.

A swelling over the lower gum was noted by a midwife on the initial check and the neonatal team was called to review. The swelling was round, solitary, 2x2cm in size and arising from the lower alveolar ridge over future canines (FIGURE 1). The swelling was firm in consistency with a smooth surface and non-tender on palpation. The parents were understandably anxious, especially due to

the family history of lymphoma. She was reviewed by the dental team and the diagnosis of congenital epulis was confirmed.

The plan was for conservative management. The child would be actively monitored for change with regular review of the size/shape of the epulis, evidence of bleeding or pain, difficulty in feeding or breathing, weight gain and dentition. The management plan was explained to the parents and they were reassured about the benign nature of the mass and likelihood of self-resolution.

Discussion

Congenital epulis of the newborn

Congenital epulides are benign mesenchymal tumours arising from the anterior alveolar ridge, composed of large cells with a coarse granular cytoplasm. They were first described by Neumann in 1871 and later explained by Zuker and Buenecha¹ in 1993. They are usually present at birth or appear soon after. Growth generally starts antenatally around



FIGURE 1 Congenital epulis arising from the lower alveolar ridge.

the 22nd week of gestation, hence they are unlikely to be diagnosed on anomaly scans performed between 12 to 20 weeks.² Large lesions may be picked on late antenatal scans and MRI is the preferred modality to assess the extent of the lesion and help plan delivery.

They appear as smooth, non-ulcerated polypoid masses with a broad-based attachment to the alveolar ridge, with preference for the maxilla over the mandible and overlying the future canine and lateral incisor teeth. Although usually singular, about 10% of reported cases are multiple or a cluster of small tumours with potential to cause mechanical obstruction, respiratory and feeding difficulties.³

Congenital epulides are rare with an incidence of 0.0006%.^{4,6} Females are affected more often than males (9:1).⁷ Various aetiologies are proposed including odontogenic, neurogenic, myoblastic, endocrinologic, fibroblastic or histiocytic origin.^{8,9} Female predominance may favour the endocrine theory of aetiology.

Epulides are usually benign, but the obstructive masses may cause difficulty with respiration and feeding. They stop growing at birth and often regress over time. Local conservative resection, under local anaesthesia for small epulides, is curative and only required if causing difficulty in feeding or breathing. Surgical excision is required if affecting the growth of the bone or the eruption of teeth. Even after incomplete excision, they do not recur.¹⁰⁻¹²

Oral masses in the newborn period

A wide range of diseases present as oral lesions in the newborn period; fortunately, the majority are benign. A thorough clinical examination and knowledge of the oral mass is essential for precise diagnosis and management. Oral lesions in neonates include:

- cysts
- infections
- traumatic lesions
- tumours
- autoimmune diseases.

Malignant tumours of soft tissue, salivary glands and bones must be considered.

The list of conditions to consider in the differential diagnosis for a congenital oral mass is long but identification based on anatomical location in the oral cavity (TABLE 1) and consistency (FIGURE 2) helps to narrow the diagnosis.

Location	Differential diagnoses
Midline palate	Epignathus/teratomas, malignant tumours/mass
Soft tissues of the maxilla or mandible	Epulis
Multiple neck spaces	Mostly vascular anomalies, malignant mass
Anterior two-thirds of the tongue/ base of mouth	Oral foregut cyst, ranula
Posterior one-third of the tongue	Choristomas (hamartomas, lingual thyroid, etc)

TABLE 1 Differential diagnoses for an oral mass based on anatomical location in the oral cavity.¹³

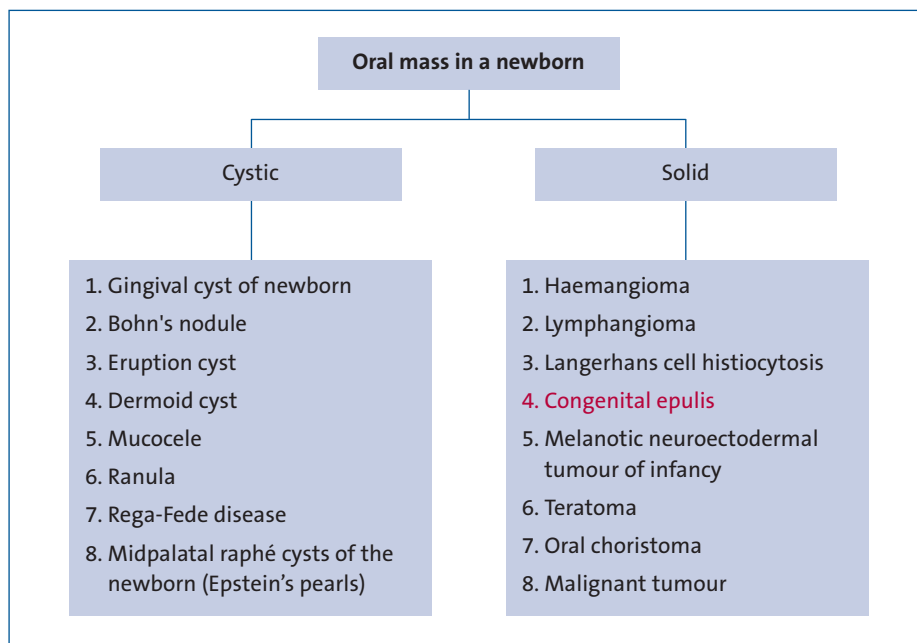


FIGURE 2 Differential diagnoses for an oral mass based on consistency.

Cystic oral masses (TABLE 2)

Gingival cyst of the newborn

Also known as dental lamina cyst. A small, benign oral mucosal lesion, usually multiple and nodular. White to cream-coloured lesions.¹⁴⁻¹⁷

Bohn's nodule

A keratin-filled nodular cyst that presents as small, whitish lesions that do not increase in size. When present on the anterior mandibular ridge they may be misdiagnosed as natal teeth.¹⁵⁻¹⁸

Eruption cyst

Presents as a bluish, dome-shaped, translucent, compressible swelling within the mucosa, overlying an erupting tooth.^{15,19}

Dermoid and epidermoid cysts

Rare benign, asymptomatic and slow-growing cysts. Diagnosed if/when enlarged with a potential risk of respiratory distress and feeding difficulty.^{16,17,19}

Mucocele

Presents as a bluish (depending on the proximity to the surface), translucent and

fluctuant swelling that may cause mechanical obstruction during feeding.^{17,20}

Ranula

A painless swelling in the floor of the mouth commonly caused by extravasation of mucin.²⁰⁻²²

Rega-Fede disease

Appears as an ulcerated/cystic or fibrous mucosal mass. The neonatal type is benign and commonly associated with natal/neonatal teeth. Unifocal/multifocal and occasionally painful.^{20,23,24}

Midpalatal raphé cysts of the newborn (Epstein's pearls)

Small (1-3mm) yellow-white papules along the midpalatal raphé, especially at the junction of the hard and soft palate. The cysts are filled with keratin.^{20,25,26}

Solid oral masses (TABLE 3)

Haemangioma

The most common vascular benign mass. Manifests as a rapidly growing macule followed by regression into spotted pigments.^{27,28} Can be present at birth but

tends to develop a few weeks after birth. There are three phases of development:

1. rapid proliferating phase (0-1 year)
2. involuting phase (1-5 years)
3. involuted phase (5-10 years).

Lymphangioma

Benign congenital tumours of the lymphatic system. Oral lesions present with macroglossia, dysphagia, ulcerations, deformity of jaws, and feeding problems. A lymphangioma can be superficial (slow, progressive lesion with superficial blue-black or red haemorrhagic elevated

nodules), deep (soft, diffuse growths with normal colour) or cystic (soft, painless fluid-filled lesions, characteristically located in the neck).²⁸

Langerhans cell histiocytosis

Congenital self-healing reticulohistiocytosis presenting at birth and showing complete involution within 2-3 months. Characteristic features include; erythematous vesicular-pustules with/without crusting and eczematous scaling of the skin; oral petechiae, swelling of the gingiva; X-ray lytic lesions. Depending on severity

and extent, systemic findings may include hepatomegaly, splenomegaly, pancytopenia, and central nervous system involvement.^{29,30}

Melanotic neuroectodermal tumour of infancy

Characterised by benign, painless, non-ulcerative, expansile, rapidly growing, pigmented lesions.³¹⁻³²

Teratoma

Presents as an oral mass with mass effect – eg poor feeding, breathing difficulty. May have an intracranial component.^{33,34}

Cystic oral mass	Incidence	Sex	Origin	Location	Diagnosis	Treatment
Gingival cyst of the newborn ¹⁴⁻¹⁷	Frequent in newborn = 13.8%	Affects both genders equally	Dental lamina	Crest maxillary and mandibular dental ridges	Clinical	None, self-resolution, reassure parents
Bohn's nodule ¹⁵⁻¹⁸	High prevalence = 47.4% (79% has been reported in Taiwanese neonates)	Affects both genders equally	Derived from minor salivary gland structures	Nodules at the junction of the hard and soft palate	Clinical Histological	None, self-resolution, reassure parents
Eruption cyst ^{15,19}	Rare with few case reports	Affects both genders equally	Degenerative cystic changes in the reduced enamel epithelium Remnants of the dental lamina secondary to an impediment of eruption by overlying dense fibrotic mucosa	Alveolar ridge	Clinical (consider fine needle aspiration or biopsy)	Marsupialisation or surgical extraction
Dermoid and epidermoid cyst ^{16,17,19}	7% in the head and neck region	No specific predilection	Dermoid cysts are epithelial-lined with some skin appendages eg hair, hair follicle, sebaceous and sweat glands Epidermoid cysts are lined by epithelium only	The floor of the mouth Submental region	Ultrasonography MRI/CT	Surgical enucleation (recurrence is rare)
Mucocele ^{17,20}	Approximately 2.7%	No specific predilection	Commonly arise due to alterations in the minor salivary glands	Lower lip is the most common site	Fine needle aspiration/ biopsy	Surgical excision
Ranula ²⁰⁻²²	Rare – incidence is approximately 0.74%	No specific predilection	Salivary gland damage	Floor of the mouth	Mostly clinical Confirmed by MRI Histology of a surgical specimen	Observation for asymptomatic cases If treatment is required: cryosurgery, marsupialisation or surgical excision
Riga-Fede disease ^{20,23,24}	Rare benign mass	No specific predilection	Oral mucosa secondary to repeated trauma from teeth	Ventral surface of tongue, lip, palate, vestibular mucosa, floor of the mouth	Clinical examination	Conservative management in neonates or dental extraction
Midpalatal raphé cysts of the newborn (Epstein's pearls) ^{20,25,26}	Very common Prevalence = 65-85% of neonates		Epithelial remnants along the line of fusion of nasal septum and hard palate		Clinical	No treatment indicated; self-resolution

TABLE 2 Cystic oral mass in the newborn period – differential diagnoses.

Solid oral mass	Incidence	Sex	Origin	Location	Diagnosis	Treatment
Haemangioma ^{27,28}	Most common vascular benign mass Prevalence = 2-3% in term (higher in preterm infants)	Female predilection (3:1-5:1)	Abnormal angiogenesis	Commonly head and neck regions including lips, buccal mucosa and tongue	History Colour doppler ultrasonography	Medical (propranolol, corticosteroids, alpha-interferon) Lasers Surgery
Lymphangioma ²⁸	Prevalence = 1-3 per 10,000 live births 50% of cases noted at birth 90% develop before age 2	Affects both genders equally	Incomplete development; ectopic deposition of lymph tissues; congenital obstruction or sequestration of the primitive lymphatic channels	Dorsum of the tongue, lips, buccal mucosa, soft palate, and floor of the mouth	Clinical Ultrasound with doppler CT/MRI for extent of involvement Histopathology and immunohistochemistry	Steroids Cryotherapy Embolisation Radiofrequency tissue ablation Radiation Surgery
Langerhans cell histiocytosis ²⁹⁻³⁰	Frequency = 1 in 200,000	Male predilection	A wide group of disorders but always histiocyte as the common cell of origin	Posterior mandible Skull, femur, pelvis and vertebrae	Histopathology and immunohistochemistry CT/MRI to evaluate extent of involvement	Surgery Chemotherapy Radiotherapy
Congenital epulis	Incidence = 0.0006%	Female > male 9:1	Various aetiologies proposed: odontogenic, neurogenic, myoblastic, endocrinologic, fibroblastic or histiocytic origin	Gingival mucosa of the maxilla or mandible	Prenatal diagnosis Histopathology and immunohistochemistry	Often regress over time Local conservative resection Surgical excision
Melanotic neuroectodermal tumour of infancy ³¹⁻³²	Rare Occur in the first 6 months of life	Male predilection	Neural crest cells	Tongue, palate, buccal mucosa, and floor of the mouth	CT/MRI Histopathology	Surgical excision Chemotherapy and radiotherapy (alone or combined)
Teratoma (epignathus) ^{33,34}	Exceptionally rare Incidence = approximately 1 in 35,000 to 200,000 live births	Female > male 3:1	Pluripotent cells of one or more of the three germ layers	Base of the skull, usually the hard palate or mandible	Ultrasonography, CT/MRI Bloods: elevated maternal serum alpha-fetoprotein	Surgery Prognosis is poor and mortality rate is high in fetal life/neonates
Oral choristoma ^{20,32,33,35,36}	Extremely rare	Male > females 1.6:1	Pluripotent stem cells of the first and third pharyngeal arches Histologically can be respiratory and gastrointestinal mucosa, glial, cartilaginous, thyroid or glandular in origin	Head and neck region, tongue (lingual choristoma), floor of mouth, pharynx, and hypopharynx	Clinical Biopsy	Complete surgical excision
Haemangioendothelioma of the parotid gland ³⁷	Rare Present at birth or may present at 3-4 months	Female > male: 3:1	Parotid glands	Rapidly growing multiple parotid masses	Ultrasound MRI	May involute with time or Surgical excision (with risk of facial nerve palsy)
Malignant oral lesions in neonates ^{17,18,20}	Extremely rare	No reports	Hard palate, tongue, face and neck muscles	Head and neck	Mostly diagnosed on antenatal scan Antenatal fetal MRI Postnatal CT, MRI	Require a multidisciplinary approach at delivery Depends on type of malignancy

TABLE 3 Solid oral mass in the newborn period – differential diagnoses.

Oral choristoma

Abnormal location of histologically normal tissue. Mostly asymptomatic but with space-occupying effects. Can be cystic or solid depending on the tissue of origin.^{20,32,33,35,36}

Malignant tumour

A solid space-occupying lesion mostly diagnosed on antenatal scans. The mass may interfere with feeding and respiration, depending on location. For example, malignant melanoma of palate and spindle cell sarcoma of tongue.^{17,18,20}

Patient consent

The authors received written consent to publish this report from the patient's parents.

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