International Journal of Pediatric Otorhinolaryngology 97 (2017) 66–71

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: http://www.ijporlonline.com/

Teratoma of the neonatal head and neck: A 41-year experience

Jacob R. Brodsky, MD [a, b, *], Alexandria L. Irace, BA [a], Amanda Didas, PA-C [a], Karen Watters, MB BCh BAO, MPH [a, b], Judy A. Estroff, MD [c], Carol E. Barnewolt, MD [c], Antonio Perez-Atayde, MD [d], Reza Rahbar, DMD, MD, FACS [a, b]

[a] Department of Otolaryngology and Communication Enhancement, Boston Children's Hospital, United States
[b] Department of Otolaryngology, Harvard Medical School, United States
[c] Department of Radiology, Boston Children's Hospital, Harvard Medical School, United States
[d] Department of Pathology, Boston Children's Hospital, Harvard Medical School, United States

ARTICLE INFO

Article history:
Received 18 January 2017
Accepted 11 February 2017
Available online 14 February 2017
Presented at the European Society of Pediatric Otolaryngology (ESPO) Meeting, Dublin, Ireland; June 2014.

Keywords:
Teratoma
Germ cell tumor
Head and neck
EXIT

ABSTRACT

Objective: To review our institution's experience with the presentation, evaluation, and management of teratoma of the head and neck in the neonatal population.
Design: Retrospective case series (November 1970 through September 2011).
Setting: Tertiary care children's hospital.
Patients: 14 patients (12 boys and 2 girls).
Intervention: Detailed review of presentation, diagnostic approaches, surgical management, and outcomes.
Main outcome measures: Anatomic sites, use of pre and post-natal imaging, use of EXIT (ex utero intrapartum treatment) procedure, presenting symptoms, surgical approaches, additional therapeutic modalities, and outcomes are reviewed.
Results: Seven patients were diagnosed prenatally, while the remaining 7 patients were diagnosed at birth or shortly thereafter. The tumor emanated from the neck in 9 patients, the nasopharynx/oropharynx in 3 patients, the external nose in 1 patient and the face in 1 patient. Nine patients had associated upper airway obstruction. Four underwent an EXIT procedure, with 3 requiring intubation and 1 requiring tracheostomy. All patients underwent surgical resection. One patient demonstrated recurrence at follow-up.
Conclusions: Teratoma of the head and neck, though rare, is an important part of the differential diagnosis of neck masses in children, particularly in the perinatal period. The ability to make this diagnosis prenatally with high-resolution fetal ultrasound and MRI (magnetic resonance imaging) permits planning for airway and tumor management prior to delivery. An EXIT procedure should be considered when airway compromise by tumor compression is suspected. Early surgical excision is the treatment of choice and recurrence is rare when a complete resection is achieved.

1. Introduction

Teratoma is a rare tumor of the neonatal head and neck. Teratomas most commonly present in the saccrococcygeal region [1]. Although it is typically a benign congenital neoplasm, teratoma presenting in the neonatal period can be associated with a considerable risk of mortality, due to fetal hydrops and premature delivery resulting from the sheer volume of the tumor. Teratomas of the head and neck also carry a risk of partial or complete airway obstruction. Recent advances in prenatal imaging techniques provide a degree of detail adequate to not only diagnose potentially obstructing teratomas, but also to plan airway and tumor management strategies before the child is born. Furthermore, the advent of the *ex utero* intrapartum treatment (EXIT) procedure has allowed for the successful management of neonates with obstructive teratomas that would have been unlikely to survive without perinatal intervention.

This study describes a series of neonatal patients with teratoma

* Corresponding author. Department of Otolaryngology and Communication Enhancement, Boston Children's Hospital, 300 Longwood Avenue, LO-367, Boston, MA 02115, United States.
E-mail address: jacob.brodsky@childrens.harvard.edu (J.R. Brodsky).

http://dx.doi.org/10.1016/j.ijporl.2017.02.011
0165-5876/

of the head and neck seen at Boston Children's Hospital over a 41-year period. Presentation, imaging evaluations, and surgical management are reviewed. Prenatal diagnostic and management techniques are highlighted and examined.

2. Methods

Patients treated at Boston Children's Hospital for benign teratoma of the head and neck presenting in the perinatal period were included in this study. Any patients with malignant germ cell tumors or patients presenting beyond the first month of life were excluded. A total of 14 patients met this criteria and were included in the study. These patients' dates of initial presentation ranged from November 1970 to September 2011. The medical records of included patients were reviewed for demographic features, age at presentation, prenatal diagnosis, tumor site, airway management, presenting symptoms, preoperative/prenatal imaging, surgical management, adjuvant therapies, outcome, and duration of follow-up. Approval for this study was obtained from the Institutional Review Board at Boston Children's Hospital and Harvard Medical School.

3. Results

Fourteen patients met inclusion criteria. Clinical presentation and imaging evaluations are summarized in Table 1. Patients included 12 males (86%) and 2 females (14%). Seven patients (50%) presented prenatally, while the other half of patients presented at birth or shortly thereafter. Interventions and clinical outcomes are summarized in Table 2. Four patients (29%) underwent an EXIT procedure for surgical excision of the mass, with 3 requiring intubation and 1 requiring tracheostomy. All patients underwent surgical resection. Three patients (21%) required palatoplasty following excision of teratoma. Eleven patients (79%) presented with an immature teratoma, while 3 (21%) presented with a mature form. Follow-up imaging and clinic appointments were available to be reviewed for 10 patients that were managed post-operatively at our institution. The average length of follow-up was 58 months. One of these ten patients (10%) showed recurrent teratoma since their most recent excision.

4. Discussion

4.1. Epidemiology

Teratoma is the most common neonatal tumor (including all benign and malignant tumors), accounting for approximately 25% of tumors presenting in infancy [2], and it is the most common extragonadal germ cell tumor in children. The most frequent anatomic site is the sacrococcygeal region with an incidence of approximately 1 in 40,000 live births [3]. Head and neck teratomas are predominantly cervical in location and account for 2–9% of all congenital teratomas. The incidence has been reported to be greater in females with a male to female ratio of approximately 1:3 [1,2], though our series conversely shows a male predominance of 6:1.

4.2. Pathogenesis

The pathogenesis of teratoma is unclear and multiple theories have been proposed. In 1964, Batsakis et al. proposed that the lesions derive from a sequestration of pluripotent stem cells isolated during embryologic development [4]. Thirty years later, Kountakis et al. suggested that teratomas originate from foci of embryonic tissue that fail to migrate appropriately during development and escape the influence of their regional primary organizer [5]. They noted that the most common sites of teratomas within the head and neck can be predicted by the areas where all three germ cell layers closely approximate during embryological migration. Thus, the abnormal descent of Rathke's pouch endoderm and neuroectoderm with trapped elements of adjacent endoderm and mesoderm may form a nasopharyngeal teratoma. In addition, the abnormal descent of primitive thyroid tissue and trapped elements of adjacent ectoderm and mesoderm from the foramen cecum may form a cervical teratoma. Notably, all 9 of the cervical lesions in our series involved the thyroid gland.

The tendency of cervical teratomas to contain thyroid tissue or even replace part of the thyroid gland has led to much speculation about whether such tissue simply represents a well-differentiated component of the tumor or demonstrates that these lesions actually arise from the thyroid gland itself [6–8]. In 2005, Riedlinger et al. proposed that both situations exist, but that teratomas truly originating from the thyroid anlage may be over-reported in the literature, since many cases demonstrate a clear separation from the gland by a capsule or pseudocapsule on close histological examination [6].

4.3. Presentation

The presentation of teratoma of the head and neck varies by anatomic site and by whether the lesion is diagnosed in the pre- or post-natal period. The exact distribution of anatomic sites among

Table 1
Presentation of neonatal teratoma of the head & neck in 14 patients.

Patient	Sex	Year	Timing of Diagnosis	Tumor Site	Airway Obstruction	Thyroid Involvement	Pre-natal Imaging	Post-natal Imaging
1	M	1970	Birth	Right face	N	N	None	Skull X-ray
2	M	1979	Birth	Left maxilla, buccal mucosa, and left nasopharynx	N	N	Unknown	Unknown
3	M	1984	Prenatal	Left anterolateral neck	N	Y	US	None
4	F	1987	Birth	Right anterior neck mass	Y	Y	None	CT; Thyroid scan
5	M	1994	Prenatal	Nasal tip	N	N	US	CT
6	M	1996	Prenatal	Left neck	Y	Y	US; MRI	CT; Chest X-ray
7	M	2002	Birth	Nasopharynx/oropharynx	Y	N	None	MRI; CT
8	F	2002	Prenatal	Nasopharynx/oropharynx	Y	N	MRI; CT	None
9	M	2003	Prenatal	Right neck	Y	Y	US	MRI
10	M	2007	Prenatal	Left neck	Y	Y	US; MRI	CT
11	M	2007	Birth	Left anterior neck	Y	Y	None	US; CT
12	M	2010	Birth	Left anterior neck	Y	Y	US (normal)	MRI; US
13	M	2011	Birth	Right anterior neck	N	Y	None	US; MRI
14	M	2013	Prenatal	Left neck	Y	Y	US; MRI	None

Abbreviations: MRI, magnetic resonance imaging; CT, computed tomography; US, ultrasound.