Branchial apparatus anomalies

R3 林哲儀
- Branchia: gills
- 30% of congenital neck masses
- Present as cyst, sinus, or fistula
  - Cyst: without external opening
  - Sinus: with external opening
  - Fistula: persistent of branchial groove → pharyngocutaneous fistula
- Male : Female = 1:1
Embryology

- Branchial arches (apparatus)
  - End of 4th gestation, 4 well defined pair of arches and 2 rudimentary arches developed
  - Ectoderm
  - Mesoderm: artery, nerve cartilage, muscle
  - Endoderm
  - Clefts, externally, ectoderm
  - Pouches, internally, endoderm
  - Obliterate to form head and neck structure
  - Incomplete obliteration: branchial anomalies
Table 1  Simplified table of branchial apparatus derivatives: six pairs of branchial arches separated by five paired pharyngeal pouches internally and five paired branchial clefts externally

<table>
<thead>
<tr>
<th>Cleft</th>
<th>Arch</th>
<th>Pouch</th>
</tr>
</thead>
<tbody>
<tr>
<td>1st</td>
<td>EAC</td>
<td>Eustachian tube, tympanic cavity, mastoid air cells</td>
</tr>
<tr>
<td>2nd</td>
<td>Rudimentary</td>
<td>Palatine tonsil</td>
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<tr>
<td>3rd</td>
<td>Rudimentary</td>
<td>Inferior parathyroid glands, thymus, pyriform sinus</td>
</tr>
<tr>
<td>4th</td>
<td>Rudimentary</td>
<td>Superior parathyroid glands, apex pyriform sinus</td>
</tr>
<tr>
<td>5th</td>
<td>Rudimentary</td>
<td>Parafollicular cells of the thyroid gland</td>
</tr>
<tr>
<td>6th</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td></td>
<td>Arytenoids and cricoid cartilages and cranial nerve X</td>
<td>–</td>
</tr>
</tbody>
</table>
Pathology

- Branchial anomalies lined with respiratory or squamous epithelium
  - Cyst: squamous epithelium
    - May mixed with follicular-lymphoid tissue, sebaceous gland, salivary tissue, cholesterol crystals
  - Sinus: ciliated or columnar epithelium
    - Nonfollicular lymphoid tissue
  - Squamous cell carcinoma: rare, but most in adults
Diagnosis

- Upper airway endoscopy
  - Pharyngeal opening
  - Tonsillar fossa
  - Pyriform sinus
- Fine needle aspiration
  - For rule out metastatic carcinoma
  - Avoid incision biopsy
- CT is first choice
- MRI, sonography
- Esophagography: 80% sensitivity for 3rd and 4th fistula
Treatment

- Complete surgical excision
- Timing of operation
  - Early resection for prevent infection
- Acute infection
  - Antibiotic treatment first
  - Incision and drainage
  - Complete resection after resolution
First branchial cleft anomaly

- Rare, 1% of branchial cleft malformation
  - Female > male
  - May involve EAC, tympanic membrane or middle ear
  - Close to the parotid gland, superficial lobe
  - Traveling through above, or below the facial nerve branches → opening at mandible angle
  - Symptom: Otorrhea, parotid swelling, mandible pit discharge……
  - Histology: most commonly lined by stratified squamous epithelium

- 2 types, Arnot, 1971, base on anatomic location
Type I
- Lower pole of the parotid gland
- Closely to lower branches of facial nerve
- Passing between the mandible and the stylohyoid ligament

Type II
- Anterior and below the mandible angle
- Ending at bony part of EAC
• Type I (Work, 1972)
  • duplications of the membranous external auditory canal, parallelly
  • Composed of ectoderm only
  • Within parotid gland, lower pole
  • Superior to the facial nerve, and between the mandible and stylohyoid ligament
Type II (Work, 1972)
- Composed of ectoderm and mesoderm, contain cartilage, hair, or glands
- End at bony part of EAC
- Substance within parotid gland and close to the facial nerve
- Present as preauricular, infraauricular, or postauricular
- Inferior to the angle of the mandible, anterior to SCM
Treatment

- Standard cervico-mastoid-facial parotidectomy incision with dissection of the facial nerve and superficial parotidectomy
- Assisted by injection of methylene blue or probe guide to identified the tract
- Recurrence: common, average receive 2.4 procedure
Second cleft anomalies

- ~90% of all branchial cleft malformation,
- Incidence: cyst > fistula
- Present as recurrent unilateral tonsillitis or parapharyngeal abscesses
- Cyst:
  - Most diagnosed at age of 30~ 50
  - Acute enlargement after a URI
  - Lead to airway compromised, torticollis, dysphagia
- Fistula
  - Most diagnosed at infant
  - Present with chronic discharge along anterior SCM in lower 1/3 neck
Traverse deeply to second arch structures and superficially to third arch structures

- external opening → pierce platysma → run superiorly along the carotid sheath → turn medially to hypoglossal nerve → beneath digastric muscle (post belly) → course between the internal and external carotid arteries → cross glossopharyngeal nerve → internal opening in the tonsillar fossa

- most common: lateral to internal jugular vein (carotid bifurcation)
- **Type I**
  - anterior to the SCM, not contact the carotid sheath

- **Type II**
  - the most common
  - deep to the SCM, either anterior or posterior to the carotid sheath, lateral to IJV
- **Type III**
  - between the internal and external carotid arteries, adjacent to the pharynx

- **Type IV**
  - medial to the carotid sheath, close to the pharynx, adjacent to the tonsillar fossa.
- **Treatment**
  - Transverse incision over skin fold
  - Exploration of an associated fistula with complete excision
  - Monofilament or probe for cannulating the fistula tract
  - Finger assisted to identified the internal opening in tonsillar fossa
  - Cysts lying medial to the carotid sheath may be more easily removed transorally
Recurrent rate, Mayo Clinic
- 21% when there was a history of prior surgery
- 14% when there was a history of infection
- 3% for cases with no history of prior surgery or infection

Infection and prior surgery may cause adhesions and distortion of tissue planes, which may lead to higher recurrence rates and an increased risk of damage to nearby nerves and vessels.
Third and fourth branchial anomalies

- **Sandborn, 1972**, the first description
  - sinus tract originating from the piriform fossa might represent a branchial anomaly

- **Takai, 1979**
  - Persistent sinus of pyriform fossa with acute suppurative thyroiditis

- Rare, 3\textsuperscript{rd} branchial anomalies representing 2\% to 8\% and 4\textsuperscript{th} branchial anomalies representing 1\% to 2\%

- Pouch: from pharynx to hypoid bone $\rightarrow$ open into pyriform sinus
Third branchial anomaly

- pyriform fossa → pierce thyrohyoid membrane → passing cranially to the superior laryngeal n. → pass superficial to the hypoglossal n., deep to the glossopharyngeal n. and the ICA → platysma → anterior border of SCM at lower neck
Fourth branchial cleft anomaly

- Pyriform sinus → circothyroid joint → between sup. laryngeal nerve and recurrent laryngeal nerve →
  - Left site: → descending along trachea and esophagus to mediastinum → looping aorta anteriorly
  - Right site: → descending along trachea and esophagus → looping subclavian artery anteriorly
- Ascending the neck → posterior to the internal and common carotid arteries → exit anterior to SCM in the lower neck
Distinguish from 3\textsuperscript{rd} from 4\textsuperscript{th} branchial anomaly

relationship of the anomalous tract to the superior and recurrent laryngeal nerves
- inferior to the superior laryngeal nerve and superior to the recurrent laryngeal nerve $\rightarrow$ 4\textsuperscript{th} pouch origin
- pass cranial to the superior laryngeal nerve and inferior constrictor muscle $\rightarrow$ 3\textsuperscript{rd} pouch origin

Histology finding
- Ectopic thymic tissue(+), inferior parathyroid tissue(+), superior parathyroid gland involvement(-) $\rightarrow$ 3\textsuperscript{rd} arch anomaly

Case of 4\textsuperscript{th} pouch sinus descending below clavicle is rare
4th branchial pouch anomaly
- most in children, female predominant
- 97% left predominant,
  - Related to asymmetric development of 4th arch vascular structure
  - In neonatal: present as lateral neck cyst or abscess with obstructive airway symptoms
  - In children or adult: recurrent lateral cervical abscess and recurrent suppurative thyroiditis
  - Retrograde migration of infective material from hypopharynx
  - May present as recurrent retropharyngeal abscess
- **Special exam**
  - Palin film: air in cyst
  - Flexible nasopharyngoscope
  - Pharyngoesophagogram with barium study
    - may be performed at least 6 week later than acute infection resolution
  - CT scan, FNA
  - Thyroid scan
  - Reflux of pus under laryngoscope
Hae-EL G, 1991:
- Suggest an immediate search for an internal pharyngeal sinus after a child's first episode of acute thyroiditis
Treatment

- External approach
  - Excision the tract with cannulation under endoscopy assisted
  - Ligating and dividing the tract
  - Ipilateral hemithyroidectomy with partial resection of thyroid cartilage for 4th pouch anomaly

- Internal approach
  - Endoscopic electrocauterization, 2004, D J Verret
  - Endoscopic chemical cauterization, 2008, K D Pepeira
Chemical cauterization by AgNO₃
Complications

- Permanent recurrent laryngeal nerve paralysis
- Post operative esophagocutaneous fistula
- Hypoglossal palsy
- Head and Neck Anomalies Related to the Branchial Apparatus David L. Mandell MD Otolaryngology clin N am 33(6)