Case Presentation

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Division of Pediatric Surgery
July 7th 2006
Case Presentation

• x year old African American male seen in Pediatric Surgery Clinic
  – History of midline neck mass since 2 years of age
  – Denied history of swelling, drainage or erythema
  – Mild enlargement of mass with age

• History:
  – Chronic Otitis Media: B/L myringotomy & drainage
  – Asthma

• NKDA
Case Presentation

• Physical Examination:
  – 2x1 cm midline solid mass
    • Not mobile
    • Overlying the hyoid bone
  – Mass moved cephalad with protrusion of tongue & swallowing
  – No evidence of sinus tract or external opening
  – Preoperative Labs:
    • WNL
Case Presentation

• Sonogram:
  – 1.2 x 0.9 cm midline cystic structure
  – Superior to and separate from thyroid
  – Thyroid: WNL
    • Impression:
      – Midline neck cyst most consistent with thyroglossal duct cyst.

• Surgical Excision:
  – Excision of Thyroglossal Duct Cyst: “Sistrunk Procedure”
Case Presentation

OPERATIVE TECHNIQUE

Thyroglossal Duct Cyst
Line of incision through skin over cyst
Figure 3  After splitting strap muscles down the middle, the cyst can be grabbed and lifted up as it is dissected free from the surrounding tissues.
**Figure 4**  Once freed up inferiorly, the cyst can be seen pedicled to the hyoid bone superiorly. Also notice the close relationship between the cyst and the underlying airway.

**Figure 6**  The tract can be followed to the foramen cecum and excised.
Resecting middle part of hyoid bone with cyst
Cyst & bone segment isolated on duct
**Figure 5**  The hyoid bone is then cut on each side of the midline.

**Figure 7**  The opening at the foramen cecum is then closed.
Case Presentation

Figure 8   The strap muscles are reapproximated and the skin then closed.
Case Presentation

• Pathology:
  – Thyroglossal Duct Cyst

• Follow Up:
  – POD 7: No complaints
  – POD 21: No complaints
Evaluation of Pediatric Neck Masses

ALIREZA SADEGHI MD
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July 7th 2006
Introduction

• The evaluation and management of pediatric neck masses requires extensive knowledge
  – Anatomy and embryology of the head & neck
• Congenital masses, inflammatory & infectious lesions, benign neoplasms & vascular malformations are more common than malignant neck lesions
• A thorough history and physical examination along with a focused and coordinated work up are essential in the management of pediatric neck masses
### Differential Diagnosis of Neck Masses in Children

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## Differential Diagnosis of Neck Masses in Children.

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<td>Sialadenitis</td>
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Pediatric Neck Masses

• Initial Evaluation
  – History & Physical Examination

• Site & Duration of symptoms:
  – Systemic Signs Illness: Fever, Weight loss, Night sweats
  – Associated illness: Otitis media, Streptococcal pharyngitis
  – Short vs. long duration (Recurring?)
  – Presence at birth? Mostly Benign & Congenital

• Distribution: Benign vs. Malignant
  – 80 % vs. 20%

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Pediatric Neck Masses

• Radiographic Studies
  – USG: Distinguishes Solid vs. Cystic & Evaluate Thyroid
    • Also Doppler can distinguish vascularity of the mass
  – CT Scan: More useful if a tumor is diagnosed or suspected
    • Delineates the depth, anatomic details and thoracic extension
  – MRI: 3D anatomic detail especially in spinal cord or intracranial extension
    • T2 identifies areas of high vascularity
    • T1 enhanced with gadolinium localizes CNS tumors
  – PET: Maps location & concentration of radionuclide-labeled tracer. Metabolic activity.

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Pediatric Neck Masses

• Fine-Needle Aspiration
  – Performed in office setting under local anesthesia
  – Lower cost than open biopsy & Dx within 24 hrs
  – No scarring & reassuring to parents/pediatricians
    • A lymph node > 1 cm persisting or growing for 6 weeks despite of antibiotic therapy

• Open Biopsy
  – Sampling error on FNA
    • LN or neck mass continues to enlarge over 2-3 weeks or persists in size for 3 months after FNA

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Work up of Pediatric Neck Mass

[Diagram of workup process]

**FIGURE 56-5.** Algorithm for the evaluation and treatment approach to neck masses in children. FNA, fine-needle aspiration; AMB, atypical mycobacteria; TB, tuberculosis; CSD, cat-scratch disease.

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Introduction

• Definition:
  – Cyst
    • A mucosal or epidermal lined structure without any external opening
  – Sinus
    • A tract, with or without a cyst, that communicates to either the skin (External) or the mucosa (Internal)
  – Fistula
    • A patent duct-like structure having both an external (cutaneous) and internal (pharyngeal) orifice

Introduction

• Congenital cysts & sinuses appearing in the neck result from embryonic structures
  – Failure to mature
  – Persist in an aberrant fashion
    • Incomplete closure or resorption

• Diagnosis & Therapy depend on a working knowledge of the embryologic origin & differentiation of the head and neck structures
  – Complete surgical resection → Avoid Recurrence
Embryology

• During the 4th – 8th week after fertilization
  – In the lateral cervicofacial region (side wall of primitive pharynx) human embryo:
    • Branchial arches
      – 6 pairs of well developed ridges
    • Branchial clefts
      – Externally
    • Branchial pouches
      – Internally

Embryology
Embryology

• Branchial Arches
  – Mesodermal core surrounded by epithelium of endodermal & ectodermal origin
  – Arches are separated by deep grooves: Clefts
  – Outpocketings on the lateral wall of pharynx: Pouches
  – Pouches gradually penetrate the surrounding mesenchyme
  – Pleuripotential mesodermal core
    • Cartilage, muscles or arteries
  – Each arch consists of a nerve, artery, and cartilaginous structures
Development

Figure 2  Skeletal and ligamentous derivatives of the branchial arches.

Embryology

The first four branchial arches have clinical significance. During development, the second arch predominates. Indentations between each arch form clefts on the external surface of the embryo and pharyngeal pouches internally. Incomplete closure or resorption/fusion of these primitive branchial clefts may lead to cysts, fistulae or sinus tracts.


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During the 5th week, the ventral aspect of the second arch enlarges and grows caudally to overlap the 2nd, 3rd & 4th arches. By the 7th week, the cervical sinus and the 2nd to 4th grooves disappear, giving a smooth contour to the neck. The remaining 1st groove forms the External Auditory Canal (EAC).

Development

Forebrain prominence
Nasal placode
Nasal pit
Oral plate
Oral opening
Maxillary process
Mandibular arch
Hyoid arch

Development

Development

The Branchial System

• First Branchial System:
  – First Arch
    • Mandibular (Meckle’s) and maxillary process of the upper jaw
    • Muscles- Temporalis, masseter, pterygoids, mylohyoid, ant belly of digastric, tensor tympani, tensor veli palatini
    • Nerve- 5th cranial nerve
    • Artery- Maxillary artery
  – First Pouch
    • Persists as the Eustachian tube, middle ear, portions of the mastoid bone.
  – First Cleft
    • Persists as the external auditory canal, and tympanic membrane
The Branchial System

- Second Branchial System:
  - Second Arch
    - Bones- Upper body & lesser horn of the hyoid, the styloid process and stylohyoid ligament, stapedes superstructure
    - Muscles- platysma, muscles of facial expression, posterior belly of digastric, stylohyoid, and stapedius
    - Foramen cecum & base of tongue
    - Nerve- 7th cranial nerve
    - Artery- stapedial artery
  - Second Pouch
    - Palatine tonsil
    - Supratonsillar fossa

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The Branchial System

• Third Branchial System:
  – Third Arch
    • Lower body of the hyoid and greater cornu.
    • Muscles- stylopharyngeus, superior and middle pharyngeal constrictors.
    • Nerve- 9th cranial nerve
    • Artery- Common carotid and proximal portions of the internal and external carotid.

– Third Pouch
  • Inferior Parathyroid Glands
  • Thymus

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The Branchial System

• Fourth Branchial System
  – Fourth Arch
    • Muscles- cricothyroid, Inferior Pharyngeal Constrictors
    • Thyroids cartilage
    • Nerve- Vagus & Superior Laryngeal Nerve
    • Artery- Right Subclavian, Aortic arch

  – Fourth Pouch
    • Superior Parathyroid Glands
    • Parafollicular thyroid cells
    • Part of thymus

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**Parathyroid Embryology**

- **Thyroid Origin:**
  - **Thyroid Primordium**
  - Endoderm of the floor of mouth between 1st & 2nd arches
  - Origin is Foramen Cecum.
Pediatric Neck Masses

• Lateral Neck Masses
  – Branchial cleft anomalies
    • Fistula > Sinuses > Cysts
  – Lymphatic & Vascular anomalies
  – Dermoid & Teratoid Cysts
  – Torticollis

• Midline Neck Masses
  – Thyroglossal Duct cyst
  – Cervical Lymphadenopathy
  – Plunging Ranula
Branchial Apparatus Remnants

- First Branchial Cleft Anomalies: Located close to the parotid gland
  - **Type I: Cystic mass**
    - Ectodermal duplication anomaly of the EAC
    - The cyst is lined by squamous epithelium without skin appendages
    - These lesions track parallel to the EAC and middle ear
    - Surgical excision in the noninfected state is the treatment of choice

Branchial Apparatus Remnants

• First Branchial Cleft Anomalies
  - Type II:
    • Cyst, fistula and/or sinus and the opening is localized in the anterior neck, always superior to the hyoid bone
    • Ectodermal and mesodermal in origin
    • The tract courses over the angle of the mandible, through the parotid gland, and terminates near the bony-cartilaginous junction of the EAC
    • The definitive excision requires identification of the facial nerve and a superficial parotidectomy is typically required.

Branchial Apparatus Remnants

- **Second Branchial Cleft Anomalies**
  - Most common (90%) of all branchial anomalies
  - They usually present spontaneously as a painless, fluctuant mass of the anterior triangle in infants
  - These lesions may fluctuate after a URI
  - The tract courses deep to the second arch derivatives and superficial to the third arch derivatives
  - If an external opening is present, it is most consistently found at the anterior border of the SCM
  - It then courses anterior to the SCM, superficial to the IX, XII nerve, to turn medial and pass **between the internal and external carotid artery**.
  - The tract is surgically excised.

Second Branchial Cleft Anomalies


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Branchial Apparatus Remnants

• Third Branchial Cleft Anomalies
  – Rare (<2%).
  – They may present in a similar fashion as Second BCC.
  – The internal opening is located within the pyriform sinus. The tract pierces the thyrohyoid membrane cephalad to the superior laryngeal nerve, lateral to the hypoglossal nerve, medial to the glossopharyngeal nerve, and posterior to the internal carotid artery.
  – Surgical approach is usually via a standard thyroidectomy approach to fully visualize the recurrent laryngeal nerves

Summary BCC

Digital pressure applied on tonsillar fossa aids
dissection & ligation of duct near carotid bifurcation
Surgical Excision

Figure 3.54 A branchial sinus tract is dissected from the lower neck to the level of the hypopharynx. "Stepladder" transverse incisions are used to expose the entire tract.

Figure 3.56 Branchial Cleft Cyst. The palpable cyst drained mucus and occasionally pus onto the skin. A lacrimal duct probe enters the tract for identification and ease in dissection.

Figure 3.57 The 4-cm cyst and draining sinus are excised.

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Figure 9  Incision for second branchial cleft cyst.

Figure 10  A stair step incision.

Figure 15  The hypoglossal nerve.

Figure 16  Dissection of the duct.

Thyroglossal Duct Cyst

- The most common midline mass encountered
- As the bilobed thyroid diverticulum descends from the floor of the pharynx, a thyroglossal duct is created
  - Failure of involution of any portion of this tract can lead to a thyroglossal duct cyst
  - The pyramidal lobe, found in 40% of patients, is the failure of closure of the inferior most portion of the thyroglossal duct

Thyroglossal Duct Cyst

Figure 1  Origination and descent of thyroglossal duct cyst.
Foley DS et al. Thyroglossal duct and other midline congenital anomalies. Seminars in Pediatric Surgery. 2006;15;70-75.
Thyroglossal Duct Cyst

• Presentation:
  – The majority of thyroglossal duct cysts present in children and adolescents
    • Asymptomatic midline mass at or below the hyoid bone (i.e. cyst)
    • ¼ present with draining sinus tract (spontaneous drainage)
  – They commonly elevate with tongue protrusion.
  – External sinuses involving the pharynx may rarely occur, and these may become infected & fistulize
  – An ultrasound is the best study to document the presence of normal thyroid tissue

Thyroglossal Duct Cyst

- Due to anatomic proximity with the oral cavity, TDC are prone to infections.
- 1/3 of patients with concurrent or previous history of infection in the cyst.
- The risk of infection is the primary indication for surgical excision.
- **Microbiology:**
  - H. Influ, Staph Auerus and Strep Epidermidis.

Thyroglossal Duct Cyst

• Primary management of thyroglossal duct cysts is with surgical excision.

• Simple cyst excision results in a high rate of recurrence (50%)
  – This procedure has been largely replace by the ‘Sistrunk procedure’

• During this procedure, the central portion of the hyoid is included in the en bloc excision of the cyst, and dissection is carried up into the tongue base.
  – Recurrence rates with this procedure dropped to 4-6%.

• Patients who are at greatest risk for recurrence are those who have had recurrent infections, externally draining sinuses, or prior incision and drainage.
  – For these patients, a modification of the Sistrunk procedure to include en bloc anterior dissection including portions of the strap muscles is advocated to reduce recurrence.

Dermoid and Teratoid Cyst

• Rare causes of neck masses
• Developmental anomaly
  – Composed of different germ cell layers.
• Etiology:
  – Isolation of Pluripotent stem cells
  – Closure of germ cell layers within points of failed embryonic fusion lines.
• Classified according to composition.

Dermoid Cyst

• Mesoderm and Ectoderm in origin
  – Contain Sebaceous glands, Hair follicles & Connective tissue

• Midline or Paramedian
  – Painless masses that usually do not elevate with tongue protrusion

• Commonly misdiagnosed as Thyroglossal Duct Cysts

• Treatment is simple surgical excision
  – Sistrunk is unnecessary

Teratoid Cyst

• Composed of all three germ cell layers
  – Endoderm, mesoderm and ectoderm.
• Larger midline masses, present earlier in life
  – Aero digestive compressive symptoms
• 20% associated maternal polyhydramnios
• Unlike adult teratomas, they rarely demonstrate malignant degeneration.
• Well encapsulated and poorly vascularized
  – Surgical Excision is ideal.

Ranula

• Ranula is a mucus retention cyst arising from an obstructed sublingual gland.

• A simple ranula is confined to the oral cavity as a cystic unilateral mass of the floor of the mouth
  – Sublingual > Submandibular > Parotid

• A plunging ranula may pierce the mylohyoid and present as a paramedian or lateral neck mass with or without an obvious oral cavity ranula

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Ranula

• Simple Ranula:
  – Unroof into the oral cavity & dome of cyst is marsupialized to prevent recurrence
  • The ranula cavity obliterates over 1-2 weeks

• Plunging Ranula:
  – Marsupialization has led to a high rate of recurrence of these lesions
  – Meticulous dissection of the cyst & excision in continuity with the sublingual salivary gland of origin

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Fibromatosis colli (Torticollis)

- Involves the tightness of one SCM following an idiopathic inflammatory process in the newborn period
- Noted by the parent as a painless firm lateral neck mass
  - Birth Trauma?
    - Pregnancy or delivery characterized by breech, unusual presentation, forceps assistance or prolonged labor

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Fibromatosis colli (Torticollis)

• On Examination:
  – Characteristic pose with the chin tipped away from the lesion & the head pulled down to the side of the mass.

• Treatment:
  – Passive (ROM) exercise & stretching + massage with positional changes
  – Surgical division/resection of the SCM for recalcitrant cases

Figure 3.61 Torticollis This newborn with a firm mass in the right sternocleidomastoid muscle tilts his head to that side and lacks full neck rotation to the opposite side.

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Vascular Malformations

• Lymphangioma & Cystic Hygroma
  – At about the 6th week of gestation a system of clefts develops in the cervical mesenchyme → Lymph channels → Jugular lymph sacs → Cervical lymph nodes & lymphatics that drain into IJ Venous system
  – Definition
  • Masses of disorganized, dilated lymph channels that arise when communications with the IJ system fail to develop in portions of the lymphatic channels

Lymphangioma & Cystic Hygroma

• Theory of evolution of lymphangioma
  – The embryological development of the lymphatic system is theorized by the centrifugal theory and the centripetal theory.
  – The centrifugal theory states that lymphatic channels grow outward from venous channels.
  – The centripetal theory states that lymphatic channels grow independently of venous channels.

• Regardless of the theory, the lymphatic cysts become either totally or partially isolated from the venous system

Cystic Hygroma

- Lymphatic malformations
  - Vary in size and location
    - Few centimeters to massive tumor like lesions extending into the mediastinum
      - Macrocystic dilated channels → Cystic Hygroma
  - Location:
    - Lateral cervical & submandibular region
      - Posterior Neck Triangle
    - Extremities, Mediastinum, Retroperitoneum & Trunk

Lymphangioma & Cystic Hygroma

- Lymphatic malformations
  - Most are evident early in life
    - 65% present at birth
    - 80% evident by 12 months of age
    - 90% diagnosed by age of 2 years
  - Presentation:
    - Asymptomatic painless mass lesion and can be located anywhere on the body
    - Hemorrhage can occur within the lesion → rapid enlargement
    - Infection → cellulitis → abscess
    - Pain associated with hemorrhage and infection

Lymphangioma & Cystic Hygroma

• Lymphatic malformations
  – The larger size and obstructive potential of lymphangiomas may lead to CHAOS (Congenital High Airway Obstruction Syndrome).
  – Emergent airway management at the time of delivery is key for survival.
  – Management involves elective Caesarean section with establishment of the airway while still on placental oxygenation.

Lymphangioma & Cystic Hygroma

- **Treatment**
  - Spontaneous regression is rare
  - Surgical excision for obstructing lesions is the treatment of choice
    - Since benign lesions, radical extirpation resulting in loss of function or deformity is not necessary
    - Usually 2/3 amenable to resection
  - Recurrence rates are generally high because of the poor encapsulation and dissection planes

Lymphangioma & Cystic Hygroma

• Treatment
  – Radiation
    • Of no benefit & causes significant morbidity in children
  – Injection of sclerosing agent
    • Bleomycin: High toxic
    • Alcohol
    • OK-432: A monoclonal antibody produced by incubation & interaction of *Streptococcus Pyogenes* with Penicillin
      – Good outcomes but largely experimental at this time

Cervical Lymphadenopathy

• Benign cervical lymphadenopathy
  – Most common neck mass in childhood
    • Head contains many orifices through which bacteria/virus may enter the body
    • Etiology is often viral or is related to Upper Respiratory Tract, Pharynx or skin infection
    • Atypical mycobacterial infection, mononucleosis & cat scratch disease
      – Rare: TB,
    • Reactive Hyperplasia: Adenitis resolves spontaneously
    • Mostly bilateral enlarged nodes in anterior cervical nodes basin
      – Easily palpable between the ages of 2-10
      – In age <2 is most likely a cystic hygroma, TGD cyst, dermoid cyst or a branchial cyst.

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Cervical Lymphadenopathy

• Acute Suppurative Cervical Lymphadenitis
  – Infection of oropharynx
  – Penicillin resistant Staph Auerus & Step Hemolyticus
  – Anaerobes are not uncommon
  – Treatment:
    • Antibiotics
    • Aspiration & Drainage: Diagnostic & Therapeutic
    • May need formal Incision & Drainage.

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Acute Suppurative Cervical Lymphadenitis

Cat Scratch Disease

• Transmitted by cat, dog or monkey

• Organism is Bartonella henselae
  – Self limited regional lymphadenopathy
    • Mild symptoms
    • Complications: Encephlaitis, neuroretinitis & conjuctivitis
  – Treatment:
    • Antibiotics: Azithromycin, Gentamicin, Rifampin
    • 10% of cases will need Incision & Drainage.

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Conclusion

• Lateral Neck Masses
  – Branchial cleft anomalies
    • Fistula > Sinuses > Cysts
  – Lymphatic & Vascular anomalies
  – Dermoid & Teratoid Cysts
  – Torticollis

• Midline Neck Masses
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  – Plunging Ranula
Conclusion

• The evaluation and management of pediatric neck masses requires extensive knowledge
  – Anatomy and embryology of the head & neck

• Diagnosis & Therapy depend on a working knowledge of the embryologic origin & differentiation of the head and neck structures
  – Complete surgical resection → Avoid Recurrence