Congenital cystic lesions of the neck - the role of diagnostic imaging

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Learning objectives

- Describe the general imaging features of congenital cystic neck lesions.
- Recognise typical imaging features of complicated congenital cystic cervical lesions.
- Review relevant imaging features relative to embryology and anatomy of neck for correct diagnosis, in most cases with histopathological correlation.
Background

The congenital cystic neck lesions are generally uncommon malformations connected with defective/abnormal embryonic development. Most of congenital cystic cervical lesions manifest in childhood or adolescence, and rarely in young adulthood.

The congenital cystic lesions of the neck according to their localisation include thyroglossal duct cyst, branchial cleft cyst, cystic hygroma, thymic cyst, laryngocele, cervical bronchogenic cyst, dermoid and epidermoid cyst. The most common are thyroglossal duct cyst and second branchial cleft cyst.

In the evaluation of congenital cystic neck lesions the most important factors are clinical history and physical examination. First important diagnostic information is the patient’s age. Next clinical information is commonly a slow-growing mass observed on the neck. In most cases painless or fluctuant palpable cervical neck mass is the first clinical manifestation. In other cases cystic cervical neck lesion is the accidental finding in diagnostic imaging. In most cases congenital cystic neck lesions are benign lesions. However, there are complications such as inflammatory conditions and less frequent malignant transformation.
Findings and procedure details

A. Procedure details:

Ultrasonography (US) is an ideal initial imaging method for congenital cystic lesions of the neck. It is non-invasive examination, without radiation exposure and sedation of patients. Ultrasonography of cervical palpable mass is able to confirm the cystic nature of lesions, and distinguish between solid lesions of the neck. In addition, it is a low cost examination. Limitations of ultrasonography are lack of information about the structure of the wall of the cystic lesion, exact location in deeper structures, post-contrast attenuation and internal architecture. If inflammation is present, low-level internal echoes may be seen.

Multidetector computed tomography (MDCT) and magnetic resonance imaging (MRI) furthermore characterise the location, visualise the characteristic features, potential vascularisation and complications /inflammatory conditions, malignant changes of cystic lesions/. MDCT is processed in non-contrast phase, but also a post-contrast phase is required. MDCT is preferable for detecting calcification. MRI can be principally relevant in cases of expansion into the mediastinum or deep neck spaces. Complicated cystic neck lesions are characterized by increased attenuation caused by intracystic bleeding, solidification of cystic fluid, thickening of the cystic wall /well- or ill-defined borders/ in cases with infected cystic lesions or malignant transformation of cystic neck lesion.

In some cases fine-needle aspiration cytology may be required for the confirmation of the lesion. Extirpation of the neck cystic lesion with exact histological examination is necessary and recommended in all cases except hygroma/lymphangioma/ in special cases.

B. Findings:

1. Thyroglossal duct cyst

Thyroglossal duct cyst (TGDC) is the most common congenital cystic lesion of the neck, with typical midline anterior location at or below the level of the hyoid bone (Fig. 1.), sometimes in a paramedian location (Fig. 2.) or between strap muscles (Fig. 3.) /70% from all congenital cystic neck anomalies/.

Thyroglossal duct is a result of an embryonic migration of the thyroid gland - it descends from foramen caecum located at the base of the tongue to its final position. Thyroglossal duct cyst is the remnant of the thyroglossal duct. TGDC may contain thyroid gland tissue /
ectopic rests from thyroid gland descending/ and that is the reason, why in 1% TGDC is associated with thyroid carcinoma. In all cases complete excision is the recommended surgical method.

Typical anechoic cystic appearance on US is only in 42% cases. Other US findings show hypo-echoic echo pattern, internal echoes, proteinaceous content. On MDCT, typical thyroglossal duct cyst is homogeneously hypo-attenuated well-circumscribed, thin-walled lesion /attenuation like fluid/. On post-contrast MDCT scans it may show peripheral wall enhancement. In cases with complication there may show higher attenuation, septations, solid component, higher peripheral enhancement, reaction of around soft tissue (Fig. 4.). On MRI, it is hyper-intense on T2-weighted sequences, on T1- weighted images signal intensity is more variable.

2. Branchial cleft cyst

Branchial cleft anomalies are the most common congenital anomalies in children. Branchial cleft anomalies such as cyst, sinus or fistulous tract arise from incomplete obliteration of a part of any branchial apparatus. According to the cleft of origin branchial cleft anomalies are classified as first, second, third or fourth arch anomalies.

2.a. First branchial cleft cysts are residual embryonic tract of the first branchial cleft. A bit higher prevalence is seen in middle-aged women. In clinical history there is important information about recurrent inflammations or recurrent parotid abscesses unresponsive to antibiotic therapy or drainage. The first branchial cleft cyst is located from the external auditory canal through the parotid gland to the submandibular triangle. It may be superficial to, or deep to the parotid gland. The first branchial cleft cyst has very variable appearance on US, MDCT or MRI, because of recurrent infections. In the differential diagnosis radiologist have to consider cystic masses of parotid or peri-parotid region.

2.b. Second branchial cleft cysts /lateral cysts/ are the most common of the all branchial cleft anomalies /95%/ . According to Bailey, second brachial cleft cysts are divided into four types:

- type I. - superficial, anterior to the sternocleidomastoid muscle and deep to the platysma
- type II. - deep anteromedial to the sternocleidomastoid muscle, lateral to the carotid space and posterior to the submandibular gland (Fig.6. and 7.)
- type III. - medially between the bifurcation of internal and external carotid arteries
- type IV. - in pharyngeal mucosal space medial to the carotid sheath
On US, branchial cleft cysts have typical cystic appearance. On MDCT, branchial cleft cysts are well-circumscribed, homogeneously hypo-attenuated lesions with thin wall. On MRI, these lesions appear like typical cystic masses. Depending on the presence and severity of any associated inflammatory process or malignant transformation may be seen mural thickness, increase variable peripheral enhancement or higher attenuation/variable intensity (Fig. 8. and 9.).

2.c. Third and fourth branchial cleft cysts are extremely rare. Third branchial cleft cysts are located in the posterior cervical space, posterior to the common or internal carotid artery, above the superior laryngeal nerve, below the hypoglossal nerve. Fourth branchial cleft cysts are located below the superior laryngeal nerve with various locations, including the thyroid gland and mediastinum. Both are related to the pyriform sinus in case of complete fistula. These lesions appear like typical cystic masses on US, MDCT or MRI.

3. Cystic hygromas

Cystic hygromas are common form of lymphangiomas, which are developmental anomalies of vasculolymphatic origin /early sequestration of embryonic lymphatic channels/. Cystic hygromas are characterised like expansive cystic mass localised in the posterior triangle of the neck, sublingual or in submandibular space. Cystic hygromas may have infiltrative character. In many cases these lesions do not respect fascial planes and occur in lower neck, axilla or upper mediastinum. Most of the cases are reported in infancy and early childhood /by the age of 2 years/. Cystic hygromas may be classified according size as macrocystic /more than 1 cm/, microcystic /less than 1 cm/ and mixed cystic forms. On US, cystic hygromas may be uni- or multi-locular anechoic lesion, with echogenic septations. Microcystic hygromas occur hyperechoic and can be confused with solid neck lesion. On MDCT, these lesions are uni- or multi-locular hypo-attenuated and poorly circumscribed lesion, with multiple septations in posterior triangle of the neck. In that case MRI is more sensitive than MDCT to show the relationship of the lesion to the surrounding soft tissue.

4. Dermoid and epidermoid cysts

Dermoid and epidermoid cysts are sequestration of ectoderm tissue, localised in floor of the mouth, sublingual or submandibular space in the midline. Dermoid cysts contain squamous epithelium and skin appendages, but epidermoid cysts are only formed of squamous epithelium. Complex dermoid cysts contain mesodermal elements. Epidermoid cyst in 5% may lead to malignancy.
Dermoid and epidermoid cysts are seen like well-defined anechoic/hypo-attenuated/fluid intensity formation on US, MDCT or MRI. Nevertheless, dermoid cysts often have variable appearance on US, MDCT and MRI, because of heterogeneous germinal components. In some cases a typical sign called sack-of-marbles is seen, which is caused by small multiple coalescence of the fat into small nodules.

5. Thymic cysts

Thymic cysts are extremely sporadic, with a bit higher male predilection between age 2-15 years. Persistence of the thymopharyngeal duct can result in thymic cysts. Thymic cysts occur anywhere from the angle of mandible to the anterior mediastinum, however more often below hyoid bone. These lesions have very similar appearance to third or fourth branchial cleft cysts, and only the proof of thymic tissue can confirm the definitive diagnosis of thymic cyst. Thymic cysts are seen like uni-locular well-defined anechoic/hypo-attenuated/fluid intensity formation on US, MDCT or MRI.

6. Laryngoecele

Laryngoecele is congenital derivation with dilatation of the laryngeal saccule. There are three types of laryngoecele - internal, external and mixed. The clinical appearance is similar to third or fourth branchial cleft cysts arising in the larynx, on MDCT or MRI. Sometimes may be seen air-fluid level. Communication between the sack and airways needs to be confirmed for definitive diagnosis.

7. Cervical bronchogenic cysts

Cervical bronchogenic cysts are extremely rare, with a bit higher male prevalence. These lesions are as result from an anomalous foregut development. Cervical bronchogenic cysts are located in thyroid and parathyroid space, usually in tubular shape anterior to the trachea.
Fig. 1: Thyroglossal duct cyst in a 44-year-old woman with 2-weeks swallowing problems. (A.) Axial non-contrast and (B.) axial contrast-enhanced CT scans show a small well-circumscribed lesion in the anterior midline of the neck below the hyoid bone with (A.) homogeneous attenuation /around 15-18HU/ and (B.) discrete peripheral rim enhancement on contrast enhanced scan. (C.) Axial non-contrast CT scan, (D.) axial and (E.) sagittal contrast-enhanced CT scans after one year - there is no change in character of cystic lesion.

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Fig. 2: Thyroglossal duct cyst with left paramedian location in a 21-year-old woman with palpable mass on the neck observing one year. (A., B.) Axial, (C.) coronal and (D.) sagittal non-contrast CT scans show a homogenous cystic mass in left slightly off-midline location of the neck below the level of the hyoid bone, the mass is closely to the ventral part of the left strap muscles.

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Fig. 3: Thyroglossal duct cyst with left paramedian location in a 55-year-old woman with palpable mass on the neck observing half a year. (A.) Axial non-contrast CT scan shows a homogeneously attenuated cystic mass in the left strap muscles. (B.) Axial, (C.) coronal and (D.) sagittal contrast-enhanced CT scans show a small cystic mass in the left strap muscles with enhanced capsule and septations.

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**Fig. 4:** Thyroglossal duct cyst in a 56-year-old man with palpable neck mass. (A.) Axial non-contrast CT scan shows a homogeneously iso-attenuated mass in the anterior midline of the neck below the level of the hyoid bone, with attenuation around 40 HU. (B.) Axial, (C.) sagittal and (D.) coronal contrast-enhanced CT scans show homogeneous contrast-enhancement /55HU/ intracapsular, also discrete peripheral capsular rim enhancement - suspicion of intracapsular bleeding transformation or post-inflammation changes /that mass has typical cystic density at CT scans two years before/.

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Fig. 5: Thyroglossal duct cyst in a 28-year-old man with palpable mass during 5 years, without enlarging. (A.) Axial non-contrast CT scan shows a large well-circumscribed lesion with homogeneous attenuation /15-20 HU/ in the anterior midline of the neck. (B.) Axial contrast-enhanced CT scan after 35 seconds shows leading peripheral rim enhancement of thickened capsule. (C.) Axial, (D., E.) sagittal and (F.) coronal contrast-enhanced CT scans made after next 35 seconds /70 seconds after intravenous contrast injection), they show massive peripheral rim enhancement of thickened capsule /uniformly-thickened cystic wall up to 7 mm/ of the cervical mass at the level of the hyoid bone.

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**Fig. 6:** Second branchial cleft cyst in a 21-year old woman with one year painless palpable mass. Ultrasound image shows sharply defined ovoid anechoic mass with peripheral wall and distinct acoustic shadowing in right lateral cervical space.

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**Fig. 7:** Second branchial cleft cyst in a 21-year old woman with one year painless palpable mass, the same patient like on fig.6. (A.) Axial non-contrast CT scan shows a well-circumscribed, homogeneously hypo-attenuated lesion in right location. (B.) Axial, (C.) sagittal and (D., E., F.) coronal contrast-enhanced CT scans show mass surrounded by a uniformly thin wall in typical location. The lesion in typical location displaces the sternocleidomastoid muscle posterolaterally, pushes the vessels of the carotid space posteromedially and displaces the submandibular gland anteriorly.

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Fig. 8: Second branchial cleft cyst with acute inflammation in a 58-year old man with 2 months palpable painless mass. (A.) Axial non-contrast CT scan shows a well-circumscribed, homogeneously iso-attenuated lesion in right typical location /around 60HU/. (B.) Axial, (C.) coronal and (D.) sagittal contrast-enhanced CT scans show lesion surrounded by peripheral enhanced and uniformly thickened wall up to 2.5 mm.

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Fig. 9: Second branchial cleft cyst with chronic inflammation with purulent content in a 34-year old woman with enlarging painful neck mass. (A.) Axial non-contrast CT scan shows a large ill-circumscribed mass with homogeneous attenuation /18-25 HU/ in the left lateral cervical space. (B.) Axial contrast-enhanced CT scan after 35 seconds shows leading peripheral rim enhancement of thickened capsule and distinguish surrounded soft neck tissue with reactive changes. (C., D.) Axial, (E.) coronal and (F.) sagittal contrast-enhanced CT scans made after next 35 seconds /70 seconds after intravenous contrast injection) show massive peripheral rim enhancement of thickened capsule /uniformly-thickened cystic wall up to 3-4 mm/ and septation of the lateral cervical mass.

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Conclusion

Exact diagnosis of congenital cystic lesion of the neck is clear and easy when embryological knowledge and clinical history correlate with imaging findings such as location, internal architecture and post-contrast enhancement. Complicated congenital cystic cervical lesions can cause diagnostic problems for radiologist and consideration of a wider range of differential diagnosis is needed. MDCT and MRI imaging allow main information on congenital cystic neck lesion location essential for optimal preoperative planning.
References


