ABSTRACT

Objectives: To highlight the uncommon presentation of congenital thyroid teratoma undetected by prenatal ultrasound and to review airway and operative management for congenital cervical teratomas

Study Design: A retrospective case review of a single patient with congenital teratoma of the left neck.

Methods: The history, physical, imaging, operative, and follow-up findings of the patient are reviewed. Airway and operative management for congenital teratoma are discussed.

Results: A 37-week gestation female was born by spontaneous vaginal delivery and found to have a large left neck mass. One minute Apgar score was 2 and the infant was intubated with a 2.5 ETT. 18-week prenatal ultrasound had not detected a neck mass. Preoperative CT and MRI revealed a 7 x 6 x 6 cm left neck mass consistent with congenital teratoma. Initial alpha fetoprotein level was 47,000 ng/ml. On day of life #5, the patient was taken to the OR for surgical removal. Direct laryngoscopy revealed moderate tracheal compression. The mass was completely excised, appearing to arise from the left thyroid lobe. Continuous left recurrent laryngeal nerve monitoring revealed absence of activity as it was thinned by the teratoma. Pathology was consistent with primary congenital thyroid teratoma. At 18-month follow-up the patient has persistent left vocal cord paralysis with no stridor or feeding difficulties. There is no evidence of recurrent teratoma and a normal alpha fetoprotein level.

Conclusions: Congenital teratomas of the neck are uncommon anomalies that require careful management of the airway to ensure survival. Operative technique can successfully remove the mass and relieve the airway obstruction.

DISCUSSION

Cervicofacial teratomas are extremely rare neoplasms: their incidence is one in 20,000 live births. Head and neck teratomas account for approximately 5% of all neonatal teratomas. They are developmental tumors, derived from embryonic origin, comprising elements from all three germinal layers. The histologic appearance of these tumors is most often benign. Cytologic characteristics of immaturity in congenital cases is not considered a sign of malignancy. However, malignant transformation of these lesions and the occurrence of metastases at birth can occur. Cervical teratomas may cause severe airway obstruction. A 1988 review by Jordan and Gauderer described three groups with cervical teratomas: stillborns, newborns with respiratory distress, and newborns without respiratory distress. A 1988 review by Jordan and Gauderer described the mortality in three groups with cervical teratomas: stillborns (100%), newborns with respiratory distress (43.4%), and newborns without respiratory distress (2.7%). With the advent and improvement of prenatal ultrasound and subsequent fetal MRI, the detection and airway management of cervical teratomas has vastly improved. In these situations two procedures may be performed: EXIT (Ex Utero Intrapartum Treatment) or OOPS (Operation On Placenta Support). Securing the airway is then accomplished by either endotracheal via direct laryngoscopy/bronchoscopy or operative tracheostomy. In our current patient, the ultrasound at 18 weeks gestation did not demonstrate the neck mass and fortunately the neonatal team was able to secure the airway with a small ETT. Once the airway is secure, the patient should be medically stabilized and preoperative imaging obtained. Ultrasonography can reveal solid and cystic structures within a heterogenous mass with calcifications in up to 50% of cases. CT can provide more complete information regarding invasion and anatomical extension of tumor. MRI has further advantage due to its multplanar capacity, high signal intensity from fat on T1-weighted images, and high contrast resolution. The operative goal is to completely remove the neoplasm in a timely fashion. Prognosis is good with complete excision. Lymph node metastasis or local relapse suggesting metastases with elevation of alpha fetal protein after tumor removal have been reported. Our patients AFP level was elevated at diagnosis, returned to normal after resection, and continues to be normal. A recent review from Children’s Hospital of Boston described 11 cases of primary thyroid teratomas in children. As in our patient, the pathology demonstrated an intimate admixture of thyroid and other tissues with or without a surrounding fibrous pseudocapsule, strongly suggesting thyroid as origin. Congenital cervical teratomas present a significant airway challenge. When antenatal diagnosis allows, preparation for intervention with anesthesia, perinatology, neonatology, and otolaryngologist/surgeon is essential. After securing the airway, proper preoperative studies provide for a timely operative intervention and good long term prognosis for these infants.