Congenital Giant Cervicofacial Teratoma: A Case Report from Diagnosis in Utero to Surgical Excision

Heather E. Lee, M.D., Matthew B. Hirsch, M.D., Swapna K. Chandran, M.D., Jeffrey M. Bumpous, M.D.
Department of Otolaryngology, Head and Neck Surgery, University of Louisville School of Medicine

Introduction

Head and neck teratomas account for approximately 5% of all neonatal teratomas. Giant cervicofacial teratomas are a rare congenital anomaly with an incidence of one in 40,000 live births. Due to their location and size, they pose challenges to delivery, airway management, and surgical resection. We report a case of a 31-year-old G1P0 pregnant woman undergoing routine maternal ultrasound who was found to be carrying a fetus with a right cervicofacial mass. We discuss in utero diagnosis and subsequent multidisciplinary management of delivery and airway planning which involved extrauterine intrapartum treatment, or EXIT procedure. A strategy for definitive treatment by surgical excision is also presented.

Case Report

A healthy 31-year-old G1P0 pregnant woman undergoing routine maternal ultrasound was found to be carrying a fetus with a right cervicofacial mass which was further assessed with fetal MRI. The MRI showed fluid in the airway and the stomach, suggesting patency of the trachea and esophagus. However, the mother displayed clinical signs of polyhydramnios.

Prior to delivery, a multidisciplinary team included otolaryngologists, obstetricians, anesthesiologists, and neonatal intensivists was assembled for airway management. The neonate was delivered at 37 weeks gestation. Extrauterine intrapartum treatment, or EXIT procedure was performed with the patient partially delivered via hysterotomy. While still on uteroplacental circulation, the neonate’s airway was secured via oral intubation. The patient was then taken for tracheostomy and biopsy of the large cervicofacial mass. The biopsy returned as mature teratoma, negative for malignancy.

CT and MRI scans of the head and neck performed after delivery showed a large, heterogeneous right neck mass extending from the area of foramen ovale to the C4 level, occupying the masticator space, oral cavity, and oropharynx displacing the right mandibular ramus and condyle anterolaterally. The external carotid artery and its branches were laterally displaced by the lesion.

Surgical resection was performed at 3 weeks of age due to concerns for continued enlargement of the teratoma. The skin overlying the mass was first ellipseed. The facial nerve was seen to be splaying across the mass and was preserved, although resection required extensive mobilization. The mass extended inferiorly to the level of the hypoglossal nerve, which overlies the mass was first ellipsed. The facial nerve was preserved, although resection required extensive mobilization. The mass was then used as a full thickness skin graft for the intraoral mucosal defect. Final pathology returned as mature benign teratoma.

Discussion

- On fetal MRI, teratomas often appear as multilobular, heterogeneous masses with solid, cystic, and fatty components, as well as calcifications.
- Ultrasound plus fetal MRI is the preferred strategy for imaging fetal neck masses. MRI can assist in establishing a diagnosis and assessing the patency of the upper aerodigestive tract.
- In a fetal MRI, T2 hyperintense signal should fill the oropharynx, hypopharynx, trachea, and esophagus. Its absence should raise suspicion for compression of the airway by the mass.

The standard approach to the airway during EXIT procedures:

- The obstetric surgeon delivers the head, neck, and one shoulder via a low hysterotomy. The umbilical cord is left unclamped. Inhalational anesthesia is utilized to facilitate uterine relaxation. Fetal monitoring is established, and then the otolaryngologist addresses the fetal airway.

1. Direct laryngoscopy is usually first attempted. After the patient is intubated, delivery is completed.
2. If laryngoscopy is not successful, rigid bronchoscopy can be attempted.
3. An endotracheal tube can also be passed over a guidewire or flexible endoscope.
4. If the infant cannot be intubated via the above methods, tracheostomy should be performed immediately while still on maternal support.
5. Some centers have reported success utilizing transcervical ultrasound to identify the tracheal lumen followed by retrograde intubation using a Seldinger technique.

Conclusion

Benign cervicofacial teratomas are typically diagnosed with fetal ultrasound and MRI. Perinatal evaluation can give valuable information about the characteristics of the mass and its effect on the airway. This allows for appropriate multidisciplinary strategies at birth for delivery and airway management which may include an EXIT to secure the newborn's airway while still on maternal support. Surgical resection is often the mainstay of treatment.

References