Neonatal Airway Masses

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Conflict of Interest: none

Neonatal Airway Masses

Infectious Etiologies
- Laryngeal papillomatosis
- Infantile hemangioma
- Thyroglossal duct cyst
- Vallecular cyst
- Cervical teratoma
- HPV Immunization?

Vascular and lymphatic malformations
- Laryngeal cyst
- Infantile hemangioma
- Thyroglossal duct cyst
- Vallecular cyst
- Cervical teratoma

Cysts, teratomas
- Laryngeal cyst
- Infantile hemangioma
- Thyroglossal duct cyst
- Vallecular cyst
- Cervical teratoma

Neonatal Respiratory Papillomatosis

- Caused by Human Papilloma Virus
  - Most common STD in humans (1%)
  - Results in condyloma
- Recurrent exophytic lesions of the upper airway
- Hoarseness, progressive dyspnea
- Maternal transfer
- Juvenile-onset most commonly dx'd between 2-4 yrs, but can occur in neonates
- Highly variable clinical course, earlier presentation worse than later
- HPV Immunization?
**Infantile Hemangioma**

- Benign tumors of vascular endothelium
- Most common tumor of childhood (5% of newborns)
- Most common in:
  - Caucasians
  - Females: Males (6:1)
  - Premies
- 60% head and neck, 25% truncal, 15% extremities
- Natural history: rapid growth in 1st year followed by involution beginning at 18-24 months
- Typically benign course and self-limited nature
- Can result in ulceration, disfigurement, or critical dysfunction
- May herald significant abnormalities of spine, brain
- In this setting, focus on hemangioma of the airway

**Early Evolution of Infantile Hemangiomas**

**Vascular Tumors**

**Neonatal Hemangiomas**

- Congenital Hemangioma
  - Rare, benign, present and FULLY GROWN at birth
- Infantile Hemangioma
  - Common, benign, NOT present at birth, rapidly develop in neonatal period

**Natural History of Neonatal Hemangiomas**

- NICH: non-involuting congenital hemangiomas
- RICH: rapidly involuting congenital hemangiomas
- IH: infantile hemangiomas

**Neonatal Airway Masses**

CLASSIFICATION OF VASCULAR ANOMALIES

2014 INTERNATIONAL SOCIETY FOR THE STUDY OF VASCULAR ANOMALIES

- Vascular tumors
- Simple malformations
- Combined malformations
- Anomalies of major vessels
- Vascular malformations associated with other anomalies
- Infantile and congenital hemangiomas
- Cutaneous lymphatic, and venous malformations

**Neonatal Airway Masses**

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**Infantile and congenital hemangiomas**

- Capillary, lymphatic, and venous malformations

**Infantile and congenital hemangiomas**

- 2 or more histologies in the same lesion

**Coarctation**

- Sturge-Weber
- Klippel-Trenaunay

**Infantile and congenital hemangiomas**

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**Infantile and congenital hemangiomas**

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**Infantile and congenital hemangiomas**

- Early evolution of infantile hemangiomas
Natural Involution of Infantile Hemangiomas Over Time

Cutaneous hemangiomas and the airway
- Typically present cutaneously, but can present anywhere in tracheobronchial tree
- Subglottis most common
- At least 50% of pts with subglottic hemangiomas also have cutaneous lesions
- “Beard” distribution

Subglottic Hemangioma
Diagnosis:
- History
- Laryngoscopy/Bronchoscopy (posterior and lateral subglottis)
- X-ray (asymmetric narrowing of subglottis)
- CT Scan
  - Exclude mediastinal extension

Subglottic Hemangioma
- Direct laryngoscopy: compressible, asymmetric, bluish or reddish color
- Observation
- Steroids
- Interferon
- Laser
- Tracheostomy
- Propranolol

Subglottic Hemangioma

Intra-Op Subglottic Hemangioma
Studies starting in 2008 suggested oral propranolol effective 2015 study: Interim analysis of first 188 pts 60% vs 4% success 88% improved by week 5

Neonatal Propranolol for Hemangioma
- Hypoglycemia, hypotension, bradycardia, bronchospasm
- For infants < 2 months: EKG, ECHO, labs, and admitted for 2-3 d for observation
- For infants > 2 months: EKG, ECHO, labs, day hospital for 2-3 visits about 1 week apart
- Continuous telemetry, glucose checks
- Airway lesions typically advanced to 3 mg/kg/d

PHACES Syndrome
- Posterior fossa malformations
  - Dandy Walker variants
- Hemangioma
  - Large, facial lesions
- Arterial abnormalities
  - Cerebrovascular and carotid anomalies
- Cardiac abnormalities
  - Coarctation, arch anomalies
- Eye abnormalities
  - Microphthalmos, retinal vascular anomalies
- Sternal abnormalities

Neonatal Airway Masses

Lymphatic Malformation
- Dilated lymphatic channels filled with serous fluid
- Poorly defined, infiltrative
- Localized or diffuse
- Head and neck (48%), trunk and extremities (42%), intra-thoracic or intra-abdominal viscera (10%)
- Grow as child grows
- May enlarge rapidly due to hemorrhage, trauma, or infection
Lymphatic Malformation

- Males = Females
- 50-75% in posterior neck
- 60% diagnosed in utero by U/S
- Soft, compressible painless mass
- Dysphagia, airway compromise

- Macro-cystic, micro-cystic or combined
- Micro-cystic
  - < 2 cm in diameter
  - Ill-defined margins
  - Invasive
- Macro-cystic
  - >2cm in diameter
  - Well defined, encapsulated

- Supra- vs infra-hyoid
  - Suprahyoid difficult to manage
  - Often involves tongue base, pharynx, and parotid
  - Suprahyoid, microcystic disease remains a significant clinical challenge

Lymphatic Malformations

- Genetics
  - Fetal LM often have co-existing anomalies
  - 50-70% 45X
  - Autosomal trisomies
  - Associated syndromes:
    - Noonan
    - Roberts
    - Turner
    - Down
    - Hydrops fetalis
    - Fetal alcohol syndrome
- Fetal diagnosis (before 30 weeks) carries very poor prognosis
  - 10% survival
- Neonatal/Pediatric dx have very different natural history. Not usually associated with other anomalies
  - 2-6% mortality

Lymphatic Malformation

- Surgical excision is treatment of choice.
  However:
  - Difficult dissection
  - Complicated post op course, including seroma formation, infection, nerve damage, recurrence
  - 10% of cases extend into chest or mediastinum

Non-surgical treatment for Lymphatic Malformations

- Several therapies have been tried unsuccessfully including
  - Serial aspiration
  - Incision and drainage
  - Radiation therapy

- Sclerosing Agents
  - Alcohol
    - Rarely used today
    - Excessive toxicity
  - OK-432 (Picibanil)
    - Swelling, erythema, pain, fever
  - Doxycycline
    - Side effect: severe pain, swelling
    - May not be appropriate for retrobulbar or tongue masses
  - Bleomycin
    - Side effect: pulmonary fibrosis

Macro-cystic Lymphatic Malformation

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Macrocystic Lymphatic Malformation

Micro-cystic Lymphatic Malformation
Micro-cystic Lymphatic Malformation

Lymphatic Malformations of the Tongue

- Intra-oral LM's frequently involve tongue
- Both micro-cystic and macro-cystic types
- Can present dramatically
- Tend to effect anterior 2/3 of tongue
- May (or may not) obstruct airway

Neonatal Airway Masses

6 month old female

Stridor since birth

No definitive diagnosis in community hospital

Airway exam

Laryngeal Cyst

- Rare mass resulting in neonatal airway obstruction
- Presentation from Day 1 of life through 2 months of age
- Stridor, FTT, feeding difficulties, muffled cry, progressive cyanosis
Lingual Thyroglossal Duct Cyst

- Uncommon etiology of airway obstruction and stridor in newborns
- Several case reports of SIDS-like death in newborns retrospectively caused by LTGDC
- Midline thick-walled submucosal cyst at base of tongue

Vallecular Cyst

- Typically present in neonatal period
- Muffled cry
- Feeding difficulty, FTT
- Cyanosis with feeding
- Apnea
- Easily confused with laryngomalacia
- Marsupialization

Teratoma

- Majority are benign
- Unclear natural history
  - One series (J Peds Surg 2004) demonstrated mets to regional lymph nodes in 14% of cases
  - Potential for malignant transformation?
  - Many remain tumor free long-term despite these mets
  - Possible role for alpha-feto-protein monitoring

Lingual Thyroglossal Duct Cyst

- Neonatal presentation associated with significant mortality
- Presentation may be later in life, or as an “incidental” finding in older children
- Differential:
  - Vallecular cyst
  - Ectopic lingual thyroid

Teratoma

- Germ cell tumors composed of tissue foreign to that anatomic location
  - All 3 germ layers
  - Most commonly sacrococcygeal, but can occur in the neck and other areas including ovaries, testes, mediastinum
- <10% of teratomas are found in the neck
- Cervical teratomas typically bulky, extending from the mandible to the clavicle
  - Mandibular hypoplasia
  - Polyhydramnios due to esophageal compression (40%)

Cervical Teratoma

Lymphatic malformation
Epignathus: neonatal teratoma arising from oropharyngeal cavity

Conjoined Twins/Epignathus
- Conjoined “parasitic” twin
- Separated at 72 hours
- Heart failure at 3 weeks due to Pentalogy of Cantrell
- Expired

Cincinnati airway anomalies resulting in fetal intervention

Cincinnati airway anomalies resulting in Fetal Intervention

Fetal Intervention: Agnathia
Fetal Intervention: Micrognathia

CHAOS
Congenital High Airway Obstruction Syndrome

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Lymphatic malformation

Vascular and lymphatic malformations
Laryngeal cyst
Thyroglossal duct cyst
Vascular malformation

Cysts, teratomas
Vallecular cyst
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