Pediatric Teratoma and Dermoid Cysts

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KEYWORDS

• Pediatric • Teratoma • Dermoid cysts • Head & neck

KEY POINTS

- Teratomas and dermoid cysts are germ cell neoplasms that can occur in the cervical and craniofacial regions.
- Presentation of these neoplasms varies in degree of severity, from cosmetic deformities to airway distress requiring emergent intervention.
- Nasal lesions (particularly if suspicious for a nasal dermoid) require imaging before biopsy to assess for intracranial extension.
- Treatment consists of airway management if respiratory distress is present, and early surgical intervention.
- Postoperative follow-up is required to monitor for recurrence.

TERATOMAS

Epidemiology

The word "teratoma" is derived from "teraton," a Greek word meaning "a monster," highlighting what the ancients thought about these maldevelopments.[1] The incidence of teratomas is 1:4000 births.[2] No sex predilection for teratomas in the head and neck region has been observed.[3] The etiology of teratomas is not fully understood; however, they likely occur in part when individual pluripotent cells fail to complete migration and continue dividing in an aberrant location, typically along the midline.[4] Teratomas have been found to occur in isolation or in association with other anomalies. Examples of previously reported comorbidities include central nervous system lesions, Klinefelters, Trisomy 13, Trisomy 21, congenital heart defect, Beckwith-Wiedemann syndrome, and cleft lip and palate.[1]

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Abbreviations	
AFP	Alpha-fetoprotein
CT	Computed tomography
EXIT	Ex utero intrapartum treatment

Classification

Teratomas are classified by the anatomic location in which they occur. General anatomic classifications include the following:

- Gonadal: teratomas involving testis or ovaries
- Extragonadal: teratomas located in regions such as sacrococcygeal, mediastinal, gastric, retroperitoneal, intracranial, cervical, and craniofacial.[1]

The sacrococcygeal region is the overall most common location. This article focuses on cervical and craniofacial teratomas.

Teratomas in the head and neck region account for approximately 2% to 5% of all germ cell neoplasms, with cervical teratomas being the most common. The incidence of cervical teratomas is estimated to be between 1:20,000 and 1:40,000 live births and are often identified on a prenatal ultrasound.[5] Cervical teratomas may extend into the mediastinum or displace the trachea, causing pulmonary hypoplasia.[1] As a result, there is an increased risk of respiratory-related morbidity and mortality and perinatal fetal interventions, such as an EXIT (ex utero intrapartum treatment) procedure may be required (refer to the article by Walz and Schroeder, elsewhere in this issue for further detail). If untreated, cervical teratomas have an estimated mortality rate of 80% to 100%.

Craniofacial teratomas have occurred in the orbit, pharynx, oropharynx, middle ear, sinonasal tract, and palate. A palatal teratoma is named an epignathus, and is the most common craniofacial teratoma in newborns (**Fig. 1**). These teratomas can present with fetal polyhydramnios due to impaired fetal swallowing and are often identified antenatally. Once identified, an EXIT procedure should be seriously considered. An epignathus is typically attached to the palate via a stalk. After initial resection and establishment of the airway, further imaging is necessary to determine persistence of teratoma within the infratemporal fossa and parapharynx, as demonstrated in **Figs. 1** and **3**.

Histology

Teratomas are composed of all 3 embryonic layers: endoderm, mesoderm, and ectoderm. They are further classified as being either mature or immature. Mature teratomas are the most common histologic type in children and contain only mature elements, such as skin, hair, fat, tissue, cartilage, bone, and glands.[1,6] Immature teratomas contain immature elements, such as neuroepithelial tissue and immature mesenchyme.[7] Microscopic foci of a yolk sac tumor, an immature element, have been found to be a predictor of recurrence after resection.[1] Congenital cervical teratomas have an estimated 5% risk of malignancy, with risk increasing with advanced age of diagnosis.[3,8]

Tumor Markers

Alpha-fetoprotein (AFP) is normally elevated in neonates and decreases in the postnatal period. Persistently high AFP may be suggestive of a teratoma. Additionally,