Congenital Neck Masses

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Case Presentation

- xx year old male presents with sudden onset left lower neck swelling x 1 week
- Denies pain, shortness of breath, dysphagia
- No history of trauma, recent upper respiratory infection
- NKDA
- Medications: none
Physical Exam

• Large swelling in lower half of left neck encompassing lower and medial portions of sternocleidomastoid muscle
• Does not move with deglutition
• No visible sinuses, erythema, bruit appreciated on exam
• No cervical/supraclavicular/axillary lymphadenopathy
Laboratory Values

7.0   13.5   189

41.1

142  105  19

3.8  28  1.0

92
Studies

- U/S guided needle aspiration, pathology negative for malignant cells; demonstrated keratinized squamous cell epithelial cells
- CT Neck: large left lower neck cystic mass measuring 3.5x4.2x7.2 cm, posterior to left sternocleidomastoid muscle. Mass compressing left IJ. Carotid arteries, larynx and periglottic spaces are unremarkable.
Operative Findings

- Massive, deep-seated 6x7cm branchial cleft cyst wrapped around lower 2/3 of carotid sheath
- Vagus, hypoglossal nerves were intimately associated with mass, sharply dissected and preserved
- Estimated blood loss was negligible
Post Operative Recovery

- Patient tolerated procedure well
- Immediate postoperative course complicated by urinary retention
- Discharged home postoperative day 1
- Postop office visit, pt fully recovered
- Pathology consistent with cervical lymphoepithelial cyst (second branchial cyst)
8-1E Pharyngeal arch formation and derivatives

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Branchial Apparatus

- Consists of arches, pouches and clefts
- Arches develop from buds in lateral pharynx
- Ectoderm lined clefts separate arch externally
- Endoderm lined pouches separate internally
- Maldevelopment may result in a neck mass.
8-2A Pharyngeal pouches and their derivatives
Branchial Arch Derivatives

- First Branchial arch (mandibular)
  - Meckel’s cartilage (forms malleus/incus)
  - Muscles of mastication
  - Supplied by trigeminal nerve and external maxillary artery
  - Pouch forms eustachian tube, mesotympanum & mastoid antrum
Branchial Arch Derivatives

• Second Branchial Arch
  – Reichert’s cartilage (hyoid bone, styloid process)
  – Muscles of facial expression
  – Supplied by facial nerve and stapedial artery
  – Pouch forms mesotympanum and palatine tonsil
Branchial Arch Derivatives

• Third Branchial Arch
  – Greater cornu and body of hyoid bone
  – Stylopharyngeus, constrictor muscles of pharynx
  – Supplied by glossopharyngeal nerve and common carotid artery
  – Pouch forms thymus and inferior parathyroids.
Branchial Arch Derivatives

• Fourth Branchial Arch
  – Thyroid cartilage
  – Cricothyroid, inferior pharyngeal constrictor
  – Supplied by superior laryngeal nerve and aortic arch, subclavian artery
  – Pouch forms superior parathyroid glands
Branchial Arch Derivatives

• Fifth/Sixth Branchial Arches
  – Cricoid/Arytenoid cartilages
  – Intrinsic laryngeal
  – Supplied by recurrent laryngeal nerve and ductus arteriosus/pulmonary artery
  – Clinically silent
Congenital Neck Masses

- Branchial Cleft Cyst
- Thyroglossal duct cyst
- Cystic Hygroma
- Dermoid Cyst
- Thymic Cyst
- Ranula
Branchial Cleft Anomalies

- Majority originate from 2\textsuperscript{nd} pharyngeal cleft
- Failure of obliteration of cervical sinus of His
- Present as cysts, sinuses or fistulae
- Lined with squamous epithelium
- Often present with recurrent infection in 2-4\textsuperscript{th} decade of life
Second Branchial Cleft Anomalies

- Sinuses/fistulae occur in lower third of the neck and may be bilateral in 1/3 of cases
- Intimately associated with neurovascular bundle, unlike thyroglossal cyst
- Recurrence is rare, unless infected
- Cysts manifest deep to sternocleidomastoid, near carotid bifurcation/parapharyngeal space
Branchial Cleft Sinuses/Fistulae

- Sinuses: openings on the skin or tonsillar fossa
- Fistulae: communication between skin and pharynx
- Less common than cysts
- Present within first decade of life
- Are bilateral in 20% of cases
- Slight female predominance
Thyroglossal Duct Cyst

- Appear as a painless midline mass
- Present at birth, may not become apparent until adulthood
- Excision indicated, risk of infection/malignancy
- Thyroglossal duct carcinoma arises within thyroid tissue often accompanying epithelial cystic remnants
Diagnosis

- Diagnosis is based almost entirely on history and physical.
- Elevation and movement of lesion with deglutition and protrusion of tongue is pathognomonic.
- Intraoperatively appear thin walled and translucent.
Surgical Treatment

• Sistrunk Procedure involves complete excision of mass, including central portion of hyoid
• Prior to excision, need to confirm presence of normal thyroid tissue.
• Stalk dissected to level of foramen cecum and suture ligated
• Care should be taken not to rupture cyst
Dermoid Cysts

- Consists of tissue for all three germinal layers
- Typically present as midline neck masses
- Rarely present while acutely infected
- Do not move with swallowing
- U/S may not be able to distinguish between dermoid cysts and thyroglossal duct cysts
- Treatment is cystectomy alone
Cystic Hygroma

- Lymphatic malformations from failure of drainage into venous system
- Represent less than 5% of congenital neck masses
- Tend to develop in left posterior triangle
- Present in the first two years of life
- Associated with spontaneous hemorrhage
Clinical Presentation

- Soft, compressible, cystic masses that distort surrounding structures
- Multiloculated cystic structures on u/s or CT
- Vascular malformations may be identified within cystic hygroma
Surgical Resection

- Intimate anatomic involvement may impede complete excision
- Do not follow native tissue planes
- Drainage often utilized to prevent seroma formation in resection of moderate to large lesions
- Variable recurrence rate (6-50%)
Thymic Cyst

- Occur anywhere in lower neck, mostly left side
- Present during the first decade of life
- Male predominance, nearly twice as frequent
- Firm, compressible round lesions, confused with branchial cleft anomalies
Ranula

- Post inflammatory retention cyst of sublingual gland
- Usually involve major salivary glands
- Can fluctuate rapidly in size
- Treatment involves either excision or marsupialization
Summary

• Congenital masses arise from maldevelopment of branchial apparatus or thyroid
• Presentation can vary based on location
• Knowledge of anatomy and embryologic variants is vital to surgical technique
• Imaging studies aid in determining resectability
References

- Pediatric Otolaryngology, The Requisites, Wetmore et al.
- Otolaryngology, 5th Edition, Cummings et al.