OVERVIEW NECK CYSTIC MASSES - DIFFERENTIAL DIAGNOSIS

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Branchial cysts-distribution

Cystic masses of the neck are either congenital or acquired. The latter are usually of inflammatory or neoplastic nature. Branchial cleft cysts (BCCs) are congenital developmental malformations. However, they are not often noticed before adolescence. They are painless, cystic swellings in the neck of about 2cm to 4cm. However, may significantly enlarge and become symptomatic after respiratory tract infections.

- The 1st branchial cleft cyst is less common than the 2nd BCC. Three groups are identified according to the anatomic location. Type I: medial-inferior- posterior to the auricle type II: Between the angle of the mandible and the auricle III: in the parotid region.
- The 2nd branchial cyst (90%) occurs usually below the angle of the mandible anterior and deep to the SCM muscle (at the level between its upper and middle third) and lateral to the carotid sheath and less commonly extends between the bifurcation of IC and EC or even medially to the carotid sheath.
- The 3rd and 4th branchial cleft cysts are exceedingly rare.
- 3rd BCCs are posterior to the common or internal carotid artery and the sternocleidomastoid muscle, between the hypoglossal nerve below and the glossopharyngeal nerve above.
- 4th BCCs are generally sinus tracts or fistulas and arise from the pyriform sinus, pierce the thyrohyoid membrane, and descend along the tracheoesophageal groove. Branchial cleft cysts may become infected and exhibit thickening and enhancement of the cyst wall, mimicking a suppurative lymph node.
Evaluation of the cystic neck mass

- **Evaluation includes a complete history and physical examination.** 1. duration of the mass 2. development of pain or fever and enlargement on upper RT infections 3. The patient’s age, rapid increase in lesion’s size, past history of radiation, malignancy, or immunodeficiency disorders are strong predictors of a potentially malignant lesion.

- **Upper airway endoscopy** is a useful diagnostic method in exploring for a pharyngeal opening, particularly in third and fourth anomalies and should be performed on children with recurrent neck abscess as well as on signs of suppurative thyroiditis. Endoscopy and **barium swallow** increase the identification rate of pyriform sinus fistula (PSF) to almost 100%. Direct laryngoscopy has a positive predictive value of 82% for third and 90% for fourth branchial anomalies.

- High-resolution **ultrasound (US)** is an ideal initial imaging investigation for neck masses as it reveals the cystic nature in most cases and localizes the mass in relationship to the surrounding structures.
Imaging and d.d. from metastatic carcinomas

- The 2nd BCCs is mostly seen as a well-marginated anechoic mass with a thin, well-defined wall at the anteromedial border of the sternocleidomastoid muscle.

- The CT scan may show thickened walls or internal septations and fat stranding after recurrent inflammation.

- MRI in delineating fistula or sinus tracts is preferable though not always accurate.

- Fine-needle aspiration cytology (FNAC) may be required as a supplementary test to confirm diagnosis. Identification of cholesterol crystals is considered pathognomonic for B.C.C. However, cytology cannot always distinguish cyst from carcinomatous lymph node metastasis and is not considered diagnostic by itself.

- Cytology in infected branchial cyst might show atypical epithelial cells.

- High concentration of C.E.A in the cystic fluid might play a role in differential diagnosis.

In patients >40y there is higher malignancy risk and degenerative metastatic lymphadenopathy (head/neck SCC or thyroid carcinoma) should always be considered in differential diagnosis.
Enucleation is the treatment of choice

38y male with prominent swelling R neck. Marking: angle of the mandible, horizontal incision. Exposed large branchial cyst on retraction of the anterior border of SCM. Since a true fistula towards the tonsillar fossa is been eliminated no recurrence is expected.

Insertion of drain tube and overnight stay in the hospital is advisable
The proximity of the XI and XII nerves demands cautious dissection of a large 2\textsuperscript{nd} branchial cyst.
Represents 70% of the congenital neck anomalies. The cyst can develop at any point along the residual tract (duct remnants) left over after the completion of thyroid gland formation and descent from the foramen cecum (tongue base) to its final position at inferior neck.

- About 90% of cases become apparent in childhood especially during the first decade. However, a significant number of patients are first diagnosed in adulthood.
- No gender predilection has been reported. The cyst develops at the level of hyoid bone (15-50%), infrahyoid (25-65%), or in supraphyoid location (20-25%).
- Typically the cyst is deep to or embedded in the infrahyoid strap muscles.
- The more inferior the cyst occurs, the more likely to be off the midline.
- In adults, thyroid carcinoma can develop in a TGDC, with an incidence of <1%.
  Most common type is the papillary carcinoma.
- The Sistrunk procedure is considered as adequate treatment for most patients with clinically and radiologically normal thyroid gland and incidentally diagnosed carcinoma in the TG cyst wall.

❖ 25y female with a history of midline painless lump at the level of hyoid bone. Shown the pathognomonic elevation on tongue protrusion. On CT imaging the lesion was off midline. Sistrunk procedure to include enucleation of the cyst and partial excision of the central part of the hyoid bone.
Imaging of TGDCs

• On all imaging modalities, TGD cysts appear as cystlike masses of the anterior neck deep to platysma, usually at the level of the hyoid bone or within the strap muscles. Approximately 75% percent of all TGDCs are located in the midline, with 25% within 2 cm of midline.

• With imaging, TGDCs generally appear as smooth, well-circumscribed lesions containing simple fluid.

• Thyroglossal duct cysts may exhibit peripheral enhancement following intravenous contrast administration. With recurrent inflammation, the cysts may become more heterogeneous in appearance and develop internal septations.
Histopathology

**Branchial cyst**
- Branchial cysts are lined by stratified squamous epithelium, or rarely by ciliated columnar epithelium, subepithelial abundant lymphoid tissue. If the cyst is infected there is significant attenuation of the epithelial lining. Carcinoma arising in these cystic lesions has been identified in adults as a rare finding.

**Thyroglossal duct cyst**
- Evidence of respiratory / squamous epithelial lining and thyroid follicles as well as inflammatory cells.
Dermoid and epidermoid cysts

- Dermoid cysts usually occur during the second and third decades of life as a slow-growing mass in the sub-mandibular or sublingual space. Epidermoid cysts appear earlier in life and quite often are evident since infancy.

- The distinct characteristic between epidermoid and dermoid cyst is that the latter contains skin appendages to include hair follicles and sebaceous glands within the cystic wall.

- Approximately 7% occur in the head and neck region and there is preference for the skin area lateral to the eyebrow. They develop in the oral cavity with predilection of the floor of the mouth. In such case there is progressive increase in size along with elevation of the floor of the mouth as well as difficulties related to speech, chewing and swallowing.

- Has been noted that there is a risk up to 5% for malignant transformation especially in large, long-standing dermoid cysts in older adults. The majority of malignancies are proved to be epithelial in origin. However, carcinosarcomas have also been described.
Dermoid and epidermoid cysts

- Ultrasound is the initial imaging modality. Epidermoid cysts are seen as well-defined cysts with multiple well-defined dependent echogenic nodules within the cyst.
- Computed tomography scan shows a unilocular cyst with homogenous, hypo-attenuating (0-18 HU) fluid material that contains multiple hypo-attenuating fat density nodules giving a “sack of marbles” appearance; this is a pathognomonic feature for a dermoid cyst.
- MRI facilitates visualization of the exact location and extent of cystic lesions in the floor of the mouth and is useful for determining their relationship to the surrounding muscles.
- Lesions above the mylohyoid muscles are operated via intraoral approach, whereas those below the muscle are removed via an incision in the neck. However, if there is a very large sublingual cyst above the mylohyoid muscle, an extraoral approach may be preferred.
Gardner’s syndrome

- **Gardner’s syndrome (GS)** is a hereditary disorder inherited as autosomal dominant with complete penetrance and variable expression. GS is a variant of familial adenomatous polyposis (F.A.P) characterized by extracolonic manifestations to include osteomas skull (frontal region, mandible), dental anomalies (supernumerary teeth, odontomes, multiple teeth impaction, high index of caries), abnormality of skin pigmentation, change of facial skeleton proportions due to mandibular osteomas and epidermoid cysts.

- The Dentist plays critical role in early diagnosis. “Gardner syndrome teeth” might be the first sign of the inherited Gardner syndrome. However, having a family history warrants a consultation with a genetic specialist.

**Prognosis:** the polyps in the colon and rectum are likely to show malignant change by early 30-40s. The number of polyps in the intestines increases by age and most people with F.A.P. eventually undergo preventive removal of the large intestine. The polyps in the duodenum are also prone to malignant change but they can be managed by careful monitoring and selective removal accordingly.
Lymphangioma

- **Lymphangioma (Cervicofacial)**
  - Rare malformation due to early sequestration of embryonic lymphatic channels, more often developing along the jugular chain. 50% of cases are noted at birth and the rest before the 5th year of age.
  - Different types of lymphangioma are described based on microscopic size of dilated lymphatic channels to include: *cystic hygroma, cavernous lymphangioma, capillary lymphangioma, and vasculolymphatic malformation, lymphangioma circumscriptum* (cluster of tiny abscesses like lesions that contain lymph fluid)
  - **Congenital Cystic hygromas** are the most common form of lymphangioma. 75% of these occur in the posterior triangle of the neck. Could be associated with karyotypic anomalies or congenital syndromes (Down or Turner syndrome)
  - Treatment: Dye laser, sclerotherapy, surgery.
Ranula: Cystic-like lesion of the floor of the mouth, that develops more often in childhood as a secondary manifestation to trauma and mucus extravasation or obstruction of the sublingual duct. Therefore, these are also called sublingual gland mucocele or mucous retention cyst.

- Either “simple” confined to the sublingual space or “plunging-diving” extending posteriorly into submandibular space or via a gap in mylohyoid muscle.

Thymic cysts Uncommon lesions that arise from remnants of the thymopharyngeal duct. They occur adjacent to the carotid sheath anywhere from the hyoid bone to the anterior mediastinum. The common age of presentation is 2-15 years, with slight male predilection. The thymic cysts may have a similar appearance to third and fourth BCCs,

Laryngocele

- A laryngocele is an uncommon congenital malformation. However, it is usually identified in adulthood around 40s.
- It is a benign dilatation of the saccule, a small pouch arising from the roof of laryngeal ventricle, laryngoceles have dilatation of the saccule on both sides of the thyrohyoid membrane. Fifteen percent of laryngoceles are associated with a tumor occluding the orifice of the laryngeal ventricle. Laryngocele may become infected.

Cervical bronchogenic cysts. Extremely rare lesion as a result of foregut developmental anomaly.

- They are usually located in the thyroid or para-tracheal region, rarely in the suprasternal or supraclavicular location.
- To confirm the diagnosis of bronchogenic cyst histopathological examination is required.

Metastatic lymph nodes in the neck. The vast majority (80%) of neck cystic-like masses in patients over 40 years of age are due to degenerative necrotic lymph nodes.

Inflammatory lesions to include abscess, suppurative thyroiditis

Neurogenic tumors

Vascular lesions

Thyroid gland cysts