OBJECTIVES

1. Outline modalities used for imaging head and neck tumors in children

2. Discuss fetal appearance of head and neck tumors

3. Highlight features of common head and neck tumors in children

4. Introduce a few rare but unique pediatric head and neck neoplasms
MODALITIES

• US
  - Usually initial modality
  - Quick, cheap, easily tolerated
  - Differentiates cystic from solid masses
  - Vascularity

• CT
  - Bony Details
  - Calcification
  - Ionizing Radiation
MODALITIES

• MRI
  ➢ Excellent soft tissue contrast
  ➢ Better characterization of masses
  ➢ Marrow involvement
  ➢ Diffusion
    ▪ Cellularity & Nuclear-cytoplasmic ratio of tumors
    ▪ ADC values help differentiate benign and malignant tumors in children \((1.3 \times 10^{-3} \text{ mm}^2/\text{sec})\)
    ▪ ADC values predict outcomes in adult H&N SCC and response to chemoradiation based on early change in ADC
    ▪ ADC of less than \(1.3 \times 10^{-3} \text{ mm}^2/\text{sec}\) (some authors use \(1 \times 10^{-3} \text{ mm}^2/\text{sec}\)) on follow up scans of adult H&N SCC suggest recurrence
    ▪ Distortion from air interfaces; better to use non-EPI sequences or multi-shot technique with readout-segmented EPI sequence
  ➢ Perfusion
Choice of Diffusion Sequence

Multishot with readout segmented EPI (RESOLVE, Siemens)

EPI Diffusion
Embryonal Rhabdomyosarcoma before and after chemoradiation
MODALITIES

- PET
  - Metabolic response earlier than anatomic response
  - Differentiates active disease from treated inactive tumor in residual soft tissue masses

LCH at Presentation

3 month follow up
MODALITIES

• PET
  - Metabolic response earlier than anatomic response
  - Differentiates active disease from treated inactive tumor in residual soft tissue masses
  - Helps distinguish reactive from malignant nodes
Which one is malignant?

11 yo Alveolar Rhabdomyosarcoma

4 yo Embryonal Rhabdomyosarcoma
MODALITIES

• PET
  - Metabolic response earlier than anatomic response
  - Differentiates active disease from treated inactive tumor in residual soft tissue masses
  - Helps distinguish reactive from malignant nodes
  - PET/MR as a combined study
MODALITIES

• PET
  - Metabolic response earlier than anatomic response
  - Differentiates active disease from treated inactive tumor in residual soft tissue masses
  - Helps distinguish reactive from malignant nodes
  - PET/MR as a combined study
  - Brown fat: winter; more common in children; ↓ sensitivity to residual neck disease

10 yo Hodgkin Disease
MODALITIES

• Angiography

  ➢ Preoperative embolization of feeding vessels significantly reduces bleeding during surgery and allows a more complete tumor resection.
TERATOMA

• Tissues of all three germ cell layers

• Second most common location for teratoma in early infancy

• Mature, immature, and teratoma with malignant transformation
  ➢ Histological immaturity in congenital teratoma does not imply an adverse outcome (different from adolescents and adults)
TERATOMA

• Heterogeneous with solid and cystic components
  ➢ Calcifications in 50%
  ➢ Sometimes fatty tissue

• Compression of the airway may necessitate EXIT (Ex Utero Intrapartum Treatment) procedure
  ➢ C Section and establishing an airway (Intubation or tracheostomy) while maintaining placental support
  ➢ Around 60 minutes
4 week old Immature Teratoma
LYMPHATIC MALFORMATION

- Trans-spatial multilocular cystic mass
- Fluid-Fluid level
- Airway compression may necessitate EXIT procedure
EPIGNATHUS

• Teratoma involving the oral cavity and oropharynx

• Arise from the craniopharyngeal canal
MATURE TERATOMA (EPIGNATHUS)

Predominantly fatty

Predominantly cystic
EPIGNATHUS

- Association with duplication of the pituitary gland, intracranial malformations, and midline facial defects
EPULIS

• Congenital benign granular cell tumor

• Homogeneous mass involving the anterior alveolar ridge

• More posterior extension suggestive of epignathus
HAIRY POLYP

- Rare fat-containing benign tumor of neonates and infants

- Bi-germinal layer origin characterized by ectoderm covering a mesodermal core of adipocytes, smooth muscle and cartilage

- No endoderm (differentiates it from mature teratoma)

- Most common in the nasopharynx but can also occur in the palate, tongue, tonsils, and eustachean tube/middle ear cavity

HEMANGIOMA

• Most common benign tumor in the head and neck

• Endothelial proliferation, hence a neoplasm

• Treatment: Expectant. If symptomatic: Steroids, alpha interferon and propranolol
**HEMANGIOMA**

<table>
<thead>
<tr>
<th>Infantile Hemangioma</th>
<th>Congenital Hemangioma</th>
</tr>
</thead>
<tbody>
<tr>
<td>More common</td>
<td>Rare</td>
</tr>
<tr>
<td>Present at birth in 20-30% of cases</td>
<td>Always present at birth</td>
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<tr>
<td>Proliferate in the first year of life then involute over several years</td>
<td>Proliferate in utero</td>
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<tr>
<td>GLUT-1 positive</td>
<td>GLUT-1 negative</td>
</tr>
<tr>
<td><strong>PHACES</strong> syndrome</td>
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<tr>
<td>Posterior Fossa malformations</td>
<td>Rapidly Involuting Congenital Hemangioma (RICH): usually disappear by 14 months</td>
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<tr>
<td>Hemangiomas</td>
<td>Non Involuting Congenital Hemangioma (NICH)</td>
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<tr>
<td>Arterial anomalies/Aortic Coarctation</td>
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<tr>
<td>Eye Anomalies</td>
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<td>Sternal defects/Supraumbilical raphe</td>
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Proliferating Infantile Hemangioma

Prominent vascularity
Hyperintense on T2
Avid enhancement

11 week old female with parotid infantile hemangioma
INVOLUTING INFANTILE HEMANGIOMA

Decrease in size
Increased fatty component
Diminished vascularity and enhancement
SUBGLOTTIC HEMANGIOMA
22 weeks Gestation & Neonatal Imaging. Congenital Hemangioma
PHACES

Post Gad T1

Post Gad SPGR

ASL Perfusion

Fat Sat Post Gad T1

ASL Perfusion

TOF MRA

T2
TUFTED ANGIOMA & Kaposiform Hemangioendothelioma

- Kaposiform Hemangioendothelioma
  - Aggressive vascular tumor of infancy of intermediate malignancy
  - Ill-defined mass with stranding in the subcutaneous fat, calcifications, prominent feeding and draining vessels, and bone destruction

- Tufted Angioma: More superficial

- Associated with consumption coagulopathy (Kasabach-Merritt)
LYMPHOMA

• Most common head and neck malignancy in children

• Hodgkin lymphoma (HL): more common; early adolescence

• Non-Hodgkin lymphoma (NHL): throughout childhood

• Necrosis and cystic changes are rare

• FDG PET has higher sensitivity and specificity
HODGKIN LYMPHOMA

- Unilateral adenopathy
- 80% of patients with cervical HL have disease outside H&N
- Prior EBV infection has been implicated
NON-HODGKIN LYMPHOMA

• 70% of NHL have disease outside of H&N
• Extranodal NHL involves Waldeyer ring, sinonasal region, thyroid, or orbit
• Common histological subtypes in children
  ➢ Burkitt lymphoma
  ➢ Lymphoblastic lymphoma
  ➢ Diffuse large B-cell lymphoma
  ➢ Anaplastic large cell lymphoma

3 yo Lymphoblastic lymphoma
NASOPHARYNGEAL CARCINOMA (NPC)

• Most common carcinoma in H&N alongside papillary thyroid cancer

• Adolescents

• Related to EBV

• Presentation
  ➢ Nasopharyngeal mass
  ➢ Lymphadenopathy
  ➢ Unilateral painful otalgia and mastoid effusion
  ➢ Rhinorrhea and nasal obstruction
Two different 15 year olds. NPC
• Often metastatic from adrenal primary (lymph nodes and osseous)

• Primary cervical location in 5% of cases arising in the sympathetic chain and causing Horner syndrome.

• Mass with restricted diffusion, calcifications, cysts, and dumbbell masses with intraspinal component

• Iodine-123 MIBG scintigraphy
NEUROBLASTOMA

7 month old Poorly differentiated neuroblastoma
JUVENILE NASOPHARYNGEAL ANGIOFIBROMA (JNA)

- Highly vascular locally aggressive neoplasm
- Adolescent males
- Epistaxis and nasal obstruction
- Arises along the posterolateral wall of the nasal cavity at the sphenopalatine foramen and grows laterally into the pterygopalatine fossa
JUVENILE NASOPHARYNGEAL ANGIOFIBROMA (JNA)

- Erosion and bowing of the bony structures
  - Characteristic anterior bowing of the posterior maxillary sinus wall

- Intraorbital and intracranial extension

- Variable signal intensity on T2
  - Often hypointense because of fibrous tissue

- Prominent vascularity with flow voids

- Intense enhancement
JUVENILE NASOPHARYNGEAL ANGIOFIBROMA (JNA)

- Angiography: Intense tumor blush with enlarged arterial supply via branches of the internal maxillary and ascending pharyngeal arteries
LANGERHANS’ CELL HISTIOCYTOSIS (LCH)

• Abnormal clonal proliferation of highly differentiated histiocytes (Langerhans cells)

• Unclear whether process is inflammatory or neoplastic

• Lymphadenopathy (most often cervical) usually part of multisystem LCH

• Areas of necrosis simulating infection
LANGERHANS’ CELL HISTIOCYTOSIS (LCH)
NERVE SHEATH TUMOR

• Solitary and plexiform neurofibroma & malignant peripheral nerve sheath tumor (MPNST) in NF1. Schwannomas with NF2

• Solitary neurofibroma and schwannoma are sharply demarcated and have variable signal
  ➢ Fibrous tissue in neurofibromas and Antoni A tissue in schwannomas → Dark T2 signal (simulates high nuclear-to-cytoplasmic ratio or highly cellular neoplasms)

• Homogeneous enhancement with cystic foci in schwannomas
Capsulated Neurofibroma in NF1

15 yo Cellular Trigeminal Schwannoma

Schwannoma in NF2
NERVE SHEATH TUMOR

- Plexiform neurofibromas
  - unencapsulated and grow along peripheral nerves
  - Multispatial
  - Hypodense on CT and hyperintense on T2

- MPNST
  - rapid increase in size
  - alteration in enhancement characteristics
  - Metastatic disease
  - PET with SUVmax above 4
9 month old. Parotid Plexiform Neurofibroma. Subsequently diagnosed with NF1
RHABDOMYOSARCOMA

• Most common soft tissue sarcoma in children (35-50% in head and neck)

• 20% of malignant head and neck tumors
  ➢ Second most frequent head and neck malignancy after lymphoma

• Bimodal distribution
  ➢ Peak in first decade around 7 years and second peak in adolescence

• Head & Neck RMS most commonly in the masticator space and orbits
RHABDOMYOSARCOMA

• Develop from undifferentiated primitive mesenchymal cells which have the capacity to differentiate into striated muscles

• Histological classification
  ➢ Embryonal (most common)
  ➢ Alveolar (older children; worse prognosis)
  ➢ Pleomorphic

• Metastasis
  ➢ Cervical lymph nodes
  ➢ Hematogenous to bone and lungs
RHABDOMYOSARCOMA

• Associations
  - NF1 (early and more aggressive tumors than non-NF1 RMS)
  - Li–Fraumeni syndrome
  - p53 tumor suppressor gene mutations

• Up to 75% of alveolar RMS have FOX01 to PAX3 or PAX7 gene fusion
  - Alveolar RMS lacking this translocation behave similar to embryonal RMS
RHABDOMYOSARCOMA

• Signs and symptoms depend on location
  ➢ Neck mass
  ➢ Nasal obstruction with epistaxis
  ➢ Proptosis
  ➢ Airway obstruction
  ➢ Otalgia from ipsilateral eustachian tube obstruction
  ➢ Cranial Neuropathies
RHABDOMYOSARCOMA

• CT
  - No calcification
  - Attenuation similar or slightly higher than muscles
  - Lytic bone destruction but can show bony remodeling
RHABDOMYOSARCOMA

• MRI
  - Isointense signal to muscle on T1w
  - Variable signal compared with muscle on T2w. Most are iso- to hypointense
  - Variable degrees of enhancement
  - Intracranial spread carries a worse prognosis
    - Anatomic sites with this potential (Parameningeal sites) are orbit, masticator space, nasopharynx, sinonasal, and middle ear
    - best seen on coronal fat suppressed post gad T1w
RHABDOMYOSARCOMA

- Characteristically have restricted diffusion

- Orbital lesions can be well circumscribed and cause bony remodeling rather than aggressive destruction
ALVEOLAR RHABDOMYOSARCOMA AND CHEMORADIATION

Before chemoradiation

After chemoradiation
• Patients with residual lesions after completing treatment were at risk of recurrence compared to patients without residual lesions on cross-sectional imaging


• Presence or absence of a residual lesion after completion of therapy did not have impact on prognosis

PRIMITIVE NEUROECTODERMAL TUMOR/EWING SARCOMA

• Peripheral primitive neuroectodermal tumors (pPNETs): Malignant tumors of neuroepithelial origin with small blue round cells that arise outside the central nervous system

• pPNET and extraosseous Ewing sarcoma are opposite ends of the same spectrum (undifferentiated cells in Ewing sarcoma and neuronal differentiation in pPNET)
  - Similar histopathologic features
  - Same chromosomal rearrangement with translocation t(11;22) (q24;q12)

• H&N location is rare (5-10% of all pPNET/Ewing)
PILOMATRICOMA

- Benign subcutaneous tumor that arises from hair cells
- Most common calcified tumor in the head and neck
- Isointense to muscle on T1 & T2
- Internal reticulations and patchy areas on T2 and post Gad
PILOMATRICOMA
LIPOBLASTOMA

• Benign mesenchymal tumor composed of immature adipose cells resembling fetal fat (lipoblasts) & mature adipocytes in addition to a plexiform vascular pattern and variable myxoid stroma

• Painless masses in young children (< 5 years)

• Majority are in the extremities but can occur in the head and neck

• Characteristic cytogenetic 8q11-13 clonal (PLAG1) chromosomal rearrangement
LIPOBLASTOMA

- Classified as focal (lipoblastoma) or diffusely infiltrative (lipoblastomatosis) forms

- Imaging features depend on the abundance of macroscopic fat, fibrous septa, cystic changes, & myxoid components

- CT: Fatty density mass

- MRI:
  - Fat signal
  - Internal fibrous septae: Dark on T1 & T2w
  - Enhancing solid components: Reflects the varying amount of myxoid mesenchymal components
SIALOBLASTOMA

• Very rare neoplasm that arises from the primitive duct epithelium of salivary glands

• First 2 years of life

• Majority in the parotid

• Local invasion and recurrence

• Distant metastasis is rare
SIALOBLASTOMA

21 months
MYOFIBROMA

• Benign fibrous mesenchymal neoplasms composed of myofibroblastic connective tissue cells

• Solitary (myofibroma) and multicentric (myofibromatosis)

• Commonest in infancy

• May spontaneously regress
SOLITARY MYOFIBROMA

- Usually homogeneous mass

- Can be associated with calcifications and necrosis

- Signal intensity varies from hypointensity to hyperintensity on T1 & T2 and variable enhancement

- Local bone destruction mimicking malignant lesion
DESMOID FIBROMATOSIS

• Also known as Aggressive fibromatosis

• Associated with Gardner syndrome

• Unencapsulated and similar to adult desmoids

• In the long bones or mandible → Desmoplastic fibroma
DESMOPLASTIC FIBROMA

• Tends to be aggressive in the mandible

• Rapidly expanding lesion typically involving the ramus and angle, although the maxilla might also be affected

• Although they lack a capsule, margins tend to be well defined

• High recurrence rate
DESMOPLASTIC FIBROMA

- Cortical breakthrough with a soft-tissue component
- Low attenuation on CT
- T2 signal is usually high due to myxoid changes and decreased collagen content
10 month old girl

T1
Fat Sat T2
Fat Sat T2
ADC
Early Fat Sat Post Gad
Delayed Fat Sat Post Gad
DIFFERENTIAL DIAGNOSIS

Destructive Sinonasal mass
1. Rhabdomyosarcoma
2. Other sarcomas (Particularly pNET/Ewing)
3. Lymphoma
4. Leukemia

Nasopharyngeal Mass
1. Nasopharyngeal carcinoma (Teenagers)
2. Rhabdomyosarcoma
3. Lymphoma
DIFFERENTIAL DIAGNOSIS

Masticator Space/Parapharyngeal Mass

1. Rhabdomyosarcoma
2. Other sarcomas (Particularly pNET/Ewing)
3. Lymphoma
4. Neuroblastoma
5. Fibrous Tumors in infants