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Perinatal management of fetal oral tumors

Alberch A, Bonmatí A, Martinez- Diago C, Rodó C, Arévalo S, Laguillo G, Adrados C, Teixidor M, Maroto A
Hospital Universitari de Girona Dr Josep Trueta, Girona, Spain

Objective

The aim of this study is to review fetal oral tumors based on a clinical case and to show the prenatal imaging progression.

Methods

Case description of an oropharyngeal teratoma diagnosed prenatally and review of the literature of congenital oral tumors.

Results

A 34-year-old pregnant woman, with a first trimester ultrasound within normality, was screened for fetal malformations at 20 weeks of gestation at Hospital Universitari Dr Josep Trueta, showing an exophytic pedunculated solid-cystic multilobed tumor without vascularization of 34 x 14 x 10 mm. The tumor seemed to affect the upper gum and palate and did not involve the dental alveoli, tongue or impediment to swallowing. Genetic testing was performed by amniocentesis with normal results. The ultrasound follow-up showed a progressive growth of the tumor. At 30 weeks of gestation, the measurement of the external part of the tumor was 45 x 20 mm. Although it was impossible to determine the internal extension of the mass, a polyhydramnios was identified. The fetal MRI reported an extensive tumor located in the oro-nasopharynx and oral cavity with probable nasosinusal extension, which protrudes externally through the mouth and completely obstructs the airway, suggesting a possible epignathus teratoma. In view of these findings, the case was discussed with a multidisciplinary team of Hospital Universitari Vall d'Hebron in Barcelona to assess the best perinatal care management, with an EXIT (Ex Utero Intrapartum Treatment) procedure being proposed. At 33 weeks of pregnancy an amniodrainage was performed due to the woman's discomfort because of polyhydramnios. At 36 weeks, she was admitted to the Hospital Vall d'Hebron for a premature rupture of membranes. A new MRI was performed showing an increase of the fetal tumor volume (by 50% compared to the previous MRI). An EXIT cesarean section was performed delivering a newborn of 2700g. A tracheostomy was performed before the cord clamping. After 24 hours, a fragmented resection of the mass was executed until reaching the pedicle. During the surgery, a cleft palate secondary to the tumor was found. The pathological anatomy result showed a mature teratoma. The hospital discharge was at 58 days. Currently, the child is 10 months old, he is receiving speech therapy and he is pending surgery of the rest of the nasal part of the teratoma. Congenital oral masses are uncommon and mostly benign. However, the potential airway obstruction of these tumors may result in adverse perinatal outcomes. Epignathus teratomas are rare tumors which originate in the region of the palate or pharynx and are known as Rathke pouch. This malformation is found in approximately 1: 35.000–1: 200.000 live births, accounting for 2–9% of all teratomas. They may be associated with other malformations such as a cleft palate and bifid tongue or nose. Prenatal imaging by ultrasound and MRI allow establishing the extension of the tumor and planning the perinatal management. Early intervention for ventilation is necessary, and immediate tracheostomy is often required.

Conclusion

The prenatal diagnosis of epignathus teratoma and the perinatal coordination are crucial to transform a potentially fatal neonatal emergency into a controlled environment in order to ensure a better outcome.