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Obstructive Congenital Epulis: Prenatal Diagnosis and Perinatal Management

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Objective: **To describe a multidisciplinary approach for delivery room management of congenital epulis.** ***Study Design:*** **Case reports.** ***Methods:*** **Reporting of two cases of congenital epulis and review of the literature.** ***Results:*** **Antepartum ultrasonography demonstrated massive intraoral masses in two fetuses whereby concerns regarding the patency of the airway at birth necessitated development of a multidisciplinary team of maternal–fetal medicine, neonatal–perinatal medicine, anesthesiology, and otolaryngology. Surgical excision was performed before delivery in one infant and after complete delivery in the other without a need for endotracheal intubation and general anesthesia. Feeding was started early, and both infants were discharged after brief hospital stays. Pathological findings were consistent with congenital epulis. Differential diagnosis and options for surgical intervention are discussed, including ex utero intrapartum treatment.** ***Conclusions:*** **A multidisciplinary approach to antenatally identified congenital intraoral masses facilitates care at birth. Surgical treatment in this milieu may be simple and complete at the time of delivery.** ***Key Words:*** **Congenital epulis, ex utero intrapartum treatment, airway obstruction.**

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INTRODUCTION

Congenital epulis, also known as congenital gingival granular cell tumor, is a rare, benign intraoral tumor found only in the newborn. It arises predominantly from the maxillary or mandibular alveolar ridges[1] and usually regresses spontaneously without recurrence. However, large or multiple lesions may interfere with respiration or deglutition. In such a case, surgical excision is the treatment of choice. We report two cases of congenital epulis identified as intraoral masses by antepartum ultrasonography that were successfully resected at the time of delivery. Both of these cases were remarkable, one for the necessity of immediate resection because of large size and possible airway compromise and the other because of early identification at 29.5 weeks of gestation. The clinical, morphological, and histological features and differential diagnosis of congenital epulis and various management options are presented.

CASE REPORTS

Case 1

A 35-year-old white woman, gravida 3, para 2, was referred to the Division of Maternal–Fetal Medicine at Long Island Jewish Medical Center, New York, when an antepartum ultrasound at 35 weeks of gestation revealed an intraoral mass. Routine antepartum laboratory test results were unremarkable. Maternal history was significant for a previous child with trisomy 21. Repeat ultrasonography at 36 weeks revealed two homogeneous, solid masses with narrow stalks protruding from the mouth (Fig. 1). The patient was followed with weekly sonograms, and the masses seemed to increase in size. Fetal swallowing appeared to be normal, and there was normal amniotic fluid volume. Color flow Doppler indicated a patent nasal airway. At 37 weeks, amniocentesis revealed a normal, 46 XX karyotype and an immature lung profile. A multidisciplinary meeting among physicians in maternal–fetal medicine, neonatal–perinatal medicine, anesthesia, and otolaryngology was held. A management plan was developed considering risks and benefits of various approaches. A decision was made to resect the lesions on delivery of the head if there was airway compromise secondary to the mass. The neonatologist and otolaryngologist discussed the plan with the family. At 39 weeks of gestation, when a mature lung profile was expected, an elective cesarean section was performed with the patient under spinal anesthesia and with the multidisciplinary team in attendance. The operating room was prepared for care of both the mother and the newborn. Equipment at hand for the infant included a radiant warming table, suction device, oxygen source, and usual equipment for resuscitation. In addition, a tracheostomy set was available. Personnel in the operating room included two teams of physicians and nurses, each team dedicated to either the mother or the child. The maternal team included obstetricians, obstetrical anesthesiologist, and surgical nurses. The other team consisted of otolaryngologists (attending and his resident), neonatologists (attending and the fellow), a pediatric anesthesiologist, and

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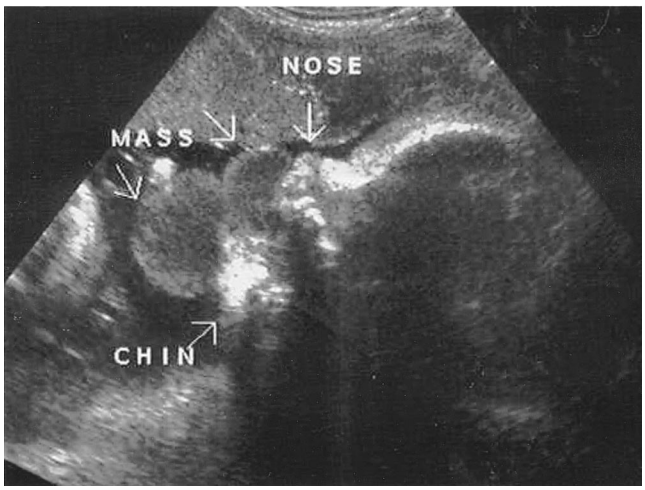

Fig. 1. Intrauterine ultrasonogram demonstrating two intraoral masses protruding from mouth of the fetus in case 1.

a neonatal nurse. On delivery of the head, two large masses attached by 2-cm stalks were observed to be arising from the mandibular and maxillary alveolar ridges and protruding from the mouth (Fig. 2). Together, the masses appeared to occlude the oral and nasal cavities. In view of impending airway obstruction, the stalk of each mass was clamped and excised before completion of the delivery while the maternal–fetal umbilical circulation remained intact. Once the lesions were excised, a baby girl weighing 3210 g was completely delivered, with Apgar scores of 9 and 9 at 1 and 5 minutes, respectively. To ensure adequate oxygenation, the neonatologist placed a nasopharyngeal tube connected to an oxygen source while the wounds were closed by the otolaryngologist. There was minimal bleeding requiring cautery and absorbable sutures to achieve adequate hemostasis. Then, the child was transferred to the neonatal intensive care unit. Feeding was started the same day, and the infant was discharged on the fourth day of life. At six months of age, the infant was doing well without any visible area of defect.

Case 2

A 40-year-old white woman, gravida 3, para 2, with gestational diabetes had an ultrasound examination at 29.5 weeks of gestation. A 1.9 $\times$ 1.4-cm intraoral mass was noted to fill the fetus's oral cavity. An amniocentesis performed at that time revealed a normal, 46 XX karyotype. Two weeks later, a repeat ultrasound found the mass to be increasing in size (2.1 $\times$ 1.9 cm). Therefore, the woman's care was transferred to the division of maternal–fetal medicine and the multidisciplinary team was consulted, as in case 1. The management plan that was agreed on was discussed with the family. At 38 weeks of gestation, an elective cesarean section was performed with the patient under general anesthesia, with the multidisciplinary team in attendance for immediate intrapartum evaluation and treatment of the infant. The operating room was equipped as in case 1. As the head was delivered, a pedunculated mass of approximately 2 $\times$ 3 cm was observed protruding from the maxillary alveolar ridge (Fig. 3). Also, there appeared to be a second raised area on the left side of the maxilla. Because these lesions were not obstructive, the delivery was completed after the obstetrician suctioned the nasal and oral cavities.

The infant cried spontaneously and was brought to a radiant warming table for further assessment and treatment. In addition to routine suctioning, only oxygen was needed through a nasopharyngeal tube. The Apgar scores were 9 and 9 at 1 and 5 minutes, respectively. On assessment of the infant by the otolaryngologist and consultation with neonatologist, it was determined that the mass could be safely resected at that time. With the patient under local anesthesia, the stalk was clamped, a ligature was placed at the bottom of the stalk, and the mass was excised. Next, small bleeders were identified, and hemostasis was obtained with a hand-controlled cautery. Then, the infant was admitted to the neonatal intensive care unit. Feeding began the next day, and the baby was discharged 2 days after delivery.

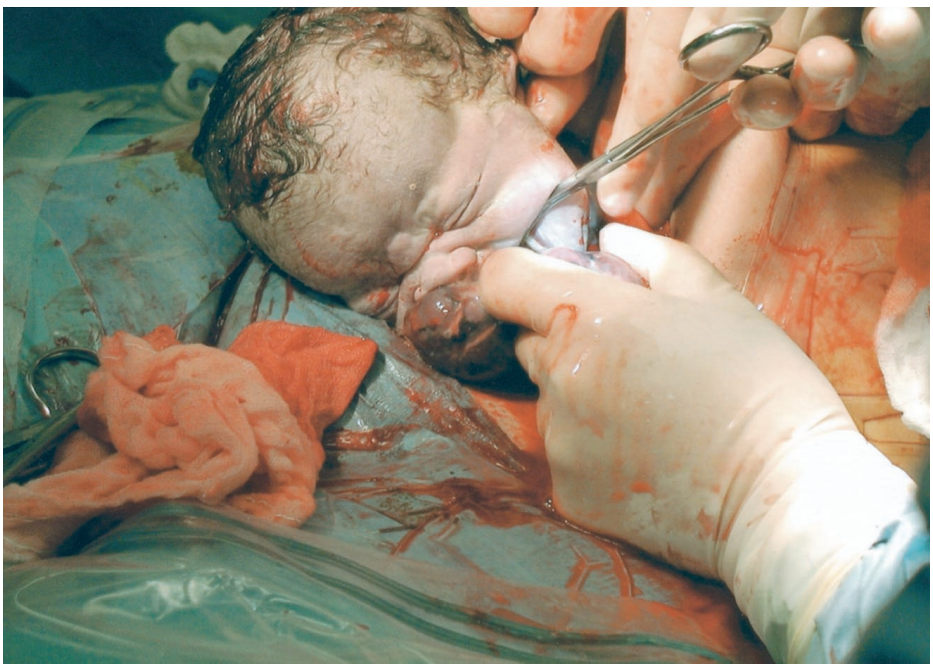

Fig. 2. Resection of intraoral masses at delivery. Only the infant's head is exteriorized from the uterus.

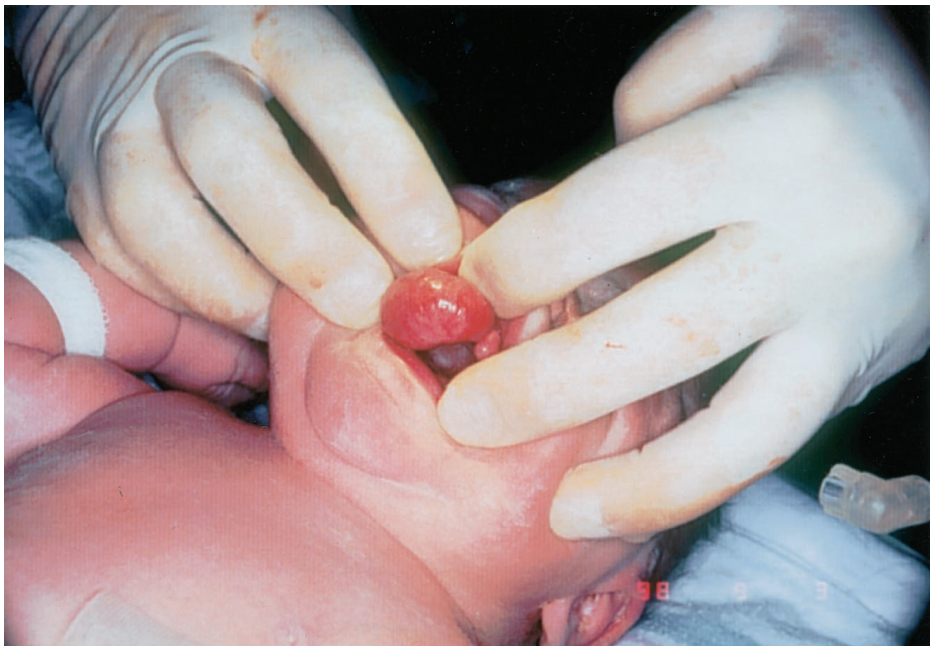

Fig. 3. Intraoral mass protruding from mouth of infant during examination after complete delivery.

RESULTS

Histopathological Findings

Pathological examination was similar in both cases. In case 1, the lesions were 5 × 5 × 4 cm and 3.5 × 3.5 × 2.5 cm (Fig. 4), whereas in case 2 the lesion was 2.5 × 2 × 1.5 cm. Histologically, both tumors were covered by a thin layer of squamous mucosa separated by a layer of fibrous tissue. They were proliferated by large polygonal cells with eosinophilic granular cytoplasm and regular nuclei with clear chromatin. There was minimal fibrous stroma. In some areas, there were abundant capillaries, some with dilated lumen (Figs. 5 and 6). Immunoperoxidase stains were negative for S-100 protein. The tumors were consistent with congenital epulis.

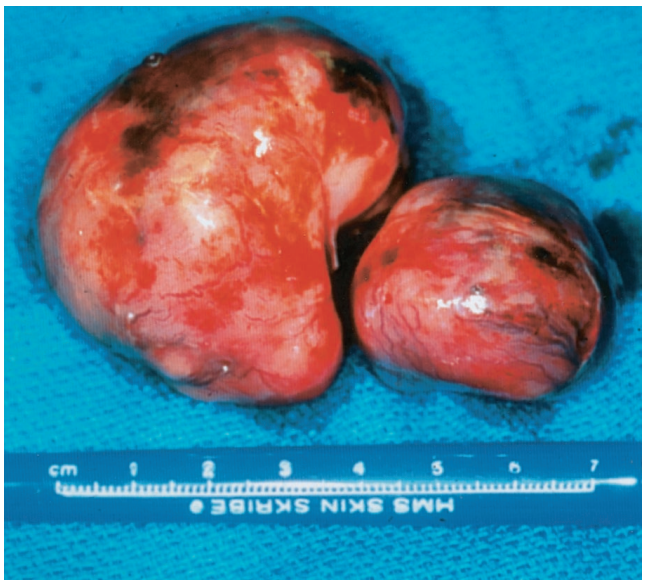

Fig. 4. Gross appearance of the cut surface of the two tumors resected from the oral cavity of infant in case 1.

DISCUSSION

Congenital epulis, also known as granular cell tumor, is a rare, benign intraoral tumor found exclusively in newborns and first described by Neumann[2] in 1871. Although there have been numerous cases, antepartum identification has been infrequent.[3] Previously, the earliest antepartum ultrasound identification of an oral mass subsequently identified to be a congenital epulis was at 31 weeks of gestation.[4] Congenital epulis has a 10:1 predilection for female newborns, and in 10% of cases there are multiple tumors.[5] Congenital epulis is usually an isolated finding, although cases have been reported that were associated with polyhydramnios,[6,7] neurofibromatosis,[8] XXX chromosome and polydactyly.[3]

In addition to congenital epulis, congenital intraoral masses include hemangioma, fibroma, granuloma, rhabdomyoma, rhabdomyosarcoma, and lymphangioma. The definitive diagnosis is determined by pathological examination. Radiographic and surgical examinations have found congenital epulis to be isolated to the soft tissue, not involving the underlying maxilla or mandible. The histogenesis of the tumor is uncertain. Theories include odontogenic, fibroblastic, histiocytic, mesenchymal, and neurogenic origins[1,9] The predilection in female newborns raises the possibility of a hormonal basis. However, immunohistochemical staining for estrogens and progesterone has been negative.[9] Size has varied from a few millimeters to 8.0 cm[6] with the tumor usually presenting as an encapsulated, multilobed, pedunculated pink mass arising from the maxillary or mandibular alveolar ridges.

Histologically, congenital epulis is similar to other granular cell tumors that may occur throughout the body. The tumor displays a tan, homogenous, smooth edge with

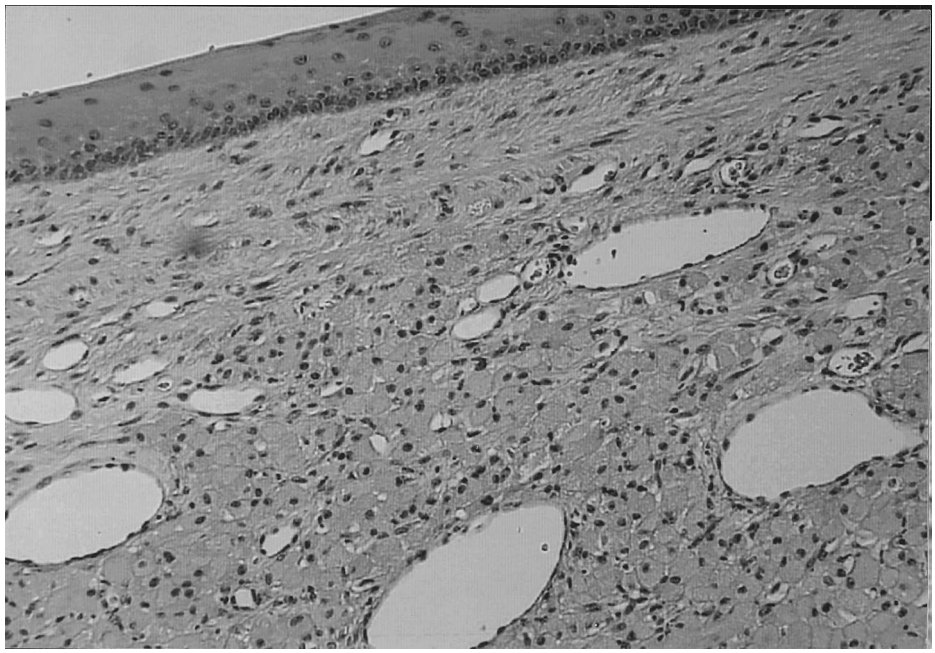

Fig. 5. Case 1: Tumor is separated from the regular covered squamous mucosa by a layer of fibrous tissue. Cells forming the tumor are large and have regular nuclei and abundant granular eosinophilic cytoplasm (H&E stain, original magnification ×125).

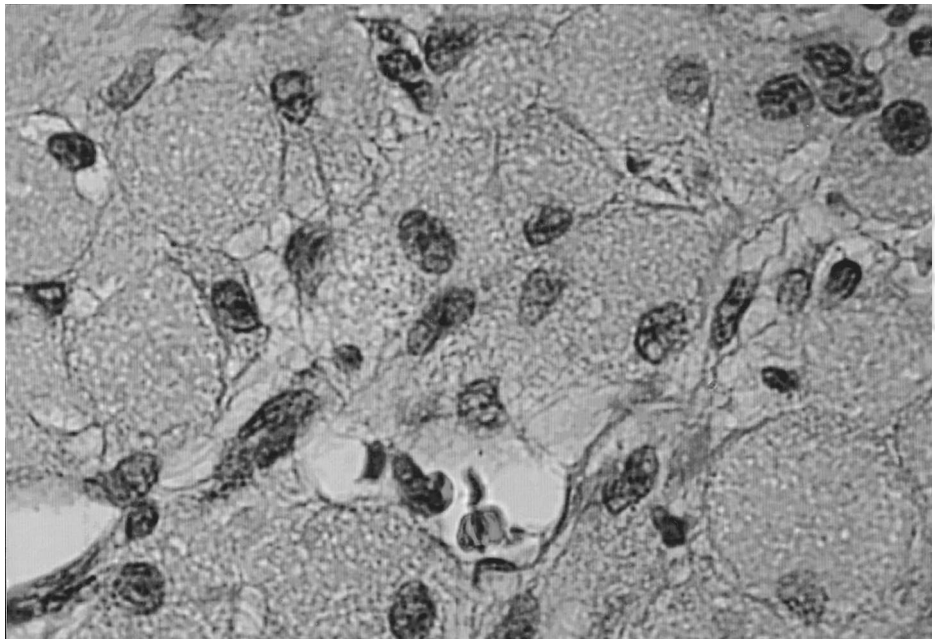

Fig. 6. Case 1: Larger magnification showing the tumor cells in case 1 with the characteristic granular cytoplasm and regular nuclei (H&E stain, original magnification ×300).

diffuse sheets and clusters of polygonal cells containing round, small nuclei with abundant, coarsely granular cytoplasm. Between granular cells there is a fine vascular network that accounts for a tendency to bleed. A congenital epulis is distinguished from more common granular cell tumors by the lack of pseudoepitheliomatous hyperplasia, absence of S-100 protein expression, and positive reaction to carcinoembryonic antigen and HLA-DR antigen.[1] Also, the tumor cells may be reactive for the macrophage marker CD68.[9,10] In contrast, noninfantile granular cell tumors usually present between the third and sixth decades in life. They occur diffusely throughout the head and neck region without predilection for either sex and show diffuse immunostaining for S-100 protein without positivity for carcinoembryonic and HLA-DR antigens.

Although congenital epulis is a benign tumor, a large epulis may cause significant problems for the newborn by interfering with respiration or deglutition, or both. Although there are reports of spontaneous regression, surgical removal is the treatment of choice for large lesions. In our two cases, antepartum ultrasound identification of these oral masses permitted assembly of a multidisciplinary team to ensure airway patency at birth and effect a rapid, simple removal of the tumors. In contrast, other authors have chosen to delay resection for several days and employ different modalities of surgery requiring gen-

eral anesthesia.[9] Resection of the masses in the operating room essentially allowed for complete management in the first minutes to the first hour of life, avoided a later return to the operating suite for definitive treatment, and resulted in a shortened hospital stay.

The option of ex utero intrapartum treatment (EXIT) should be considered in cases with possible obstructive lesions. This treatment consists of maintaining the uteroplacental blood flow during and immediately after cesarean section. First described by Zarella and Finberg[11] in 1990, EXIT has been used when significant airway obstruction was anticipated (e.g., pretracheal teratomas, epignathus, cystic hygromas, and after in utero tracheal ligation for congenital diaphragmatic hernia).[12] This consists of performing a cesarean section with the patient under general anesthesia. The use of general anesthesia and tocolytics provides adequate uterine relaxation, counteracting potential effects of intrapartum oxytocin producing uterine tone and extrusion of the placenta. Adequate uteroplacental flow can be maintained for up to an hour with maintenance of normal cord blood gases for the operative procedure on the infant. By contrast, lack of anesthesia can result in usual uterine contraction with extrusion of the placenta and with adequate fetoplacental circulation for as few as 5 minutes. Initially, only the infant's head and shoulder are delivered, and the placenta remains attached to the uterine endometrium. After the infant's airway is secured, the umbilical cord is clamped and the delivery of the infant is completed. The EXIT procedure has proven to be safe and efficacious, allowing establishment of an airway in a controlled manner as the placenta allows continued gas exchange during airway manipulation.

In our cases, an elective cesarean section was performed with a multidisciplinary team present. An EXIT procedure was not necessary because a quick, sharp resection of the lesions was possible in both cases. Indeed, we found (case 1) that definitive surgical treatment could readily be accomplished in a brief period on delivery of the head while an intact umbilical cord permitted adequate fetal–maternal circulation.

CONCLUSION

Careful antepartum imaging studies have identified in utero potentially obstructive lesions of the oral cavity. An interdisciplinary approach ensuring airway patency and simple, early resection has been presented. The ex utero intrapartum treatment of obstructive lesions should be considered. A controlled assessment and management of the airway at the time of delivery may eliminate more complicated procedures requiring general anesthesia and endotracheal intubation. From our experience, we think delivery room management of congenital epulis is most advantageous.

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