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Mature and immature pediatric head and neck teratomas: A 15-year review at a large tertiary center

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ABSTRACT

Introduction: Pediatric head and neck teratomas account for less than 4% of congenital teratomas. The distinct presentations and outcomes of mature and immature head and neck teratomas have not been well established.
Objectives: To review the management and outcomes of pediatric head and neck teratomas. To distinguish differences between mature and immature tumors with respect to the age at presentation and surgery, tumor size and location, alpha fetoprotein (AFP) levels, airway management, and recurrence.
Methods: A 15-year retrospective chart review of patients treated for head and neck teratomas at Texas Children's Hospital was performed. A total of 20 patients were included. Wilcoxon rank and Fisher's exact tests were used for statistical analysis.
Results: Immature teratomas were associated with both higher AFP levels (80800 ng/ml, $p = 0.02$) and maximum tumor dimensions (14.4 cm, $p = 0.0034$) than mature teratomas (24400 ng/ml and 6.44 cm). Patients with immature tumors were younger at the time of surgical resection (19.8 days, $p = 0.025$) compared to those with mature tumors (348 days). 89% of immature teratomas involved anterior neck localization compared to 27% for mature teratomas ($p = 0.0098$); 88% of the immature teratomas required an EXIT (Ex Utero Intrapartum Treatment) procedure compared with 40% of the mature teratomas ($p = 0.0656$). Recurrence was noted in only two cases: an immature teratoma 51 months after incomplete resection and a mature teratoma 33 months after complete resection. Long-term consequences of surgical resection included cleft palate (38.9%), dysphagia (33.3%), facial nerve paresis/paralysis (16.7%) and tracheotomy (16.7%).
Conclusion: Immature teratomas had higher AFP levels, tumor dimensions, frequency of anterior neck localization, and requirement of EXIT than mature teratomas. Given that there was no significant difference between the recurrence rates of immature and mature teratomas, follow-up vigilance should be maintained equally regardless of tumor maturity.

1. Introduction

Teratomas are usually well circumscribed masses which vary widely based on composite tissue layers; they may have both cystic and solid areas with cartilage, bone, and pigmented areas being distinguishable [1]. Based on Grosfeld et al. series, approximately 57% of teratomas were sacrococcygeal while 25% were ovarian [2]. Barksdale et al. noted the rarity of head and neck teratomas: only 6% of teratomas across 6 series were localized to the head and neck [3]. Within this subset, cervical teratomas were the most common followed by teratomas of oropharyngeal and nasopharyngeal localization [4]. There is often radiographic evidence of calcified tissue in the mass suggestive of teratomas [5]. A work-up should be initiated to rule out other types of neck masses on initial presentation: lymphatic malformation, branchial cleft cyst, thyroglossal duct cysts; in comparison with this group, teratomas are the most rare [5].

Although rare, head and neck teratomas can have major consequences especially in the perinatal period. The clinical manifestations of head and neck teratomas are a direct extension of the anatomic sites affected via mass effect: nasopharyngeal, oropharyngeal, cervical, temporal bone, infratemporal fossa, orbital, or intracranial. If the trachea is obstructed *in utero*, there is an increased risk for pulmonary hypoplasia [6]; failure of normal neonatal deglutition *in utero* often results in sonographically detectable polyhydramnios and may precipitate increasing abdominal girth and respiratory discomfort in the mother [7]. Orbital teratomas can cause pronounced exophthalmia

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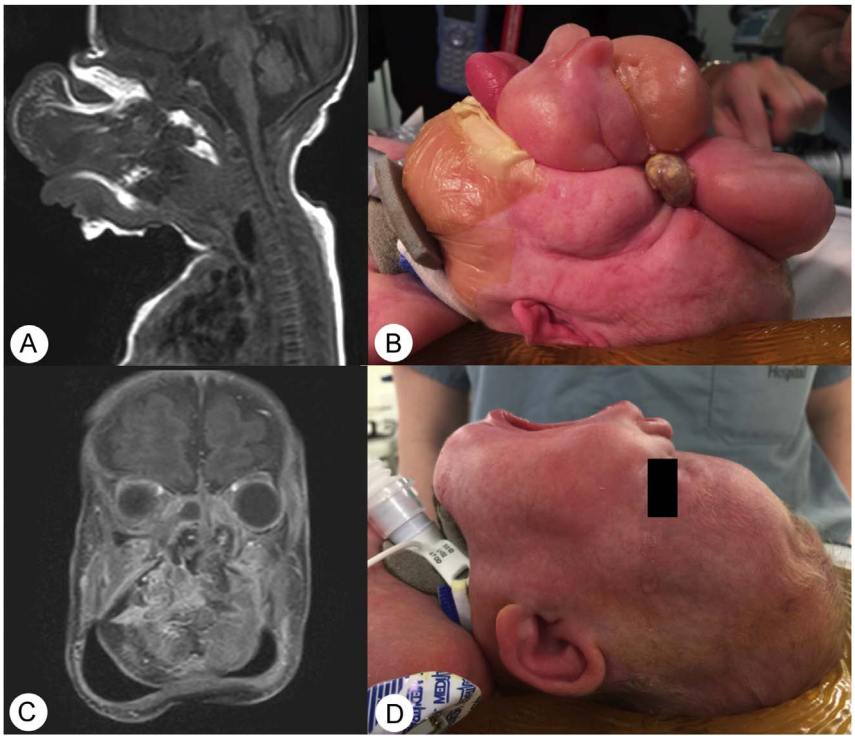

Fig. 1. This patient underwent an EXIT (*ex utero intrapartum treatment)* to airway; a tracheotomy was necessary since the ossified oropharyngeal teratoma was difficult to excise during the EXIT procedure. The patient was decannulated at 6 months with normalization of the facial structures; the cleft was repaired at 1 year of age. **(A)** Sagittal T1 and **(C)** Coronal T1 weighted images show an obstructing oral cavity and oropharyngeal teratoma. **(B)** There is extensive mass effect on the airway, oral cavity, and oropharyngeal structures that is relieved after complete teratoma resection **(D)**. Images **(B)** and **(D)** have the same orientation.

while teratomas localized to cervical, oropharyngeal, or nasopharyngeal sites may lead to dysphagia, feeding difficulties, or respiratory distress (Fig. 1) [5].

One of the most frequent complications of pediatric head and neck teratomas is airway compromise. In the neonatal period, airway obstruction can be overcome using the EXIT (*Ex Utero Intrapartum Treatment*) procedure where the airway is secured while the fetus is still on placental support [8,9]. Complete resection of a teratoma can be performed during the EXIT procedure itself or in the post-natal period [10]. Regardless of the tumor maturity, the primary goal of surgical intervention is complete resection with tumor-free margins. Failure to completely resect the tumor results in an increased risk of tumor recurrence [10,11]. However, complete tumor resection can be extremely challenging as it may lead to severe morbidity and complications from surgical intervention: cranial nerve palsies, hyponasal speech, mandibular weakness, vocal cord paralysis, facial reconstruction, and tracheotomy dependency [8,10]. After complete resection, tumor recurrence may be detected by monitoring for an increasing trend in alpha-fetoprotein (AFP) levels [12]. However, patient follow-up after tumor resection is a controversial topic; it is unclear whether tracking AFP levels, repeating imaging, or looking for changes in clinical presentation may be the best way of determining tumor recurrence. There is no consensus on the length of required patient follow-up and types of diagnostic approaches that are useful for tracking early recurrence.

On histological analysis, teratomas are identified based on the presence of at least two of the three embryonic germ cell layers: endoderm, mesoderm, and ectoderm. The degree of tissue differentiation determines the different morphologies of these tumors: a range from primitive somatic structures to well-developed fetus-in-fetu structures [13]. Teratomas may be further classified by their fractional composition of mature and immature elements. The increased presence of embryonic tissue elements such as primitive neuroepithelial and neuroglial cells indicates an immature teratoma, which is graded I to III based on criteria set forth by Norris et al.; mature teratomas have well-differentiated tissues and are grade 0 by definition [3,13,14]. A teratoma is deemed malignant based on the potential for metastasis, but malignancy does not directly correlate with tumor immaturity [15]. Metastases from congenital cervical teratomas may involve solely mature neural tissue or a mixture of mature and immature tissue elements [15]. If foci of yolk sac tumor (malignant germ cell tumor), choriocarcinoma or embryonal carcinoma are identified, the teratoma is classified as malignant [16,17].

With regards to pediatric head and neck teratomas, there have not been any prior studies that have analyzed tumor maturity versus recurrence risk. Understanding whether such a correlation exists is extremely important as the required follow-up management should be different if there is a significant difference in recurrence risk based on tumor maturity. The purpose of this review is two-fold. The first objective is to determine whether the recurrence rates and complications of mature and immature teratomas are significantly different in the long-term follow-up period. The second objective is to identify any differences between mature and immature pediatric head and neck teratomas with regards to clinical presentation: time to surgery, tumor size, AFP levels, and airway management including EXIT procedure.

2. Material and methods

The study protocol was approved by the Baylor College of Medicine Institutional Review Board. A 15-year retrospective chart review was performed at Texas Children's Hospital to identify cases of pediatric head and neck teratomas. The keyword “teratoma” was used to search the radiology (iSite) database; cases were selected from the filtered results based on suspicion of a head and neck teratoma per radiology report. The pathology database was reviewed to ensure that no cases were missed. Inclusion criteria for the study included age less than 18 years at initial presentation and teratoma documentation per pathology review. In general, patients were followed every six to twelve months to monitor for recurrence. However, the interval varied if there were complications or a requirement for a tracheostomy. Unfortunately, many patients were lost to follow-up due to non-compliance. With regards to monitoring for complications of surgical resection, patients with a tracheostomy or having any neck surgery routinely underwent flexible laryngoscopy and swallow studies. Audiograms were obtained based on the location of the teratomas. For