Complete Peripartum Airway Management of a Large Epignathus Teratoma: EXIT to Resection

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Case Report

A 34-year-old Caucasian female was referred to pediatric otolaryngology after a routine prenatal ultrasound showed a large oral mass. This was the woman’s first pregnancy and there had been no problems during the pregnancy up until that point. There was no family history of genetic disorders. Due to the findings on the ultrasound, the patient had a prenatal MRI, which showed a large oral mass, consistent with a teratoma, extending from the skull base through the oral cavity.

It was determined that an EXIT procedure would be the safest approach to ensure that the neonate had a secure airway. The EXIT procedure was performed at 37 weeks of gestation. Once an airway was secure, close examination revealed a mass that appeared to have a lip, hard palate, and uvula. The neonate was stabilized in the NICU. At 3 days of age, the infant underwent endoscopic assisted transoral resection of the mass.

Pathology results showed the oropharyngeal mass contained all three germ layers, including smooth and skeletal muscle, salivary glands, bone, cartilage, teeth, and nerves. These tissues were mature and consistent with patient’s age. Genetics evaluation revealed no chromosomal abnormalities.

Discussion

An epignathus is a teratoma of the oropharynx that arises from the skull base or hard palate. Teratomas, by definition, are neoplasms that consist of all three germ layers, and in the case of epignathus teratomas, the germ layers are mature differentiated tissue. Congenital teratomas occur in approximately 1 in 4000 live births, while epignathus teratomas are much rarer, estimated between 1 in 35,000 and 1 in 200,000 live births. These tumors can occupy a significant portion of the oral cavity and therefore, have historically had a high mortality rate secondary to asphyxiation. With improved prenatal imaging, including 3D ultrasound and MRI, these tumors are being diagnosed early in the pregnancy which allows for adequate planning of delivery to ensure a safe airway can be established. Ex utero intrapartum treatment (EXIT) procedure has become the standard of care for safe airway management in these cases.

Conclusion

Epignathus teratomas are rare congenital oropharyngeal tumors that were considered non-survivable in the past. However, since around 1990 and the development of the EXIT procedure, these tumors are becoming manageable and patients have an improved prognosis. As this case indicates, a significant amount of planning is involved for the EXIT procedure and definitive surgical treatment. A well-executed EXIT procedure can establish a safe airway and allow for stabilization of the neonate prior to proceeding with resection of the tumor.