Pediatric head and neck masses

Eileen Raynor, MD FACS FAAP
Division pediatric otolaryngology

No disclosures

Initial approach

<table>
<thead>
<tr>
<th>Congenital</th>
<th>Acquired</th>
</tr>
</thead>
<tbody>
<tr>
<td>Location</td>
<td>Location</td>
</tr>
<tr>
<td>Midline</td>
<td>Lateral</td>
</tr>
<tr>
<td>Lateral neck</td>
<td>Bilateral</td>
</tr>
<tr>
<td>Associated factors</td>
<td>Associated symptoms</td>
</tr>
<tr>
<td>Skin changes</td>
<td>Fever/pain</td>
</tr>
<tr>
<td>Change with infection (URI)</td>
<td>IL symptoms</td>
</tr>
<tr>
<td>Drainage/sinus tract</td>
<td>Dysphagia</td>
</tr>
<tr>
<td>Timing of intervention</td>
<td>Change over time</td>
</tr>
<tr>
<td>Imaging</td>
<td>Fluctuation in size or swelling</td>
</tr>
<tr>
<td>Surgery or other</td>
<td>Change in texture</td>
</tr>
</tbody>
</table>

Differential diagnosis

Benign processes more common in pediatric patients
Congenital or infectious etiologies are most likely
Congenital masses may not present until teens or early 20’s
Family history may be useful in these cases
Other conditions: ie. Hearing loss

Congenital neck masses

- Branchial cleft fistula/cyst
- Thyroglossal duct cyst
- Dermoid
- Midline cervical cleft
- Vascular malformations
  - Hemangioma
  - Lymphatic malformation

Evaluation and workup

Imaging
- Ultrasound beneficial for midline or cystic lesions
- CT with contrast
- MRI

Tissue sampling
- FNA usually most beneficial (unless pulsatile - ie. AVM)
- Excisional biopsy in specific cases
Congenital masses: midline

Thyroglossal duct cysts most common – US can often differentiate between TGD and dermoids

Dermoids also present in the midline from the frontal process to the sternal notch

Midline cervical clefts may present as a mass, draining fistula or thick tract

Thyroglossal duct cysts

• Occur as thyroid descends from foramen cecum in tongue down to 1st tracheal ring
• Usually located between cricoid and hyoid – suprathyroid can occur, hard to identify tract
• Terminate in the base of tongue (foramen cecum)
• Sistrunk procedure is surgical treatment – 5-20% recurrence rate reported (60% if midportion of hyoid not resected)
• Ultrasound to determine presence of normal thyroid


Thyroglossal duct cyst

• Usually has solid and cystic components on ultrasound
• May present with a small puncta or hair protruding through skin
• Can be difficult to differentiate intraoperatively from TGD cyst but will not find a tract leading toward hyoid
• Comprised of epidermis and adnexal structures (hair, sebaceous glands)

Dermoid cyst

• Rare lesion present at birth
• Can cause restriction of neck and mandibular growth
• Failure of fusion between 1st and 2nd arches
• Excision via double Z-plasty

Midline cervical cleft

Congenital masses: lateral neck

Lateral neck masses may initially present as pits or fistula tracts

Most common are second and third branchial cleft cysts

Location is anterior to the sternocleidomastoid and they terminate in the pharynx (tonsil or pyriform)

Hemangiomas/lymphatic malformations often present in anterior triangle but can occur anywhere in the neck
Branchial cleft cysts

- Derived from pharyngeal pouches
- First arch: ear and external canal
  - Reduplicated EAC (type 1)
  - May follow VII into parotid, end below angle of mandible (type 2)
- Second: goes between the ICA and ECA
  - Above IX and XII, enters tonsil fossa
- Third: behind ICA and ECA
  - Between IX and XII into pyriform

- Congenital masses: branchial cleft

  Fourth branchial arch anomaly
  - Opening into apex of pyriform
  - Runs between superior and recurrent laryngeal nerve
  - Opens into lower aspect of neck
  - Intimately involved with the thyroid gland

- Derivatives of Pharyngeal Arches & Pouches

- Second Branchial Cleft
  Cyst and Stem Tract

- Third Branchial Cleft Cyst
  and Stem Tract

- Pathway of a Fourth Branchial Pouch Abnormality

- Tract emerges from pyriform
  Loops around the aortic arch following RLN
  Generally cyst involves thyroid lobe
  Thyroid lobectomy should be included in resection
Vascular malformations

- May occur anywhere in the head and neck
- Can be unilateral and localized or bilateral and diffuse
- Characterized by histologic type and growth pattern
  - Hemangiomas
  - Lymphatic or venolymphatic malformations
- May be present at birth or present during infancy/childhood

Hemangiomas

- May be present at birth but usually present several weeks to months after birth
- Congenital type can be rapidly involuting (glut1 +) or non-involuting
- Majority are infantile type
- Usually do go through a proliferative phase followed by a slow involution
- May need medical intervention depending on location and size
- Imaging not always indicated but MRI with contrast would be study of choice

Subcutaneous hemangioma

- Often referred to as cystic hygromas
- May be present at birth
- Grow with the child and enlarge during concomitant illness
- Treated by surgical excision or sclerotherapy, serolimus and sildenafil both helpful adjuncts
- Can be macrocystic or microcystic or combination
- Will often change during puberty
Acquired neck masses

Lymphadenopathy
- infectious
- malignant
Thyroid
- benign
- malignant
Rhabdomyosarcoma
- Other malignancies

Cervical adenopathy
- Lymph nodes typically palpable in young children
- Often result of viral illness due to developing immune system
- May stay enlarged for several months
- Normal architecture on ultrasound

Infectious etiology: cervical adenitis
- May occur at any age
- Often associated with systemic symptoms: fever, malaise, arthralgia
- May have an overlying cellulitis and multiple sites of adenopathy
- May be self-limiting (viral adenitis) or may require antibiotics
- Can abscess and require IV antibiotics or surgical drainage

History is key
- insect bites (tick, spider, mosquito)
- cat exposure
- tuberculosis
- strep throat
- mononucleosis
- HIV exposure
Infectious etiology

Tuberculosis (scrofula)
- Necrotic adenopathy
- In children: 92% are atypical mycobacterium (MAC, MAI) with the oral cavity as the point of entry
- 95% in adults are mycobacterium tuberculosis
- Stain + for AFB
- PPD may be negative, quantiferon may be equivocal
- Surgery may still be indicated but recently antiTB therapy is mainstay of treatment if TB
- Needle aspiration or excision for atypical TB


Cat scratch disease
- Occurs most often in < 21 yo
- 22,000 cases in Us annually
- Bartonella Henselae
- Regional cervical lymphadenitis
- May cause hepatitis, retinitis, encephalitis or osteomyelitis
- Parinaud’s oculoglandular syndrome – conjunctivitis with inoculation site in eye
- Usually self-limiting, can be treated with fluoroquinolones or TMP-SMX


Toxoplasmosis
- Toxoplasma gondii
- Infects up to 60 million people in US
- Most are asymptomatic
- Found in cat faces and undercooked meat (pork most commonly)
- Flu like symptoms and generalized lymphadenopathy
- Severe cases (immunocompromised) may affect the eyes or cause encephalitis
- Pregnant women who become infected may pass it to the fetus (TORCH syndrome)
- Treatment in symptomatic patients includes pyrithiamine and a sulfonamide

Other neck pathology

Lipoma
Epidermal inclusion cysts
Sarcoidosis
Castleman’s disease – cervical and mediastinal lymphadenopathy
Neurofibromata – NF1
Post transplant lymphoproliferative disease
Langerhans cell histiocytosis

Benign thyroid lesions

- Colloid nodule
- Degenerative or hemorrhagic nodule
- Non functional goiter
- Follicular adenoma

Malignancies

Lymphoma
- Sixth most common cause of cancer
- Non-Hodgkin more common (T cell, B cell or NK cell types)
- 64,000 new cases diagnosed in 2005 (7400 cases are Hodgkin’s)
- Incidence doubled over past 35 years
- Associated with Epstein-Barr virus and Helicobacter pylori
- FNA can assist in diagnosis but may need lymph node excisional biopsy
- Chemotherapy +/- XRT

Malignancies

Hodgkin’s Lymphoma

Characterized by Reed-Sternberg cells
Spreads by nodal regions then to solid organs (liver, lung, bone marrow)
Four patterns found:
- Lymphocyte dominant
- Nodular sclerosing
- Mixed cellularity
- Lymphocyte deletion

Thyroid malignancies

- Follicular carcinoma: Diagnosed by vascular and capsular invasion. Less common in children and rare association with radiation exposure. 5-20% recurrence. Distant mets due to vascular spread.
- Papillary thyroid carcinoma: Most common type. 90-95% survival, approaches 100% in children. Usually multinodular, multicystic. Associated with autoimmune thyroid disease and prior radiation exposure. Up to 50% have regional lymph node spread.
- Medullary carcinoma: Arise from parafollicular C cells, associated with MEN syndromes. Elevated calcitonin and CEA levels helpful with diagnosis. Overall survival 75% for 10 years. Reported in children age 6 and up, usually presents late 20s-40s.

Papillary carcinoma

- Long strands or papillae of cells
- Little colloid
- Cellular atypia
- Multiple cystic spaces

Follicular carcinoma

- Atypical follicles with cellular infiltrate
- Vascular or capsule invasion diagnostic for carcinoma

metastatic nodes from papillary carcinoma of the thyroid
Medullary thyroid carcinoma

- Parafollicular C cells
- Look like "signet rings"
- Secrete calcitonin
- May produce watery diarrhea and carcinoid type symptoms

Special considerations

- MEN syndromes
  - MEN 1: parathyroid adenoma, pancreatic islet cell tumors, pituitary adenoma
  - MEN 2a: pheochromocytoma, hyperparathyroid, medullary carcinoma
  - MEN 2b: neurofibromas, medullary carcinoma, pheochromocytoma, marfinoid
- Familial medullary thyroid carcinoma (FMTC)

Disorders of the RET gene which regulates the TGF-β signaling system

Rhabdomyosarcoma

- Most common childhood soft tissue malignancy, 3-4% of all pediatric cancers
- Commonly presents in the maxilla or nasopharynx – if arise in orbit they are always embryonal type
- Histologic types
  - Embryonal – arranged in sheets and nests (most H&N)
  - Botryoid and Spindle cell – grape like (GU tract)
  - Alveolar (FOX01 rearrangement) – fibrovascular septae (extremities)

Rhabdomyosarcoma

- 7-8% associated with genetic syndromes – include Beckwith Weidemann (IGF-2 gene) and NF1 (20X increase risk)
- MRI and CT for staging – uncertain if PET useful
- LN biopsy for metastatic evaluation – less common in H&N primary
- Prognosis dependent on TNM stage, histologic type, genetic factors (presence of FOX01)

Prognosis of RMS

| Prognosis of rhabdomyosarcoma according to site of primary tumor from two different clinical trials |
|---|---|---|
| Site of primary tumor | IRS II | IRS III |
| Head and neck | 97 | 88 |
| Extremity | 94 | 91 |
| Other sites | 77 | 88 |

Consult triage

Urgent/inpt ENT evaluation
- Neck masses causing airway obstruction or failure to thrive
- Acute infection not responding to Rx
- Rapidly enlarging mass

Outpatient management
- Neck cysts (not causing airway obstruction)
- Draining pits/sinus tracts
- Hemangiomas
- Lymphatic-vascular malformation
- Lymphadenopathy
Final thoughts

<table>
<thead>
<tr>
<th>Congenital</th>
<th>Acquired</th>
</tr>
</thead>
<tbody>
<tr>
<td>Location</td>
<td>Location</td>
</tr>
<tr>
<td>Associated factors</td>
<td>Associated symptoms</td>
</tr>
<tr>
<td>Timing of intervention</td>
<td>Change over time</td>
</tr>
</tbody>
</table>