Prenatal Ultrasound Detection of Congenital Gingival Granular Cell Tumor

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Congenital gingival granular cell tumor in infants is a rare intraoral tumor of unknown histologic origin, but apparently related to the larger, but still uncommon, group of granular cell tumors seen in all age groups and multiple anatomic areas. We report the prenatal sonographic detection of such a lesion, imaged as a pedunculated extraoral mass extending from an intraoral source. Due to the potential for airway compromise, a prenatal diagnosis of a mass involving the airway should lead to delivery at a medical center prepared to handle that complication both temporarily, with airway maintenance, and ultimately, with surgical correction.

CASE REPORT

A 33-year-old gravida 2 para 1 was referred for a prenatal ultrasound (US) examination at approximately 38 weeks gestational age to assess the estimated fetal weight (EFW). Pertinent obstetrical history included a prior vaginal delivery of an 8-pound 8-ounce male, complicated by shoulder dystocia and postpartum hemorrhage. The US examination confirmed a gestation of 39 weeks based on biparietal diameter, head circumference, and femur length, with an EFW of 4270 g. The amniotic fluid volume was normal. A lobulated mass was noted arising from the right side of the oral cavity and protruding out of the mouth (Fig. 1), with distortion of the lower face and lips. The tongue was displaced to the left and posteriorly. Bright internal echoes suggested calcification and a teratoma (epignathus) was suspected.

Amniocentesis revealed an L/S ratio of 6.07 and DSPC of >600. The patient underwent elective cesarean section with delivery of a 9-pound, 11-ounce female with Apgars of 7 and 9 at 1 and 5 minutes. The infant had a spontaneous cry and good respiratory effort. A large, pink/grey pedunculated rubbery mass protruding from the mouth appeared to arise from the superior alveolar ridge on the right (Fig. 2). The palate, tongue, and lips appeared grossly intact. To insure airway patency, the infant was electively intubated.

As a teratoma was still suspected, a cranial US examination

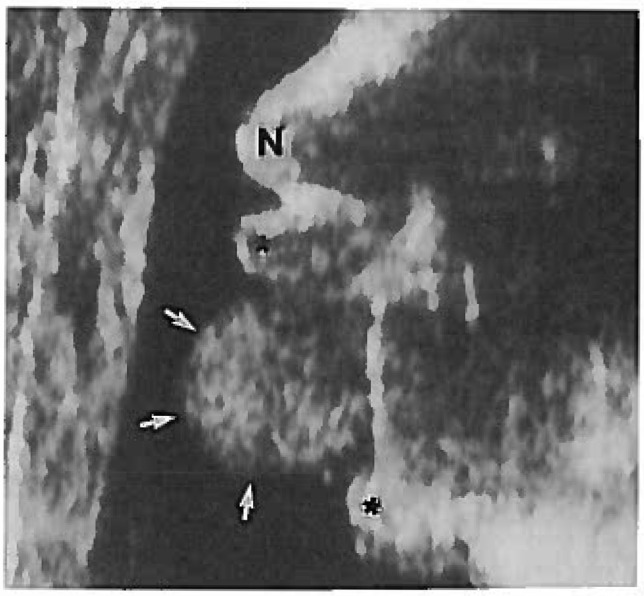

Figure 1 Midsagittal sonogram of the lower face shows the mass (*arrows*) protruding from the open mouth. N, nose; *asterisks* indicate lips.

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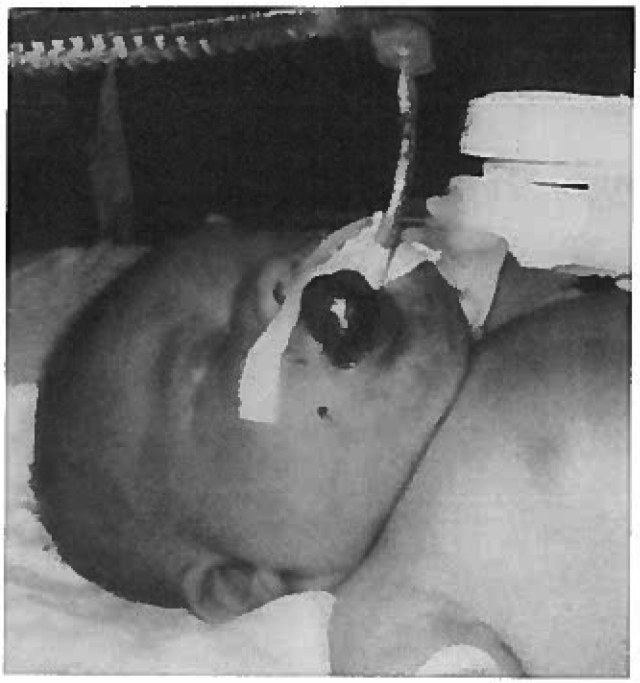

Figure 2 Intubated infant with lobulated mass protruding from the mouth.

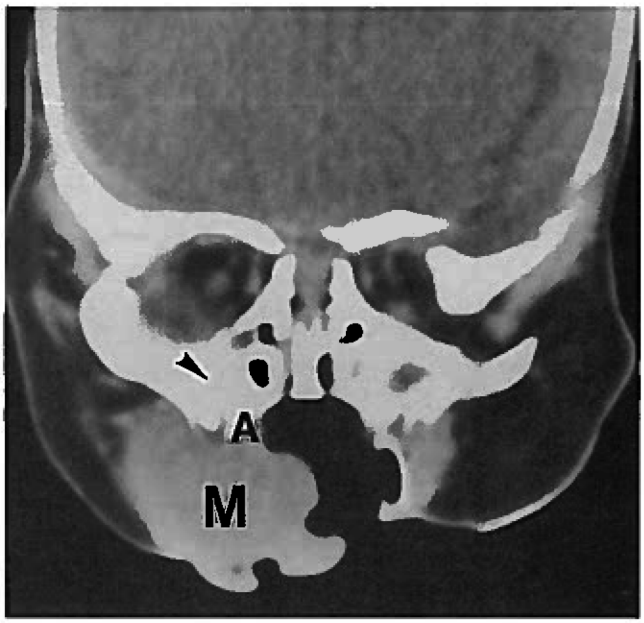

Figure 3 Coronal computed tomography scan through the face shows a lobulated soft tissue mass (M) extending from the right superior alveolar ridge (A). *Arrowhead* indicates a small tooth bud.

was performed to rule out intracranial extension, and was normal. For further diagnostic evaluation and exclusion of subtle intracranial extension, a computed tomography scan of the head and face was performed, confirming a lobulated 3 × 4-cm soft tissue mass arising from the right maxillary alveolar ridge, without cystic areas or calcification (Fig. 3). There was no erosion or alteration of the adjacent dental structures. The mass extended anteriorly to protrude through the mouth, but the tongue and mandible were intact. No intracranial extension was noted.

Surgical excision was performed on day 1, without complication. The infant was extubated on the first postoperative day had a normal respiratory effort. The postoperative course was uneventful, and the infant was discharged on day 3.

Pathologic evaluation of the specimen showed no areas of cystic change, hemorrhage, or necrosis. Microscopic examination revealed a benign tumor composed of large cells with granular cytoplasm and rare islands of odontogenic epithelium. The findings were representative of a benign congenital gingival granular cell tumor.

DISCUSSION

Congenital intraoral tumors are rare lesions. By far the most commonly described is a teratoma (epignathus), which usually arises from the oral cavity or pharynx.[1–5] Teratomas arising in or near the oropharynx are seen on US as solid masses often with areas of calcification and cystic change. The differential diagnosis for a mass arising in or near the mouth on a prenatal ultrasound should include teratoma, hemangioma, lymphangioma (cystic hygroma), fibrous tumor, neurofibroma, and granular cell tumor. Hemangiomas may be solid or cystic but arise from the subcutaneous tissues externally.[6] Pulsations may be seen in the vascular spaces on real-time US,[7] and Doppler may be an effective way to evaluate the blood flow. Lymphangiomas are predominantly cystic on US and tend to originate posteriorly in the neck.[8–10] There is one report of a fibrous tumor arising from the floor of the mouth[11] but this did not extend externally. Facial neurofibromas are subcutaneous tumors with localized nodular or diffuse soft tissue involvement, but are not pedunculated.[12]

Congenital gingival granular cell tumor (congenital epulis) has been previously described in the literature in newborns,[13–17] but only twice antenatally.[17,18] It is a benign soft tissue tumor that usually arises from the maxillary alveolar ridge anteriorly. The tumor can be pedunculated or broad based and is covered by smooth, nonulcerated mucosa. Microscopically, large sheets of granular cells and remnants of odontogenic epithelium are seen. While these tumors may regress spontaneously,[14,19,20] surgical resection is usually employed. There are no reports of postsurgical recurrence.[20,21] Congenital gingival granular cell tumor is found in the neonate, predominantly in females, and is closely related histologically to the granular cell tumor of older children and adults. The latter, however, has no sex predilection and can be found in many anatomic sites (skin, gastrointestinal tract, respiratory tract). While the histologic origin of both of these granular cell tumors is unknown, they are identical on

light microscopy, but can be differentiated by immunochemistry and electron microscopy evaluation.[22–24]

The prenatal evaluation of any fetal mass involving the head and neck should include assessment of the amniotic fluid. Polyhydramnios suggests an obstructive component to the gastrointestinal tract that could also involve the respiratory tract at birth, leading to respiratory compromise. While no polyhydramnios was present in our case, prudence dictated elective cesarean section delivery under controlled circumstances to allow for optimal handling of the neonatal airway prior to preoperative evaluation for the definitive surgery.

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