Congenital Epulis: Unusual Etiology of Airway Obstruction and Feeding Failure in a Newborn

AUTHORS:
Cody L. Mullens, BS[a]; Luke J. Grome, MD[b]; Shilpa S. Vishwanath, MD[c]; Harold J. Williams, MD[d]; Aaron C. Mason, MD, FACS, FAAP[e]

CORRESPONDENCE AUTHOR:
Cody Lendon Mullens
West Virginia University
1 Medical Center Drive
Morgantown, WV 26505
304-723-8039
cmullen3@mix.wvu.edu

AUTHOR AFFILIATIONS:
a. West Virginia University School of Medicine – Medical Student
b. Baylor College of Medicine, Department of Surgery, Division of Plastic Surgery – Resident
c. West Virginia University School of Medicine, Department of Otolaryngology – Resident
d. West Virginia University School of Medicine, Department of Pathology – Professor
e. West Virginia University School of Medicine, Department of Surgery, Division of Plastic, Reconstructive, and Hand Surgery – Assistant Professor

Background	A 2.8 kilogram (kg) female neonate born at full-term presented with an inability to breast feed due to an obstructive mass in the oral cavity.
Summary	Congenital epulis (CE) is a rare lesion of the newborn gingiva or alveolar mucosa. These lesions cause mechanical airway or feeding obstruction. CE can impact both prenatal and postnatal development. Tumor sequelae can include polyhydramnios, respiratory obstruction, and feeding failure. Surgical resection is imperative in patients presenting with signs of airway obstruction or feeding failure. CE is very rare and remains underreported in the medical literature. We present a case of symptomatic CE. Additionally, we discuss the origin of these rare tumors, their diagnosis, and their appropriate treatment modality.
Conclusion	CE is a rare lesion that is underreported in the medical literature and can cause both prenatal and postnatal sequelae. This report describes an illustrative case, which surgeons will be able to reference in order to promptly identify and surgically intervene in order to minimize morbidity in this rare patient population.
Keywords	Congenital epulis, congenital granular cell tumor, Neumann's tumor, congenital granular cell myoblastoma

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Case Description

A congenital epulis (CE) is a rare lesion of the newborn gingiva or alveolar mucosa found most commonly in females.[1,2] Other names for this rare tumor include congenital granular cell tumor, congenital myoblastoma, and Neumann's tumor. These lesions may be large in size and act as a source of mechanical oral and/or airway obstruction. They typically arise on the alveolar median ridge of the maxilla followed by the alveolar ridge of the mandible.[3] Sequelae of this tumor may include polyhydramnios during the prenatal period as well as respiratory obstruction and feeding failure postnatally.[1,4] While some reports of CE describe spontaneous regression of the lesion,[5,6] surgical resection is the safest option and is imperative in patients presenting with signs of airway obstruction or feeding failure.[7] CE is very rare and remains largely underreported in the medical literature. We present a case of symptomatic CE with early diagnosis and expedient surgical intervention. This management allowed the patient to resume oral feeds within a week after birth, greatly improving prognosis.

A full-term, 2.8 kg female presented with an oral cavity mass obstructing her ability to breast-feed. No respiratory distress was reported or observed. On examination, the mass measured 2x2 centimeters (cm) and visibly obstructed a large portion of the oral cavity. The mass was flesh-colored, pendulous, and firmly adherent to the right maxillary alveolar margin (Figure 1).

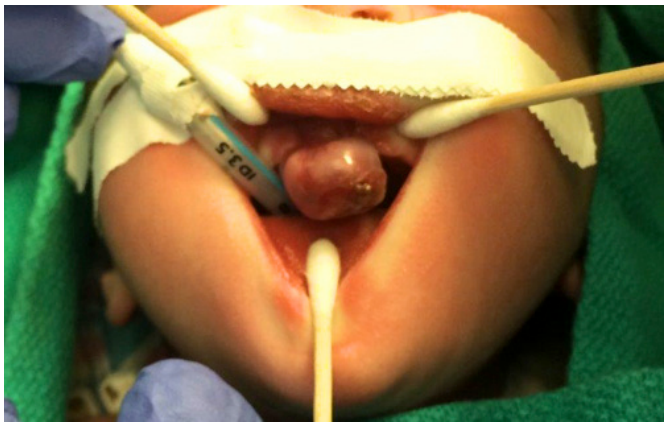

Figure 1. This image depicts the flesh colored, pendulous mass that was adherent to the right maxillary margin prior to excision.

Magnetic resonance imaging (MRI) of the lesion revealed a 2 cm exophytic structure protruding from the right maxilla, adjacent to the anterior inferior margin of the maxillary alveolar gingiva. Dental elements were not appreciated within the lesion, but components of the alveolus including fibrofatty, cartilaginous, and mucosal elements were identified (Figure 2). Due to the obstructive nature of the lesion leading to feeding failure, early surgical intervention with excision of the tumor was recommended. Intraoperative examination revealed a pendulous lesion with a 1 cm base composed of soft tissue with overlying intact mucosal surface. The lesion was excised from the margin at its point of attachment. Mucosal flaps adjacent to the base of the lesion were readily available in order to permit tension-free closure. The specimen was sent for pathologic examination (Figure 3), and the patient was subsequently able to successfully breastfeed following removal of the specimen. Follow-up without recurrence has continued for 28 months and is ongoing.

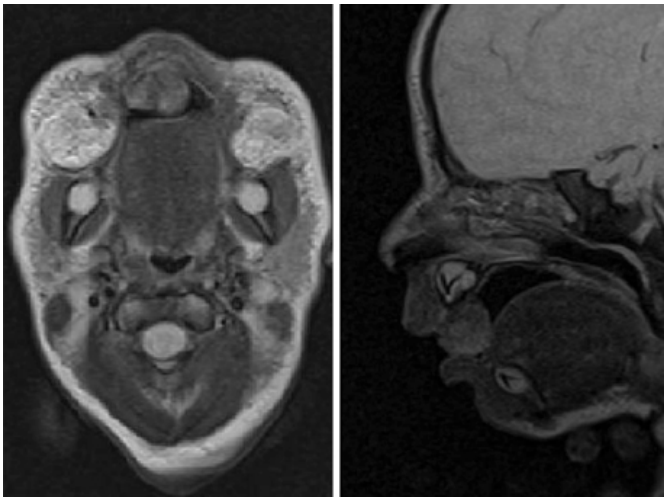

Figure 2. Preoperative MRI, which revealed an exophytic mass (see arrows) emanating from the right maxilla along the anterior inferior margin.

Discussion

The pathogenesis of congenital epulides is not known. Citing the preferential occurrence in females, some authors suggest the influence of hormones on development.[8] Potential cells of origin include fibroblasts, pericytes, epithelial and undifferentiated mesenchymal cells, myofibroblasts, and neuron-related cells.[9]

The first indication that a CE is present may be on prenatal ultrasound. In utero congenial epulis may lead to polyhydramnios. In the absence of other causes of polyhydramnios, careful evaluation of the oronasal cavity by ultrasound may be useful in the identification of CE or similar tumors of the oral cavity.[10,11] When recognized before birth, three-dimensional ultrasound may be useful to evaluate fetal swallowing and airway patency. In utero identification allows for adequate preparation at parturition for the possibility of airway obstruction.[6] The differential diagnosis of an anterior oral mass includes congenital epulis, teratoma, odontogenic cyst, dermatofibrosarcoma protuberans, and granular cell tumor.
In order to determine the definitive diagnosis, histologic factors, patient factors, gross morphologic factors, and imaging can assist in discriminating between the different potential differential diagnoses.

Histologically similar to adult granular cell tumors, CEs can be differentiated by negative S-100 staining.[9] Absence of cytoplasmic hyaline granules, solid growth pattern, pericytic proliferation, and attenuated overlying epithelium are also key CE discerning characteristics. In our case, microscopy revealed a subepithelial proliferation of cells with abundant eosinophilic cytoplasm in the absence of hyperplasia of overlying squamous epithelium is evident (Figure 3). Prominent vascular structures are typically appreciated as well.

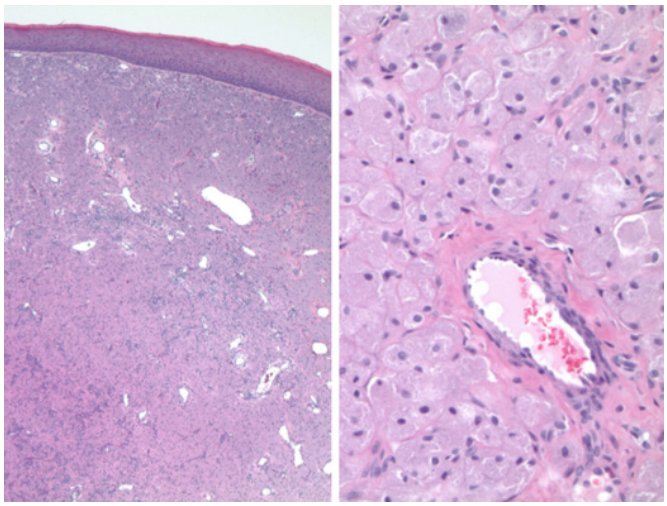

Figure 3. of hyperplasia of overlying squamous epithelium and the prominence of vascular structures. B) H&E stain, 20x. The tumor cells have abundant granular cytoplasm (low nuclear:cytoplasmic ratio) and small, uniform nuclei.

CE can also be distinguished from granular cell tumors by patient characteristics such as age. On gross inspection, a flesh colored, pendulous, and firmly adherent mass to the maxillary alveolar margin is the typical presentation.

MRI can further assist in confirming a diagnosis of CE. On MRI, a heterogeneous mass on T2 images and an isodense mass on T1 images are seen. Cartilaginous, mucosal, and fibrofatty elements are typically appreciated, while dental elements within the lesion are less common.
Surgical intervention is typically required for the treatment of congenital epulis. Expedited surgery may be required in the case of oral airway obstruction, whereas surgical excision for patients with feeding failure secondary to CE can be performed semi-electively. The size of the CE influences the degree of obstruction and its impact upon successful feeding. Larger lesions are more likely to obstruct the upper airway, leading to dyspnea and possibly hypoxemia. Suffocation from a CE has not been previously reported but is possible if the CE is large enough to obstruct the airway entirely. Although reports of spontaneous regression exist, the potential morbidity and mortality risks warrant surgical excision, especially for large lesions.[5,6] Some authors suggest conservative management of small CEs that do not compromising feeding or the neonatal airway as growth halts after birth and spontaneous involution has been reported.[12] Because of the small number of reported cases, recurrence rate after surgical excision is unknown.

Close coordination between a pediatric plastic surgeon and a pediatric anesthesiologist is useful in planning perioperative care. Pediatric airways may be challenging, and the presence of a large anterior mass further confounds securing a safe airway in some cases. Airway complications including oral bleeding, aspiration, and pneumothorax may occur, making successful intubation more of a challenge.[13,14]

Conclusion

In summary, CE is a rare and benign oral cavity lesion. Early recognition of large lesions is crucial, as airway obstruction and feeding failure may occur. We present a case wherein the lesion was quickly identified and effectively surgically managed in order to minimize patient morbidity. Physical examination, radiographic evaluation, and pathologic review are all key in evaluating and diagnosing CE. A multidisciplinary approach to these lesions permits a favorable prognosis and minimizes morbidity for the newborn patient.

Lessons Learned

A congenital epulis is a rare obstructive lesion involving the oral cavity that can have adverse effects prenatally and postnatally. Here we discuss approaches for CE diagnosis and management in order to minimize associated postnatal morbidity such as feeding failure and respiratory obstruction.

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