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Pediatric

Massive fetal cervical teratoma managed with the ex utero intrapartum treatment (EXIT) procedure

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ABSTRACT

Teratomas are rare congenital tumors typically comprising all 3 germ cell layers. Cervical teratomas arise in the neck and represent a minority of all teratomas. These are associated with high morbidity and mortality because of their propensity to cause airway obstruction. Demonstration on prenatal magnetic resonance imaging is uncommon, especially for a tumor of this size. Fetuses diagnosed with large neck masses are managed through cesarean section with the ex utero intrapartum treatment procedure to secure the airway, such as in our case of a large cervical teratoma in the female fetus of a 30-year-old mother who went into preterm labor.

Case report

A 30-year-old gravida 4, para 2 female was found to have a female fetus with polyhydramnios and a large right-sided neck and facial mass on prenatal ultrasound. Subsequent fetal magnetic resonance imaging obtained near 30 weeks of gestation demonstrated an approximate $12 \times 11 \times 9$-cm mass containing solid and cystic components (Fig. 1). It appeared to arise anteriorly within the face and cervical soft tissues, just below the orbits and nasal cavity, and extended anteriorly to the right of midline. The trachea and esophagus were displaced to the left lateral and anterior directions. Esophageal obstruction was evident with marked polyhydramnios.

Plans were made for an elective cesarean section close to term; however, the mother went into preterm labor at 30 weeks of gestation and required emergent cesarean delivery and ex utero intrapartum treatment (EXIT) procedure. During the EXIT procedure after delivery of the head first, attempts to secure the airway by direct laryngoscopy and then fiberoptic laryngoscopy were unsuccessful. An emergent tracheostomy was completed, positive pressure ventilation was started, and then the body was delivered. The Apgar scores were 5 at 1 minute, 6 at 5 minutes, and 7 at 10 minutes. She continued to require positive pressure ventilation for severe respiratory distress and was taken to the Neonatal Intensive Care Unit (NICU). The mass was a huge swelling protruding from the lower part of the face, with significant deformity of the face, airway, and neck (Fig. 2).

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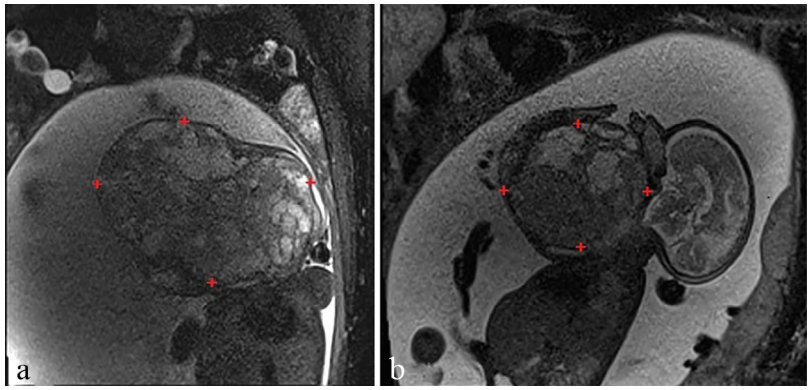

Fig. 1 – Fetal magnetic resonance imaging (MRI) axial (A) and sagittal (B) T2-weighted Half-Fourier Acquisition Single-shot Turbo Spin Echo (HASTE) images show a large 12 × 11 × 9-cm mixed solid and cystic anterior face and neck mass (outlined +).

An ultrasound of the neck (Fig. 3) performed at 2 days of age demonstrated the large mixed solid and cystic mass that also contained foci of calcifications, which was consistent with teratoma. A follow-up ultrasound performed at 2 weeks showed interval enlargement of multiple cystic components in the mass, which led to an aspiration biopsy. Histopathology of the aspirated fluid was consistent with a teratoma. She remained in the NICU requiring very high levels of ventilator support, had significant coagulopathy and superficial skin breakdown over dependent parts of the tumor, and died at 6 weeks 4 days of age.

Discussion

Teratomas are congenital tumors that contain tissue components from all 3 germ cell layers: mesoderm, ectoderm, and endoderm. They are the most common neonatal tumor with an incidence of about 1 in 20,000-40,000 live births and affect females more often than males [1,2]. Teratomas typically arise during the fourth or fifth week of gestation in midline regions at intra or extragonadal locations [3]. They are most commonly found in the sacrococcygeal region, whereas cervical teratomas represent only 2%-9% of congenital teratomas [4]. In the neonatal and infant population, these are rarely malignant but are associated with a high morbidity and mortality [5]. Cervical teratomas can compress oropharyngeal structures and impair fetal swallowing, resulting in airway obstruction and polyhydramnios. The tumor also pulls the fetal lungs up toward the apex, which prevent them from expanding and developing properly, causing pulmonary hypoplasia. Increased tumor size is associated with more severe pulmonary hypoplasia. Polyhydramnios can lead to preterm labor, and pulmonary hypoplasia can cause severe hypoxia, which can be fatal, as in this case.

The pathogenesis is not completely understood and there are several theories. These are classified histologically as mature or immature with grade based on the percentage of immature tissue in the tumor. Mature teratomas have well-differentiated tissues, which include skin, hair, nails, teeth, cartilage, bone, and nerve tissue. Immature teratomas are

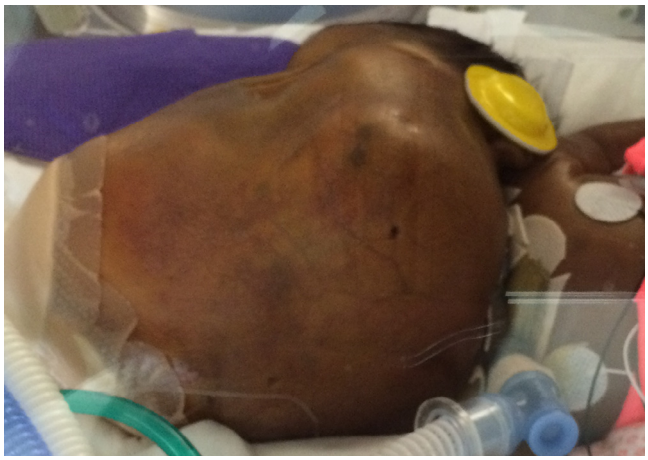

Fig. 2 – Photograph shows the gigantic mass occupying most of the face and anterior neck in this neonate. The head appears hyperextended.

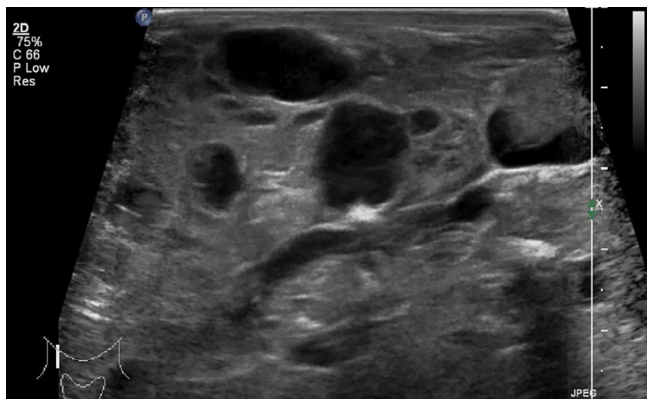

Fig. 3 – Ultrasound grayscale sagittal image of the neck shows a large heterogeneous solid and multicystic mass. There are multiple echogenic foci within, some of which display posterior acoustic shadowing, consistent with calcifications. Internal blood flow was indicated by the presence of color Doppler within (not shown).

composed of undifferentiated tissues, particularly neuroepithelial tissue [6].

On imaging, cervical teratomas appear as solid or mixed cystic midline or anterolateral neck masses commonly characterized with ultrasound or magnetic resonance imaging. Fetal magnetic resonance pulse sequences include axial and sagittal T1-weighted images, fast spin echo T2-weighted images or fat suppression T2-weighted images. After birth, contrast-enhanced sequences may be done. A teratoma appears as a well-circumscribed complex lesion, heterogeneous on T1- and T2-weighted sequences, depending on what tissue types constitutes the mass. Calcifications are often present and are relatively specific for teratomas; however, the absence of calcifications does not exclude the diagnosis. These teratomas often occur in close proximity to the thyroid gland. The ipsilateral thyroid gland might be absent or surrounding the teratoma. The main differential diagnosis for a large neck tumor is cystic hygroma/lymphangioma [6]. On ultrasound, this appears as septated fluid-filled collections and solid components are rare. It also arises more posteriorly on the neck than teratomas. Other less common differential considerations include goiter, neuroblastoma, hemangioma, and sarcoma.

Neonates diagnosed with a large cervical teratoma are best managed by C-section delivery with EXIT procedure, a specialized surgical procedure that allows the neonate to stay on placental support while their airway is secured. It was first described by Drs Adzick and Harrison in the 1990s as a method for delivering neonates with airway compression [7,8]. The EXIT procedure involves a multidisciplinary approach and is done under general anesthesia with maternal and fetal monitoring. First the placenta is mapped with ultrasound to locate an optimal incision site to avoid placental abruption. A hysterostomy is performed, and the uterine cavity is continuously filled with warmed lactated Ringer's solution to prevent umbilical cord compression. The fetal head, neck, and upper torso are delivered while the body remains in the uterine cavity. The otolaryngology team then accesses the airway via direct laryngoscopy. If the airway is difficult to access, as in our patient, rigid bronchoscopy or emergent tracheostomy is then attempted. Mechanical ventilation is started and surfactant is administered for neonates less than 34 weeks gestation [8]. Once the neonate is stabilized, the umbilical cord is clamped and the rest of the body is delivered. Care is transferred to the NICU. Management of the neonate after delivery then depends on the degree of respiratory distress [9]. If the neonate is stable and the airway is properly secured, then the teratoma can be resected soon after delivery or later. If the neonate is unstable then resection should be deferred until the baby is sufficiently improved to tolerate surgery.

For neonates who survive to delivery, airway obstruction and respiratory distress are the most life-threatening complications. Coagulopathy due to thrombosis in areas of the tumor is also a concern. Skin breakdown and wound infection may occur. Large neck masses also disrupt normal cervicofacial development leading to dysfunction of facial structures, neck musculature, and cranial nerve dysfunction from stretching of nerve fibers [10]. Survivors are at risk for disabilities from cranial nerve dysfunction, such as problems with speech and eating. Many require tracheostomy and gastrostomy tubes. Even after resection of the teratoma, most have moderate to severe disfigurement. Cervical teratomas are not associated with cognitive dysfunction provided that cerebral hypoxia and neurologic injury from the delivery are avoided.

Conclusion

Cervical teratomas are usually seen in neonates and appear as solid or mixed cystic masses, classically with calcifications. These can grow to be large enough to compromise the airway and esophagus, leading to pulmonary hypoplasia and polyhydramnios. In that situation, the EXIT procedure delivery can be attempted to secure the airway while the fetus is still supplied by the placenta. Unfortunately, the neonate is still at high risk for respiratory distress and prognosis is poor based on degree of neck mass distortion.

Consent

Parental informed consent was obtained for the publication of this case.

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