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Congenital Cystic Masses of the Neck: Radiologic-Pathologic Correlation[1]

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Cervical congenital cystic masses constitute an uncommon group of lesions usually diagnosed in infancy and childhood. The most common congenital neck mass is the thyroglossal duct cyst. The diagnosis is easily established from the presence of a cystic lesion in the anterior midline portion of the neck. The vast majority of branchial cleft cysts arise from the second branchial cleft. They can occur anywhere from the oropharyngeal tonsillar fossa to the supraclavicular region of the neck. Cystic hygroma is the most common form of lymphangioma. In the neck, cystic hygromas are most commonly found in the posterior cervical space. They typically extend into adjacent structures without respecting the fascial planes. Dermoid and epidermoid cysts result from sequestration of ectodermal tissue. The floor of the mouth is the most common location in the neck. Cervical thymic cysts are very uncommon lesions and are found anywhere from the angle of the mandible down to the sternum. Laryngoceles are classified into internal, external, and mixed types and have a frequent association with laryngeal carcinoma.

LEARNING OBJECTIVES

After reading this article and taking the test, the reader will be able to:

- Describe the current theories about pathogenesis of congenital cystic masses of the neck.
- Identify typical imaging appearances of thyroglossal duct cysts, branchial cleft cysts, cystic hygroma, dermoid and epidermoid cysts, cervical thymic cysts, and laryngoceles.
- Understand the importance of neck anatomy in establishing the radiologic diagnosis for these lesions.

■ INTRODUCTION

Congenital cystic masses of the neck include thyroglossal duct cysts, branchial cleft cysts, cystic hygromas, dermoid and epidermoid cysts, thymic and bronchogenic cysts (visceral cysts), and laryngoceles. These lesions vary in prevalence from common (thyroglossal duct cysts, branchial cleft cysts, and cystic hygromas) to very rare (thymic and cervical bronchogenic cysts). The absolute number remains unknown.

Abbreviation: H-E = hematoxylin-eosin

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Clinical Features of Congenital Cervical Lesions

Lesion	Peak Prevalence (Age)	Sex Predilection	Usual Location
Thyroglossal duct cyst	<10 y	Equal	Hyoid level or below (80%), within 2 cm of midline
Branchial cleft cyst			
First	Middle age	F > M	Parotid, external auditory canal
Second	10–40 y	Equal	Submandibular space, lateral to carotid vessels
Third	10–30 y	...	Left posterior cervical triangle
Fourth	Any age	...	Sinus tract arising from left pyriform sinus
Cystic hygroma	<2 y	Equal	Posterior cervical triangle, oral cavity
Dermoid cyst	10–30 y	Equal	Floor of mouth
Epidermoid cyst	Infancy	Equal	Floor of mouth
Thymic cyst	2–13 y	M > F	Low anterolateral neck (L > R)
Bronchogenic cyst	Any age	M > F	Low anterolateral neck
Laryngocele	Adulthood	M > F	Lateral supraglottic region

Note.—F = female, M = male, L = left, R = right.

The clinical history and physical examination of the patient are the most important factors in the evaluation of a congenital neck mass. An appropriate knowledge of the embryology and anatomy of the cervical region frequently allows the differential diagnosis to be narrowed. The clinical features and usual locations of the most common congenital cystic lesions of the neck are summarized in the Table.

The evaluation of a patient suspected of having a congenital cervical cystic mass should follow an orderly progression. Because most congenital lesions manifest during infancy and early childhood, the patient's age provides important diagnostic information. Congenital cervical cystic lesions are usually slow-growing masses and typically cause symptoms only because of enlargement or infection. A painless soft or fluctuant cervical mass is the first clinical manifestation in most cases. Following physical examination, ultrasonography (US) is usually performed. US helps define the size and extent of the mass, demonstrate its relationship to surrounding normal structures, and confirm the cystic nature of the lesion. Computed tomography (CT) also provides this information and is ideally suited for evaluation of soft-tissue planes adjacent to larger masses that cannot be entirely visualized with US. Moreover, CT is superior for detecting calcification and, when contrast material is administered, the vascularity of lesions. Magnetic resonance (MR) imaging, with its multiplanar capability and superior contrast resolution, demonstrates the full extent of the mass and gives important supplemental information for accurate preoperative planning. This can be especially relevant in cases of extension into the mediastinum or deep spaces of the neck. Furthermore, MR imaging offers superior resolution for evaluating masses located in anatomically complex areas, such as the floor of the mouth.

The purpose of this article is to present the clinical, pathologic, and radiologic features of the most common congenital cystic lesions of the neck, emphasizing their embryologic origin.

■ THYROGLOSSAL DUCT CYST

The thyroid gland begins to develop in the 3rd week of fetal life as a median outgrowth from the floor of the primitive pharynx. This thyroid primordium originates at the level of the foramen cecum, which in the adult lies at the junction of the anterior two-thirds and posterior one-third of the tongue. The primitive thyroid descends in the neck, penetrates through the underlying mesoderm of the tongue and floor of the mouth musculature, and eventually passes anterior to the developing hyoid bone and laryngeal cartilages. The anlage of the gland reaches its final position in the inferior part of the neck by the 7th week of gestation after descending anterior to the thyrohyoid membrane and the strap muscles (sternothyroid and sternohyoid muscles) (1).

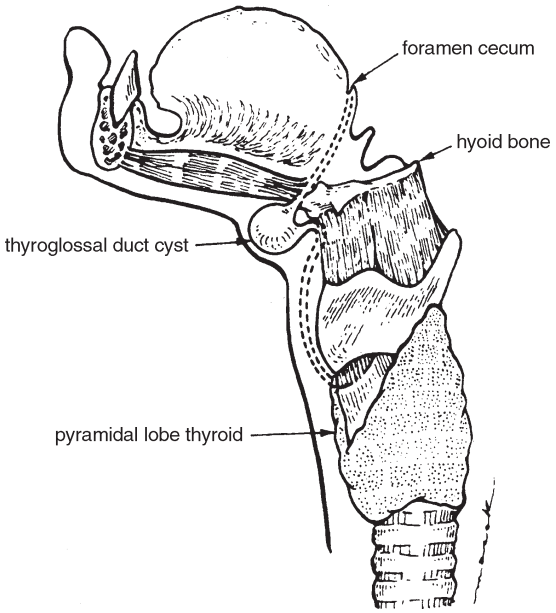


Figure 1. Pathway of the thyroglossal duct. Drawing shows a lateral view of the embryologic thyroglossal duct from the foramen cecum through the developing hyoid bone to the pyramidal lobe of the thyroid. If the duct fails to involute completely, a thyroglossal duct cyst may result, with 80% of these lesions being at or below the level of the hyoid bone. (Reprinted, with permission, from reference 5.)

During its migration, the anlage of the thyroid gland is connected to the tongue by a narrow tubular structure, the thyroglossal duct. This duct normally involutes by the 8th–10th gestational week. The site of the original opening of the thyroglossal duct persists as the foramen cecum of the tongue, with the inferior end of the canal becoming the pyramidal lobe of the thyroid gland (1–3). If any portion of the thyroglossal duct persists, secretions from the epithelial lining (likely secondary to repeated local infection and inflammation) may give rise to cystic lesions (4). The duct is intimately associated with the developing hyoid bone, usually passing through it (Fig 1) (6). As a consequence, most thyroglossal duct cysts are found either at the level of the hyoid bone (15% of cases) or in the strap muscles (65%) immediately inferior.

The thyroglossal duct cyst is the most common congenital neck mass, accounting for 70% of congenital neck anomalies (4), and the second most common benign neck mass, after benign lymphadenopathy (7). About 50% of patients present before 20 years of age, with a second group of patients presenting in young adulthood (5,8). No gender predilection has been reported (4). Rare cases of hereditary thyroglossal duct cysts have been reported; typically, these cysts have an autosomal dominant pattern of transmission and occur in prepubertal girls (9).

● Clinical Characteristics

Thyroglossal duct cysts are located in the midline (75% of cases) or slightly off-midline (25%) in the anterior neck. They are always within 2 cm of the midline (10). Of those in a paramedian location, most will occur on the left for reasons that are not well understood (11). Approximately 80% of the cysts are located either at or below the level of the hyoid bone. The remaining 20% are located above the hyoid bone. Rarely, a thyroglossal duct cyst may manifest as a mass in the floor of the mouth (3,12–15).

Most authorities ascribe inflammation as the most likely cause for a thyroglossal duct cyst. The frequency of infections in childhood may explain the increased prevalence of these cysts in this age group (4).

A thyroglossal duct cyst usually manifests as an enlarging, painless mass in a pediatric or young adult patient. The duct and cyst characteristically move upward with tongue protrusion, a reflection of the origin of the duct at the foramen cecum (6,10,16). The size of the cyst ranges from 0.5 to 6 cm in diameter, but most are between 1.5 and 3 cm (17). Many patients with thyroglossal duct cysts present clinically as a consequence of infection (17). Despite initial improvement with antibiotic therapy, the cyst frequently recurs after the antibiotic course is completed (17).

About 1% of thyroglossal duct abnormalities are associated with thyroid carcinoma arising from ectopic rests of thyroid tissue within the duct and not from the duct itself. Although most (80%) of these tumors are of the papillary type, virtually every type of thyroid malignancy has been reported in association with thyroglossal duct cysts (4,10,18–21). The presence of a carcinoma within a thyroglossal duct cyst is rarely suspected preoperatively (4). Nodal spread is much less common in these thyroglossal duct cyst neoplasms than in primary carcinomas arising in the thyroid gland itself (4).

Simple incision and drainage or partial resection of a thyroglossal duct cyst virtually always leads to recurrence (Fig 2). Therefore, complete

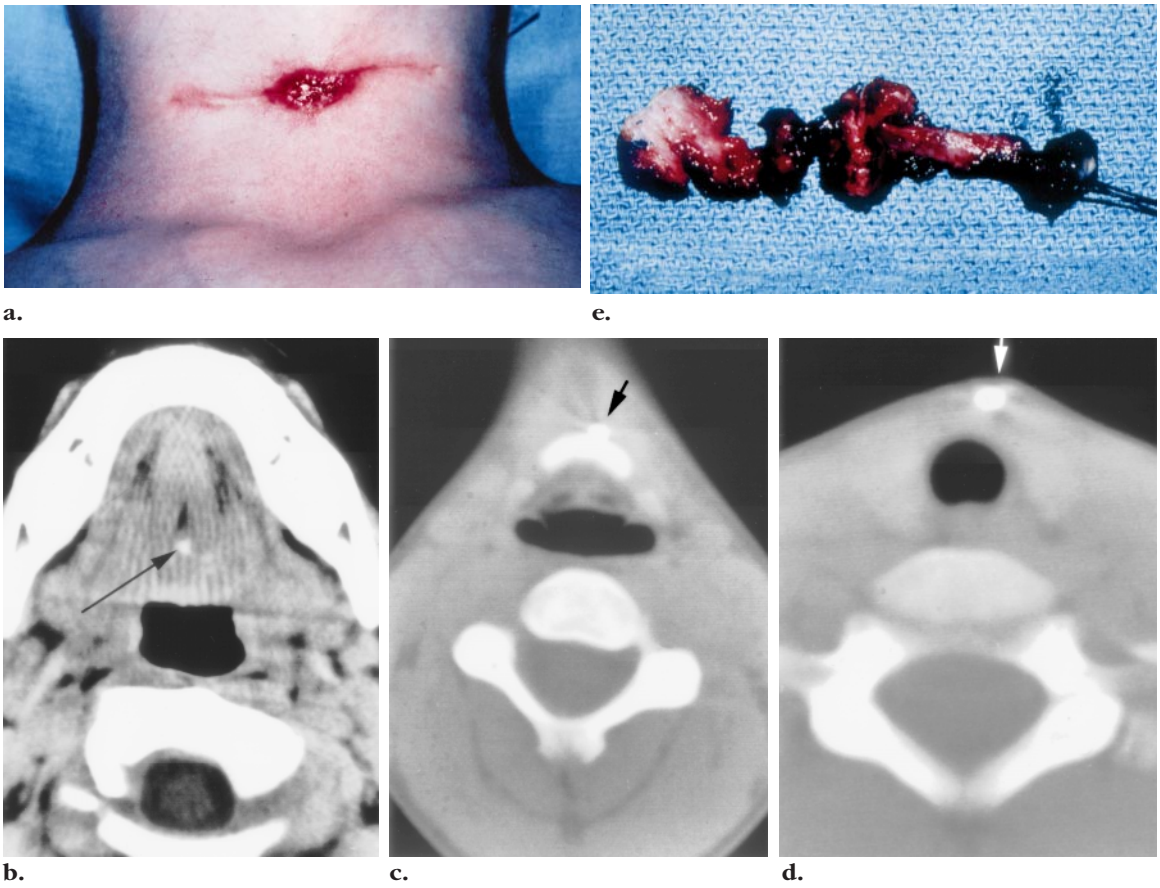


Figure 2. Persistent thyroglossal duct sinus tract in a 5-year-old boy who had undergone two partial resections for a thyroglossal duct cyst in the preceding 9 months. **(a)** Photograph of the lower neck shows a fistulous ostium with a transverse surgical scar on both sides. **(b)** CT scan obtained after ethiodized oil was injected into the fistula shows focal collection of the contrast agent in the region of the foramen cecum (arrow). **(c, d)** CT scans obtained at lower levels show the contrast material–enhanced path of the thyroglossal duct along the anterior surface of the hyoid bone (arrow in **c**) and inferiorly to the skin ostium near the pyramidal lobe of the thyroid (arrow in **d**). **(e)** Photograph of the surgical specimen reveals entire length of the fistulous tract.

excision of the cyst with the Sistrunk procedure is the recommended surgical approach. This operation involves resection of the central portion of the hyoid bone and a core of tissue following the expected course of the thyroglossal duct to the foramen cecum (4). The recurrence rate following this procedure is 2.6%, which is much lower than the recurrence rate when incomplete excision is performed (up to 38%) (15,17).

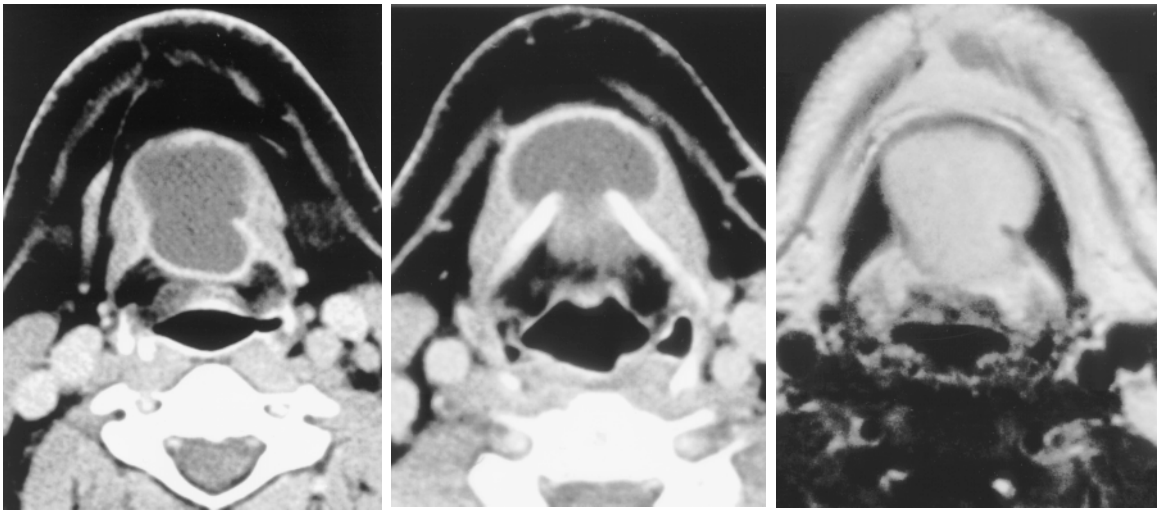
● Pathologic Characteristics

Thyroglossal duct cysts contain a colorless, viscous secretion. At histologic examination, stratified squamous epithelium or ciliated pseudostratified columnar epithelium lining the cyst wall is usually seen (16). Mucous glands may also be present (10). Ectopic thyroid tissue along the course of the duct is variably reported in up to 62% of cases (Fig 3) (19,20,22). LiVolsi et al (22) speculated that the variation in detection of this tissue was a reflection of the degree of meticulous examination by the reviewing pathologist.

● Radiologic Features

On all radiologic images, a thyroglossal duct cyst manifests as a cystlike mass either in the midline of the anterior neck at the level of the hyoid bone or within the strap muscles just off the midline.

At US, the finding of an anechoic mass with a thin outer wall in this characteristic location easily establishes the diagnosis of a thyroglossal duct cyst. However, this "classic" appearance is seen in less than half (42%) of the cases. More commonly, these cysts manifest as hypoechoic



a. b. c.

Figure 4. Thyroglossal duct cyst in a 41-year-old man. **(a, b)** Axial contrast-enhanced CT scans show a cystic mass in the anterior midline of the neck just above **(a)** and at the level of **(b)** the hyoid bone. **(c)** Axial T1-weighted MR image at the thyrohyoid membrane level shows hyperintensity of the mass, a finding suggestive of proteinaceous content or hemorrhage.

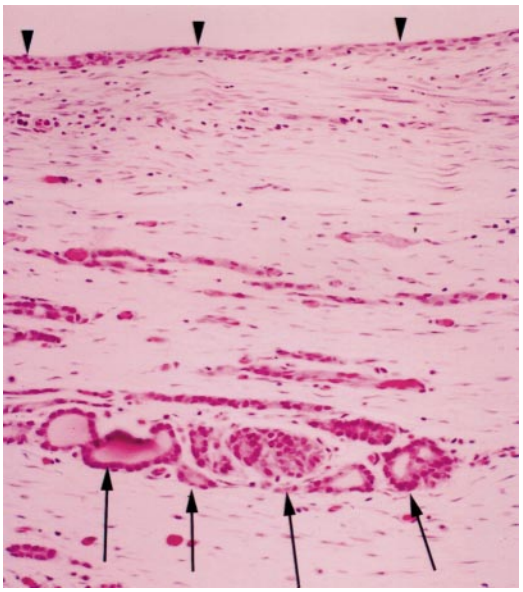


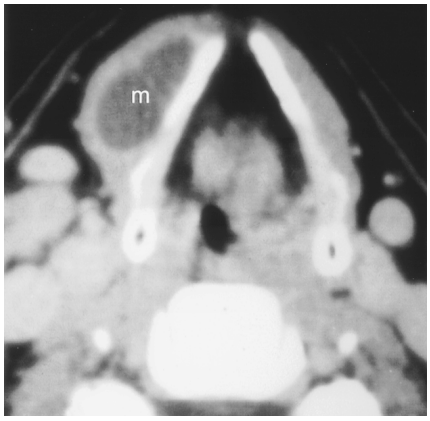
Figure 3. Photomicrograph (original magnification, ×4; hematoxylin-eosin [H-E] stain) of a thyroglossal duct cyst specimen shows squamous epithelium (arrowheads). The cysts may also be lined with respiratory epithelium. Most (about 60%) of these lesions contain some remnant of thyroid tissue (arrows).

masses, often with increased through-transmission. They may be either homogeneous or heterogeneous in appearance with variable degrees of fine to coarse internal echoes. There is no correlation between the sonographic appearance and pathologic evidence of infection and inflammation (23). Heterogeneity seen in thyroglossal duct cysts on sonograms is more likely due to the proteinaceous content of the fluid secreted from the cyst wall rather than to infection (23). Preoperative sonographic visualization of normal thyroid tissue is sufficient to exclude a diagnosis of ectopic thyroid tissue and obviates routine thyroid scintigraphy (24).

On CT scans, a thyroglossal duct cyst usually appears as a smooth, well-circumscribed mass anywhere along the vertical course of the vestigial thyroglossal duct (Figs 4–6). The mass has a thin wall and homogeneous attenuation, the values of which correspond to those of fluid (10–18 HU). Elevated attenuation values of the fluid cyst reflect its increased protein content and generally correlate with a history of prior infections. Although thyroglossal duct cysts are usually unilocular, septations may be seen occasionally. Peripheral rim enhancement is usually observed on contrast-enhanced scans (3).

An uncomplicated thyroglossal duct cyst has low signal intensity on T1-weighted images and is hyperintense on T2-weighted images, findings that reflect its fluid content. The rim will be nonenhancing unless inflammation is present (25). In case of infection or hemorrhage, a thick irregular rim may be visualized, and the signal intensity of the fluid becomes variable from the presence of proteinaceous debris (Fig 4).

Figures 5, 6. **(5)** Thyroglossal duct cyst in a 39-year-old man. Contrast-enhanced CT scan shows a hypoattenuated mass (*m*) in the right strap muscles. (Courtesy of the Department of Radiology, Neuroradiology Section, University of California at San Francisco.) **(6)** Thyroglossal duct cyst in a 71-year-old man with a 1-year history of progressive swelling on the left side of the neck. **(a)** Contrast-enhanced CT scan shows a large cystic mass arising from the left strap muscles. **(b)** Photograph of the cut specimen shows the smooth inner lining and roughened exterior surface of the cyst.



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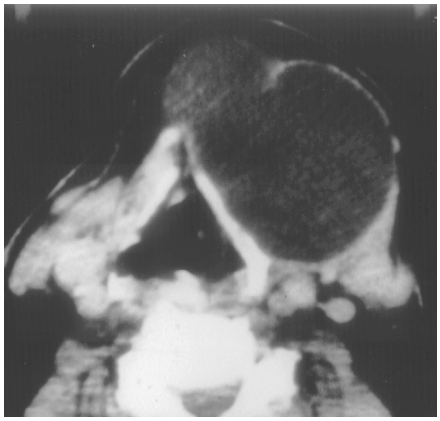
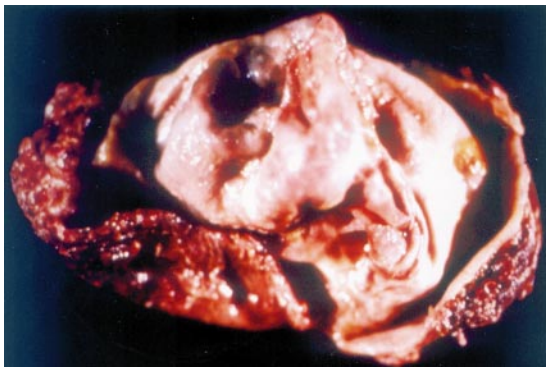


6a.



6b.

■ BRANCHIAL CLEFT ANOMALIES

By the end of the 4th week of embryonic life, the branchial arches (derived from neural crest cells) and the mesenchyme (derived from the lateral mesoderm) are easily recognizable. Five pairs of ectodermal clefts (grooves) and five endodermal branchial pouches separate the six arches, with a closing membrane located at the interface between the pouches and the clefts (1,2).

The first branchial cleft normally gives rise to the eustachian tube, tympanic cavity, and mastoid antrum and contributes to the formation of the tympanic membrane. It is the only cleft to contribute to an adult structure, the external auditory canal. The second, third, and fourth branchial clefts are part of an ectodermally lined depression known as the cervical sinus of His. As the second and fifth branchial clefts merge with each other, this cervical sinus is obliterated. The second branchial pouch, lined by endoderm, gives rise to the palatine tonsil and tonsillar fossa. The third branchial pouch forms the inferior parathyroid gland, thymus, and pyriform sinus; the fourth branchial pouch leads to the formation of the superior parathyroid gland and apex of the pyriform sinus (Fig 7) (1).

The pathogenesis of branchial cleft anomalies remains controversial. Most authorities believe that they arise from incomplete obliteration of the cervical sinus of His or from buried epithelial cell rests (27). They manifest as any combination of sinus, fistula, or cyst. Sinuses almost always open externally on the side of the neck, whereas a fistula manifests as a patent abnormal canal, opening externally on the neck surface and internally within the pharyngeal mucosa (8). In a review of 274 patients with branchial cleft anomalies at the Mayo Clinic, Telander and Deane (8) reported that most of the anomalies (75%) were cysts; 25%, fistulas; and 1%, skin tags and cartilages. Patients with branchial cleft cysts are usually older children or young adults in contrast to patients with fistulas, who are usually infants or young children (8,28). Bilateral branchial cleft anomalies are reported to occur in 2%–3% of cases and are often familial (29).

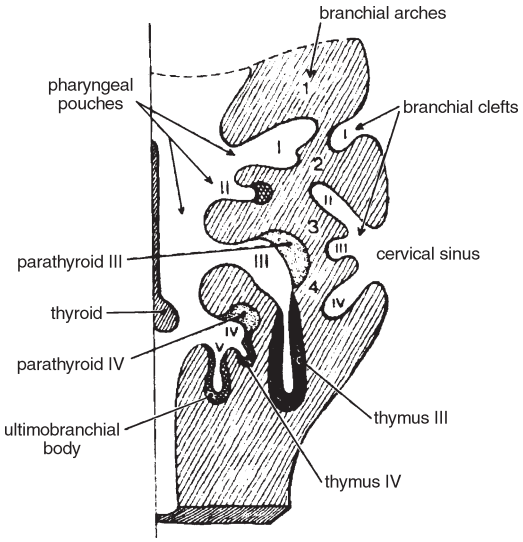


Figure 7. Branchial apparatus. Diagram illustrates the developing branchial apparatus. The branchial clefts (or grooves) are ectodermal lined, whereas the pharyngeal pouches are of endodermal origin. The branchial arches are mesodermal in nature. The second branchial arch begins to overgrow the third and fourth branchial arches to form the cervical sinus of His. In normal development, the sinus is obliterated when the second branchial arch merges with the epicardial ridge. (Reprinted, with permission, from reference 26.)

● First Branchial Cleft Cyst

A first branchial cleft cyst, or parotid lymphoepithelial cyst, arises along the residual embryologic tract of the first branchial cleft or arch extending from the external auditory canal through the parotid gland to the submandibular triangle. These cysts account for only 5%–8% of all branchial cleft defects (30) and are most commonly seen in middle-aged women (31).

Clinical Characteristics.—Cysts of the first branchial cleft usually manifest as recurrent abscesses or other inflammation (sinus tract) either around the ear or at the angle of the mandible. The patient typically has a history of recurrent parotid abscesses unresponsive to antibiotic therapy and drainage. Otorrhea commonly occurs if the cyst drains into the external auditory canal. Occasionally, a sinus tract extending to the hyoid bone may be seen. These cystic anomalies often mimic the clinical characteristics of parotid neoplasms and may even be associated with facial nerve palsy (8). Both children and adults may be affected (32,33). Complete surgical excision is the only curative therapy for these lesions (32,33).

Pathologic Characteristics.—Work (32) divided first branchial cleft cysts into two types based on embryologic criteria. The Work type I cyst is derived from ectoderm and represents a duplication anomaly of the membranous external auditory canal from the first branchial cleft. It is characteristically found medial to the concha of the ear in a parallel course with the external auditory canal but may extend into the retroauricular area. It is lined with squamous epithelium. No skin appendages are seen in Work type I cysts (32,33).

Work type II cysts arise from the first branchial cleft and arch with a possible contribution from the second branchial arch. Consequently, they are derived from ectoderm and mesoderm. They involve both the external auditory canal and cartilaginous pinna. The presence of skin appendages (hair follicles, sweat and sebaceous glands) contained in the squamous epithelial lining of these cysts allows the pathologist to distinguish Work type II cysts from type I cysts. Both types produce keratin from the squamous epithelium lining (32,33).

The first branchial cleft cysts associated with the parotid gland are classified most often as Work type II cysts because both ectodermal and mesodermal elements are present on histologic examination. They are usually distinguished from the more common lymphoepithelial cysts of the parotid gland, which lack mesodermal components (34).

Radiologic Features.—On CT scans, a first branchial cleft cyst appears as a cystic mass either within, superficial to, or deep to the parotid gland. Cyst wall thickness and enhancement are variable and increase with recurrent infections.

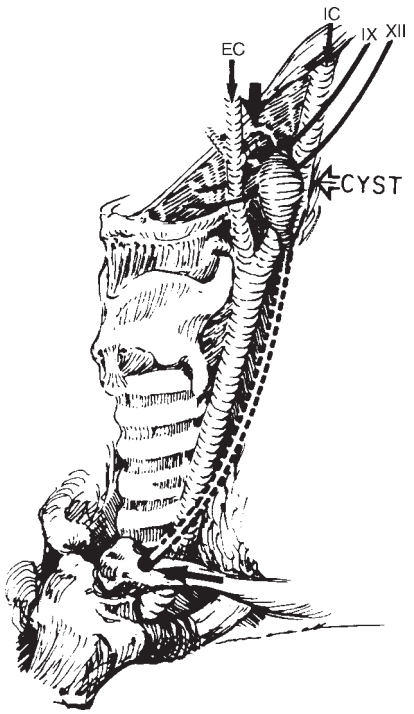


Figure 8. Second branchial cleft tract. Drawing shows the path of second branchial cleft anomalies, which can occur anywhere along a line from the supraclavicular region to the oropharyngeal mucosa. The path travels lateral to the common carotid artery, then heads medially between the external carotid (*EC*) and internal carotid (*IC*) arteries under the glossopharyngeal nerve (*IX*) and above the hypoglossal nerve (*XII*). If the tract continues further along this course, it will enter the parapharyngeal space and pierce the middle constrictor muscle before ending as an opening within the tonsillar fossa. (Reprinted, with permission, from reference 5.)

In most cases, neither the CT nor MR imaging appearance of these cysts is characteristic enough to allow differentiating a first branchial cleft cyst from any other cystic mass of the parotid gland. As with any lesion of the deep margin of the parotid gland, a first branchial cleft cyst in this location may extend into the adjacent fat-containing parapharyngeal space.

● Second Branchial Cleft Cyst

The vast majority (95%) of branchial cleft anomalies arise from the second cleft (14). At least three-fourths of these anomalies are cysts (10), which typically occur between 10 and 40 years of age, in contrast to fistulas or sinuses, which manifest most commonly during the 1st decade of life (21,35). No gender predilection has been reported (31).

Bailey (36) classified second branchial cleft cysts into four types. The Bailey type I cyst is the most superficial and lies along the anterior surface of the sternocleidomastoid muscle, just deep to the platysma muscle. The type II cyst is the most common and found in the "classic" location for these cysts: along the anterior surface of the sternocleidomastoid muscle, lateral to the carotid space, and posterior to the submandibular gland. A type III cyst extends medially between the bifurcation of the internal and external carotid arteries to the lateral pharyngeal wall. The type IV cyst lies in the pharyngeal mucosal space and is lined with columnar epithelium (36). Figure 8 outlines the path of the second branchial cleft tract in the neck.

Clinical Characteristics.—Most second branchial cleft cysts are located in the submandibular space. However, because of the anatomic relationship of the second branchial apparatus and the cervical sinus, they can occur anywhere along a line from the oropharyngeal tonsillar fossa to the supraclavicular region of the neck (30).

These cysts usually appear as painless, fluctuant masses in the lateral portion of the neck adjacent to the anteromedial border of the sternocleidomastoid muscle at the mandibular angle (13,21,37). The mass enlarges slowly over time and may become painful and tender if secondarily infected (21). In a young patient, a history of recurrent inflammation in the region of the mandibular angle is highly suggestive of a sec-

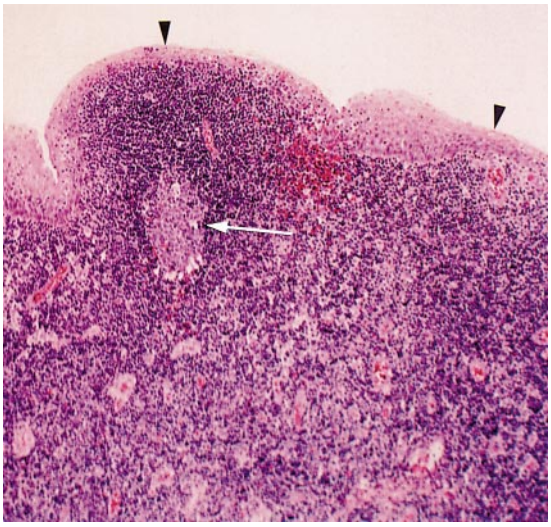


Figure 9. Photomicrograph (original magnification, ×10; H-E stain) of a branchial cleft cyst specimen shows lining of stratified squamous epithelium (arrowheads). The cyst wall usually contains a diffuse or nodular lymphoid component, often with germinal centers (arrow). The absence of sinusoids and a subcapsular sinus distinguishes this from a lymph node.

ond branchial cleft cyst. If a fistula is present, the ostium is usually noted at birth just above the clavicle in the anterior neck (5).

Surgical excision is the recommended therapy for a branchial cleft anomaly because of the increased frequency of secondary infection (8).

Pathologic Characteristics.—Second branchial cleft cysts range in size from 1 to 10 cm (10,17). They are usually filled with a turbid, yellowish fluid that may contain cholesterol crystals. Their walls are thin and lined with stratified squamous epithelium overlying lymphoid tissue (17,21,37). Columnar respiratory epithelium is occasionally present (Fig 9).

Histologic specimens treated with immunohistochemical stains show mural lymphatic sinuses with the mantle zones of the follicles always aligned toward the epithelium. This orientation is analogous to that of the normal crypt epithelium of the palatine tonsils and marginal sinuses in lymph nodes (38).

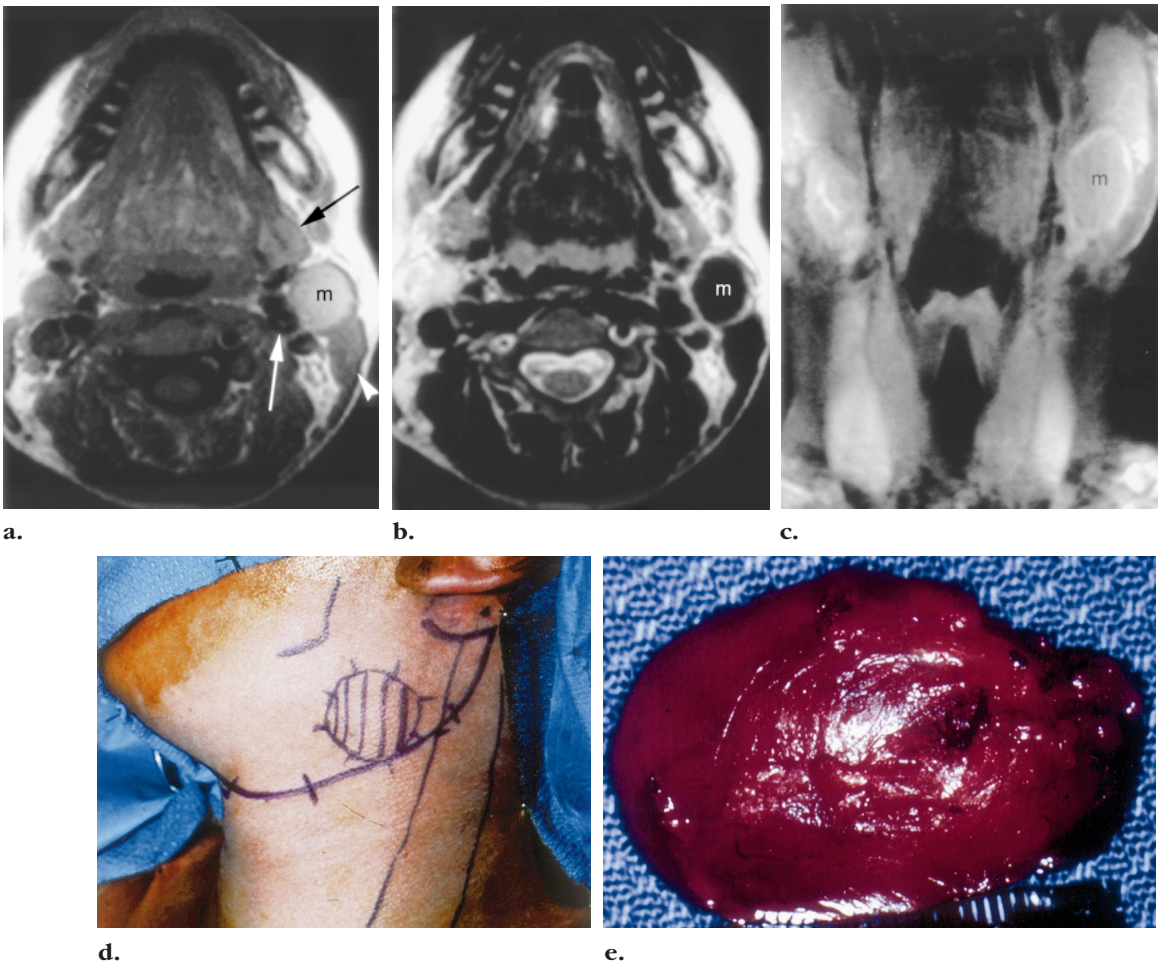
Radiologic Features.—At US, a second branchial cleft cyst is seen as a sharply marginated, round to ovoid, centrally anechoic mass with a thin peripheral wall that displaces the surrounding soft tissues. The mass is compressible and shows distinct acoustic enhancement. Occasionally, fine, indistinct internal echoes, representing debris, may be seen.

At CT, these cysts are typically well-circumscribed, homogeneously hypoattenuated masses surrounded by a uniformly thin wall (37). The mural thickness may increase after infection. The "classic" location of these cysts (at either CT or MR imaging) is at the anteromedial border of the sternocleidomastoid muscle, lateral to the carotid space, and at the posterior margin of the submandibular gland. The cyst typically displaces the sternocleidomastoid muscle posteriorly or posterolaterally, pushes the vessels of the carotid space medially or posteromedially, and displaces the submandibular gland anteriorly (Figs 10, 11) (13,30). It may also be seen more medially within the parapharyngeal space after extending through the stylomandibular tunnel and middle constrictor muscle (Fig 12) (1,39).

MR imaging better depicts the deep tissue extent of a second branchial cleft cyst, which allows accurate preoperative planning. The cyst fluid varies from hypointense to slightly hyperintense relative to muscle on T1-weighted images and is usually hyperintense on T2-weighted images (31). Mural thickness and enhancement vary, depending of the presence and severity of any associated inflammatory process (Fig 11) (30).

Occasionally, a "beak sign" may be seen on axial CT or MR images. This sign represents a curved rim of tissue or "beak" pointing medially between the internal and external carotid arteries. It is considered a pathognomonic imaging feature of a second branchial cleft cyst, specifically a Bailey type III cyst (5).

Figure 10. Second branchial cleft cyst in a 30-year-old woman with a 14-month history of a mass in the left side of the neck that was unresponsive to antibiotics and that enlarged somewhat the month before surgery. **(a)** Axial T1-weighted image shows a well-defined mass (*m*) along the anterior border of the left sternocleidomastoid muscle (arrowhead), lateral to the carotid space (white arrow), and posterior to the submandibular gland (black arrow)—the classic location for a second branchial cleft cyst. Increased signal intensity of the mass is due to either proteinaceous debris or prior hemorrhage. **(b)** Axial T2-weighted image reveals moderate to marked hypointensity of the mass (*m*), consistent with accumulation of proteinaceous debris or hemorrhage. **(c)** Coronal contrast-enhanced T1-weighted image with fat suppression shows mild rim enhancement of the mass (*m*). **(d)** Intraoperative photograph shows the mass and sternocleidomastoid muscle outlined in ink on the skin surface. **(e)** Photograph of the surgical specimen shows the well-circumscribed mass with mildly lobulated contours.



a. **b.** **c.**

d. **e.**

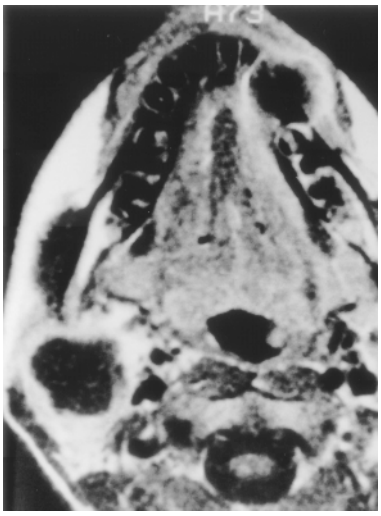
■ THIRD AND FOURTH BRANCHIAL CLEFT CYSTS

Cysts arising from the third and fourth branchial clefts are extremely rare, with only a few cases reported (27,40). These lesions have been described in children and young adults (6). Despite their rarity, third branchial cleft cysts constitute the second most common congenital lesion of the posterior cervical space of the neck after cystic hygroma (41). Anomalies of the fourth branchial cleft usually manifest as a sinus tract rather than a cyst or fistula. The vast majority of these lesions occur on the left side; of 31 cases reported, 28 have been on the left (40).

By definition, a third branchial cleft cyst must lie posterior to the common or internal carotid artery, between the hypoglossal nerve below and the glossopharyngeal nerve above. If the lesion is a complete fistula, it will pierce the thyrohyoid membrane in its course to the pyriform sinus. Most of these cysts are located in the posterior cervical space posterior to the sternocleidomastoid muscle (27).

A fourth branchial cleft sinus tract, following its embryologic derivation, arises from the pyriform sinus, pierces the thyrohyoid membrane, and begins a descent into the mediastinum, following the path of the tracheoesophageal groove. If the tract is long enough, a left-sided lesion will eventually travel under the aortic arch (or,

Figures 11, 12. **(11)** Second branchial cleft cyst in an 11-year-old boy with a mass in the right side of the neck and a history of recurrent inflammation. **(a)** Axial gadolinium-enhanced T1-weighted MR image shows a hypointense mass in the right side of the neck in the classic location for a second branchial cleft cyst. Irregular inner border of the mass suggests an associated inflammatory process. **(b)** Coronal T1-weighted MR image shows the mass at the inferior margin of the right parotid gland and lateral to carotid vessels. **(c)** Photograph of the resected specimen shows the thick capsule of the cystic lesion filled with keratinaceous debris. **(12)** Second branchial cleft cyst in a 29-year-old woman with a 1-month history of dysphagia, otalgia of the right ear, and swelling of the right side of the neck. Contrast-enhanced CT scan shows a well-defined, right-sided neck mass (*m*) deep to the carotid space and with medial extension toward the oropharynx.



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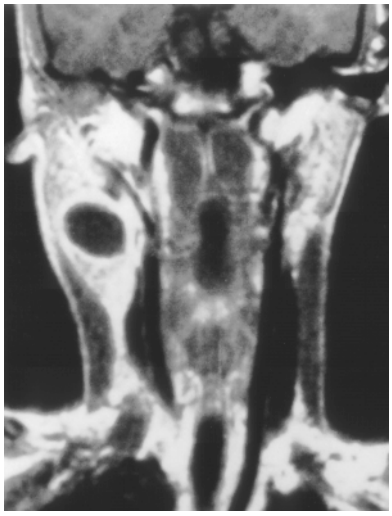

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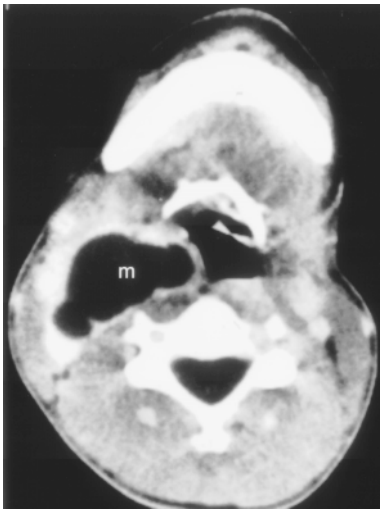

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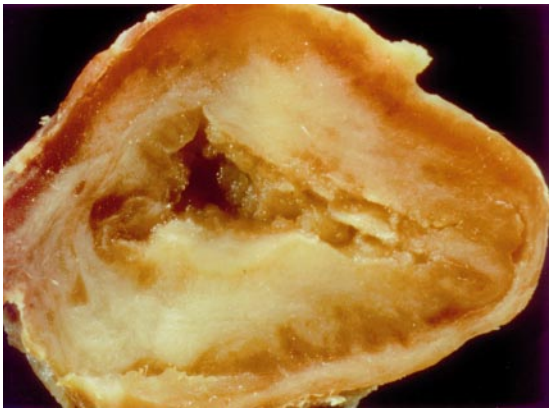


11c.

in the rare case of a right-sided lesion, under the right subclavian artery) before ascending into the neck along the ventral surface of the common carotid artery (27). Most of these lesions are short and thus sinus tracts. Presumably because of the length of this branchial cleft, a complete fourth branchial cleft fistula has not been reported (40).

Distinguishing third branchial cleft anomalies from fourth branchial cleft anomalies may be difficult, since both have relationships with the pyriform sinus. The difference between the two lesions lies in their relationships to the superior laryngeal nerve. Those above this structure are of third branchial cleft origin, whereas those below are derived from the fourth branchial cleft (40).

An ectopic or undescended parathyroid gland may be associated with either lesion, since the parathyroid glands arise from the third and fourth branchial pouches (1). Not surprisingly, parathyroid adenomas have also been reported in association with branchial cleft cysts (42).

Clinical Characteristics.—As with other branchial cleft cysts, a third branchial cleft cyst usually manifests as a painless, fluctuant mass in the posterior triangle area of the neck. Like a cystic hygroma (lymphangioma), the mass often distends during a viral infection of the upper respiratory tract. Surgical excision is the recommended therapy of choice for these lesions because of the increased frequency of secondary infection (8).

Radiologic Characteristics.—A third branchial cleft cyst most commonly appears as a unilocular cystic mass centered in the posterior cervical space on CT and MR images. As with

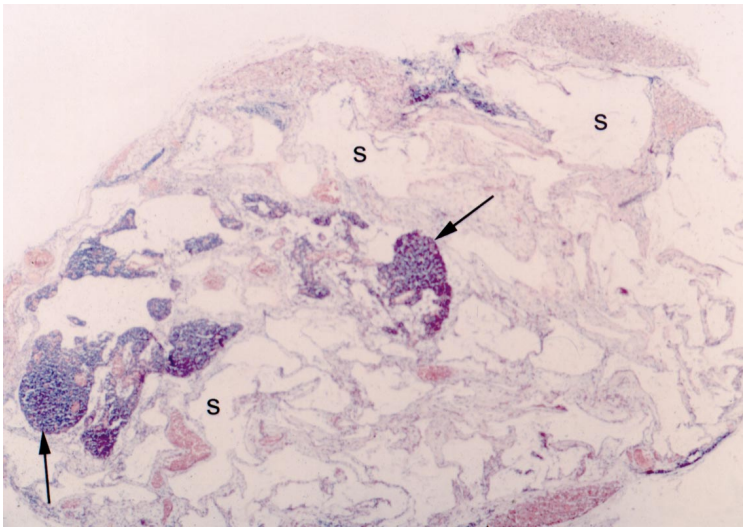


Figure 13. Photomicrograph (original magnification, ×1; H-E stain) of a cystic hygroma specimen shows multiple endothelium-lined vascular spaces. Several of the larger spaces (*S*) are indicated. The stroma includes fibrous tissue, adipose tissue, and aggregates of lymphoid cells (arrows).

other branchial cleft cysts, the cyst fluid may vary in signal intensity on T1-weighted images depending on the protein concentration and is typically hyperintense relative to muscle on T2-weighted images (31). Benson et al (27) reported a fourth branchial cleft cyst that connected with the pyriform sinus and appeared similar to an external or mixed laryngocele.

■ CYSTIC HYGROMA

A cystic hygroma is the most common form of lymphangioma and constitutes about 5% of all benign tumors of infancy and childhood (41). The overwhelming majority (about 80%–90%) are detected by the time the patient is 2 years old, the age of greatest lymphatic growth (21,43). Only a few cases have been reported in adults (5,44). No gender predilection has been reported (45).

Cystic hygroma is thought to arise from an early sequestration of embryonic lymphatic channels, as suggested by Dowd (46) in 1913 and expanded on by Goetch in 1938 (47). This sequestration apparently occurs more commonly in the developing jugular lymph sac pair than in the other four embryonic sites of the lymphatic system. From this location, the sequestered site follows the path of the surrounding mesenchyme destined for either the neck or the developing mediastinum. This accounts for the propensity of these lesions to occur in the lower neck, axilla, and upper mediastinum (48). Alternatively, a cystic hygroma may arise from a failure of the juguloaxillary lymphatic sac to drain into the internal jugular vein, producing a congenital obstruction of lymphatic drainage (7,25,49). Some authors have proposed that involution of a cystic hygroma in utero produces the "web neck" of Turner syndrome (45,50).

● Clinical Characteristics

Approximately 75%–80% of all cystic hygromas involve the neck and the lower portion of the face. In children, the most common location is the posterior cervical space, followed by the oral cavity. In adults, cystic hygromas are more commonly seen in the sublingual, submandibular, and parotid spaces (5,44). Other reported locations include the axilla (20% of cases), the mediastinum (5%), the abdominal cavity (colon, spleen, and liver), the retroperitoneum (kidneys), the scrotum, and even the skeleton (37,45).

These lesions are characteristically infiltrative in nature and do not respect fascial planes. Consequently, they may extend inferiorly from the

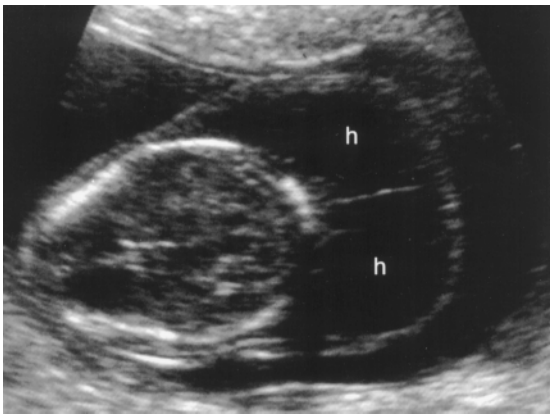


Figure 14. Cystic hygroma. Intrauterine sonogram, obtained at 14 weeks gestation to evaluate vaginal bleeding, shows a large, multiseptated, anechoic mass (*h*) of the posterior neck.

posterior cervical triangle into the axilla and mediastinum or anteriorly into the floor of the mouth and the tongue (41). If the mass is very large, it may extend across the midline (37,43). Only 3%–10% of cervical cystic hygromas are associated with extension into the mediastinum (21,43).

The majority of cystic hygromas are clinically asymptomatic and manifest as painless, soft or semifirm masses in the neck. Size is extremely variable. Very large masses may compromise the airway by extrinsic pressure resulting in death. To avoid this outcome, early tracheostomy is required (5). Although these masses usually grow slowly, they may suddenly increase in size secondary to hemorrhage or trauma or because of a viral infection when large amounts of lymphatic fluid are produced from the lymphoid follicles in the cyst wall (51). Other reported clinical manifestations include facial nerve paralysis, dysphagia, or other feeding problems (31). Chylothorax and chylopericardium may occur as complications of mediastinal involvement.

● Pathologic Characteristics

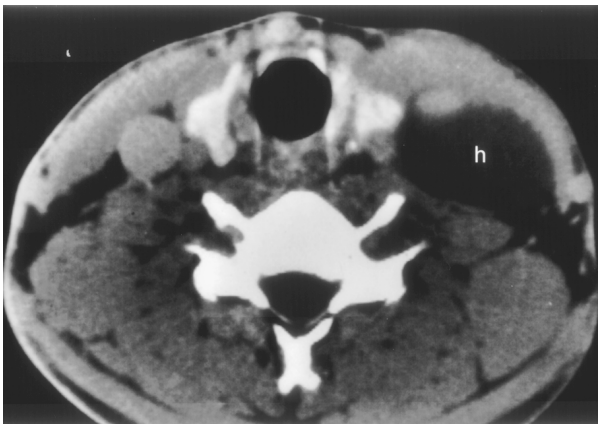
Cystic hygromas are composed of multiple dilated cystic spaces separated by minimal intervening stroma. The cysts vary in diameter from a few millimeters to more than 10 cm, and most contain chylous fluid.

These thin-walled spaces are lined by endothelial cells and supporting connective tissue stroma (Fig 13) (10,37). Lesions may extend into adjacent soft tissues, invade into muscle, and surround vascular structures.

Currently, it is believed that cystic hygromas and the other three types of lymphangioma (cavernous lymphangioma, capillary or simple lymphangioma, and vasculolymphatic malformation) are manifestations of the same disease process. This theory is supported by the frequent observation of combinations of the four types within a single lesion at histologic examination. The differences in gross morphologic features of these four types may be explained by the nature of the stroma around the malformation. If loose connective tissue surrounds the lymphatic anomaly, a cystic hygroma is more likely to be seen. Conversely, if the surrounding tissue is firmer in texture, the other types of lymphangioma should be seen (45,52).

● Radiologic Features

On US scans, most cystic hygromas manifest as a multilocular predominantly cystic mass with septa of variable thickness (53). The echogenic portions of the lesion correlate with clusters of small, abnormal lymphatic channels (53). Fluid-fluid levels can be observed with a characteristic echogenic, hemorrhagic component layering in the dependent portion of the lesion (54). Prenatal US may demonstrate a cystic hygroma in the posterior neck soft tissues (Fig 14) (50).



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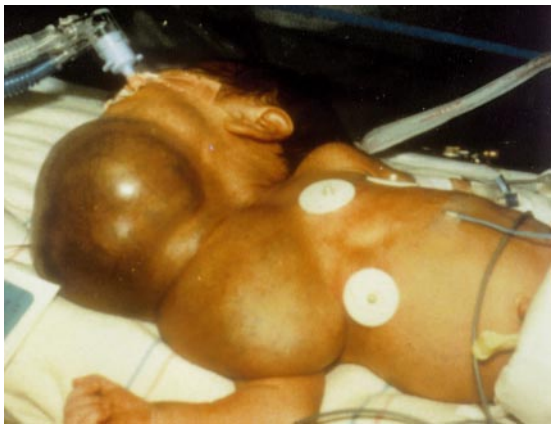

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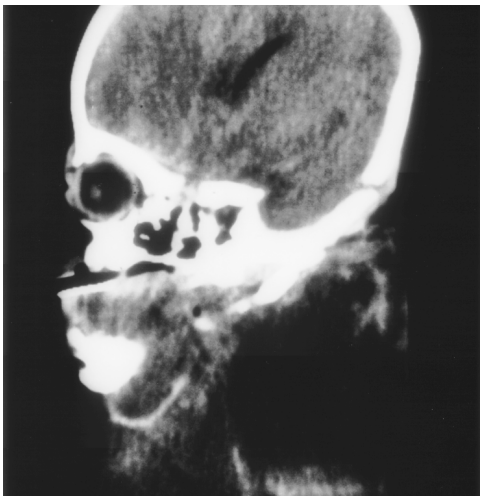


16b.

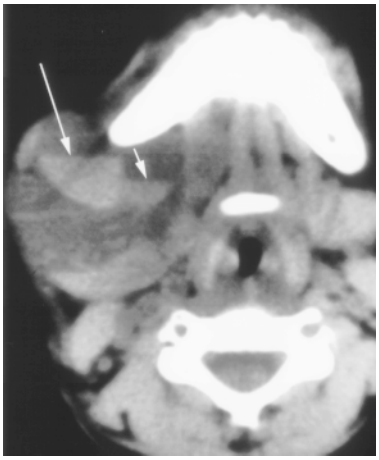
Figures 15, 16. **(15)** Cystic hygroma in a 28-year-old man with a 4-week history of painless swelling of the left side of the neck unresponsive to antibiotics. Fine-needle aspiration yielded serous fluid. Contrast-enhanced CT scan shows a hypoattenuated mass (*h*) within the posterior cervical space deep to the sternocleidomastoid muscle. At surgery, the mass was adherent to the internal jugular vein. (Courtesy of the Department of Radiology, Naval Medical Center, San Diego, Calif.) **(16)** Cystic hygroma in a newborn boy. **(a)** Photograph shows a large bilobulated mass extending from the neck into the chest wall, axilla, and lower face. **(b)** Sagittal CT scan shows erosion of the mandible and invasion of the oral cavity. Multiple septa are seen.

On CT images, cystic hygromas tend to appear as poorly circumscribed, multiloculated, hypoattenuated masses. They typically have characteristic homogeneous fluid attenuation (Figs 15, 16) (21,43). Infected lesions or those that have been in the past commonly show higher attenuation than that seen in simple fluid. Usually, the mass is centered in the posterior triangle or in the submandibular space (Figs 15, 17, 18). It is not uncommon for some of these lesions to extend from one space in the neck into another as a result of their infiltrative nature (Figs 17, 19) (21).

The relationship of a cystic hygroma to adjacent soft tissues of the neck is best demonstrated with MR imaging. The most common pattern is that of a mass with low or intermediate signal intensity on T1-weighted images and hyperintensity on T2-weighted images (Figs 18, 19). Infrequently, this lesion may be hyperintense on T1-weighted images, a finding associated with clotted blood or high lipid (chyle) content. In the case of hemorrhage, fluid-fluid levels may be observed (Fig 17) (45).

■ DERMOID AND EPIDERMOID CYSTS

Unfortunately, the term *dermoid cyst* has been applied to several different pathologic entities, including benign cystic teratomas ("true" dermoid cysts) and various sequestration-type cysts. True dermoid cysts, epidermoid cysts, and teratoid cysts compose the spectrum of teratomas, which are defined as neoplasms whose tissue is foreign to that part of the body from which the tumor arises. All three types of cysts are covered by squamous epithelium. The essential difference between a dermoid cyst and an epidermoid cyst lies in the presence of skin appendages (eg, sebaceous glands, hair follicles) within the wall of the dermoid cyst and the absence of these features in the epidermoid cyst. A teratoid cyst may also contain tissue of other major organ systems (eg, nervous, gastrointestinal, respiratory). Because of the squamous epithelium lining, all three of these cysts may have cheesy keratinaceous material within the lumen (55).



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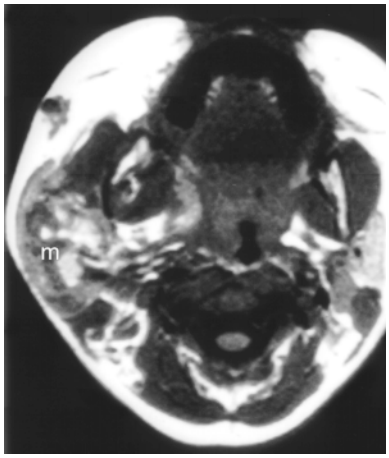

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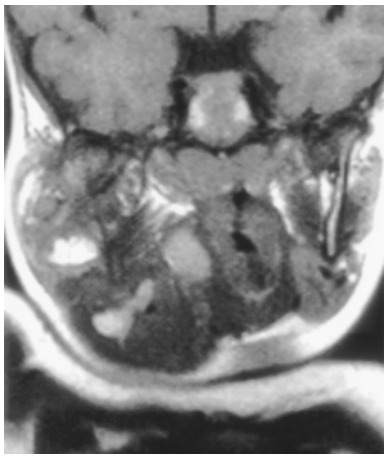

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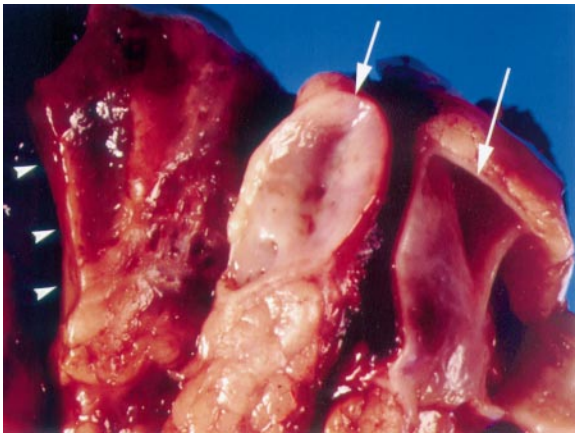

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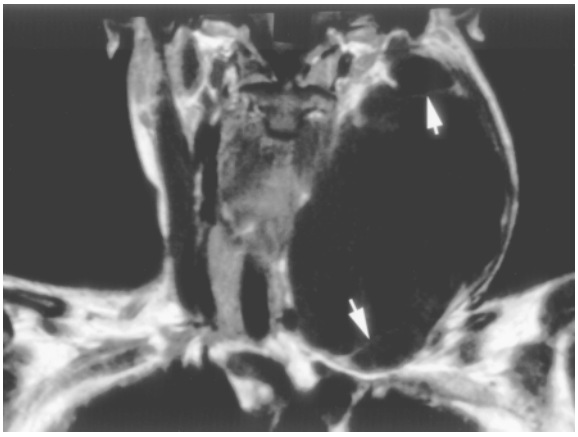

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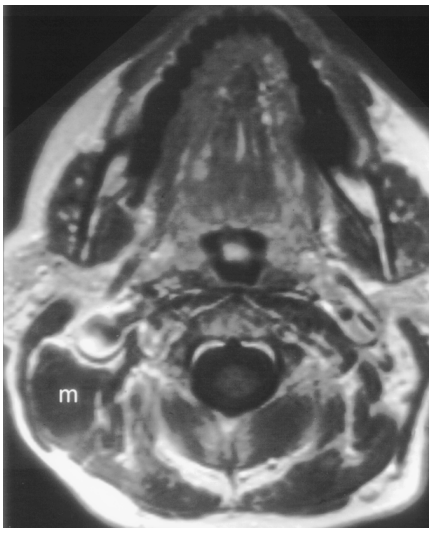


18.

Figures 17–19. **(17)** Cystic hygroma in a 20-month-old girl with swelling under the right jaw and neck noted by the mother. **(a)** Contrast-enhanced CT scan shows a large right-sided neck mass with fluid-fluid levels (arrows) indicative of recent hemorrhage. **(b)** Axial T1-weighted image shows heterogeneous signal intensity within the mass (*m*), which fills the right parotid space and portions of the mandibular space. Areas of hyperintensity correspond to regions of hemorrhage. **(c)** Coronal T1-weighted image shows extension of the mass into submandibular and sublingual spaces. **(d)** Photograph of the specimen shows the cystic lobules (arrows) of the multiloculated mass; cut surface (arrowheads) of a lobule reveals hemorrhage. **(18)** Cystic hygroma. Axial T1-weighted image shows a well-defined mass (*m*) within the right posterior cervical space that displaces the adjacent sternocleidomastoid muscle. **(19)** Cystic hygroma in a 36-year-old woman with a left-sided neck mass that enlarged during viral upper respiratory infections. Coronal T1-weighted image shows a large hypointense mass in the left side of the neck extending from the submandibular space to the thoracic inlet. Multiple septa (arrows) cross the lesion.

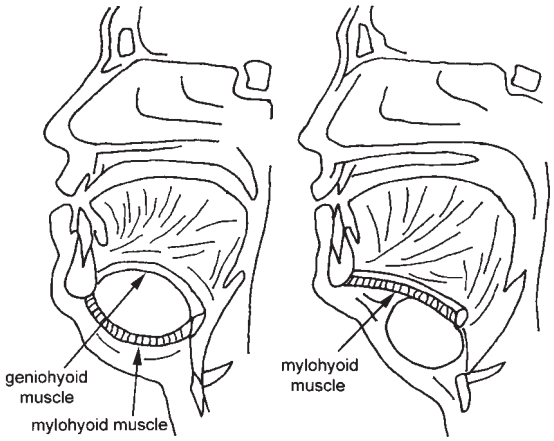


Figure 20. Location of floor-of-mouth lesions with respect to the mylohyoid muscle. (Reprinted, with permission, from reference 25.) **(a)** Diagram illustrates how a mass above the mylohyoid muscle can be resected with an intraoral approach. **(b)** Diagram illustrates how resection of a mass below the mylohyoid muscle requires an external neck approach, usually through a transverse incision.

Dermoid cysts usually manifest during the 2nd and 3rd decades of life. There is no gender predilection (21,56). Only 7% of dermoid inclusion cysts occur in the head and neck, with the lateral eyebrow being the most common location (57,58). About 11.5% of dermoid cysts of the head and neck are in the floor of the mouth, the second most common location (57).

Epidermoid cysts of the neck are rare congenital lesions and are much less common than dermoid cysts in the head and neck. They appear earlier than dermoid cysts, with most lesions evident during infancy (37).

• Clinical Characteristics

The most common clinical appearance of a dermoid cyst in the neck is a midline, suprahyoid, slowly growing mass (37). Occasionally, rapid growth may occur secondary to a sudden increase in the amount of desquamation, pregnancy, or association with a sinus tract (37). Typically, the mass is soft, mobile, and unattached to overlying skin. Unlike thyroglossal duct cysts, they have no intimate association with the hyoid bone and therefore do not move with tongue protrusion (7). The size of the cyst ranges from a few millimeters to 12 cm.

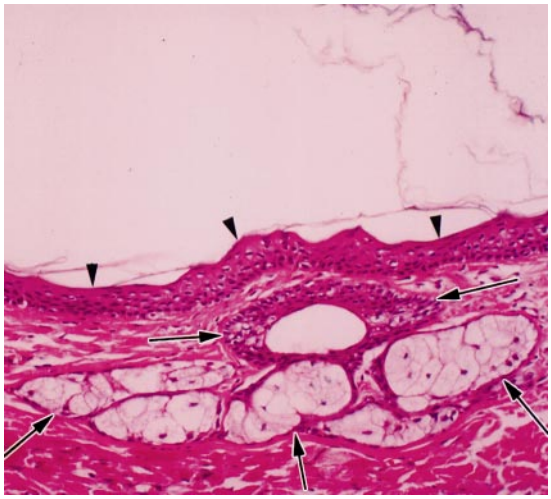


Figure 21. Photomicrograph (original magnification, ×1; H-E stain) of a dermoid cyst specimen shows epidermal appendages, such as the pilosebaceous units (arrows), in association with an epidermal lining (arrowheads).

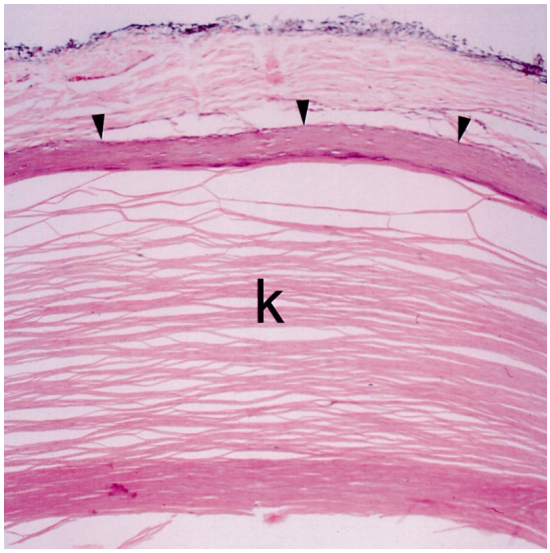
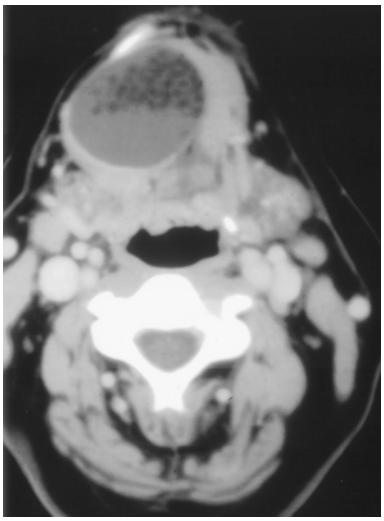


Figure 22. Photomicrograph (original magnification, ×1; H-E stain) of an epidermal inclusion cyst specimen shows its lining of stratified squamous epithelium (arrowheads). The cyst is filled with laminated keratinaceous material (*k*).

The most common cervical location of a dermoid cyst is the floor of the mouth. In rare cases, it may arise within the tongue or from the palate. Clinical signs and surgical approach are determined by the relationship of the cyst to the musculature of the floor of the mouth. If the mass

Figure 23. Dermoid cyst in a 35-year-old woman with a 1-week history of neck swelling. **(a)** Axial contrast-enhanced CT scan shows a well-defined mass in the submandibular-submental region with multiple discrete foci of hypoattenuation in the nondependent portion of the cyst. **(b)** Coronal CT scan shows the mass inferior to the mylohyoid muscle (arrowheads). **(c)** Photograph of the opened cyst shows multiple spherical masses floating in fluid contents. Scale is in centimeters.



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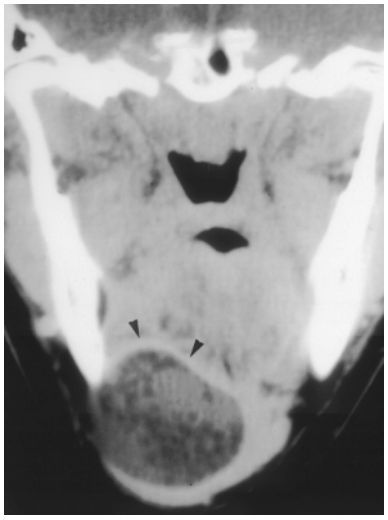

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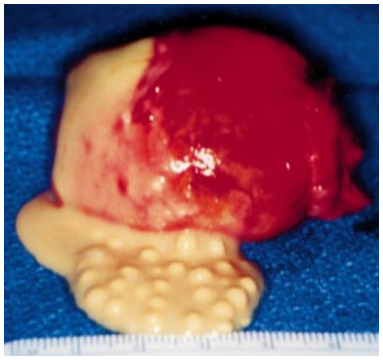


c.

is superficially located (inferior to the mylohyoid muscles), there is usually an obvious submental swelling and the cyst is externally palpable. If the mass is located between the geniohyoid and mylohyoid muscles in the sublingual space, only a small external swelling is seen (25,56). This information is crucial for optimal preoperative planning (Fig 20). About 5% of dermoid cysts undergo malignant degeneration into squamous cell carcinoma (37).

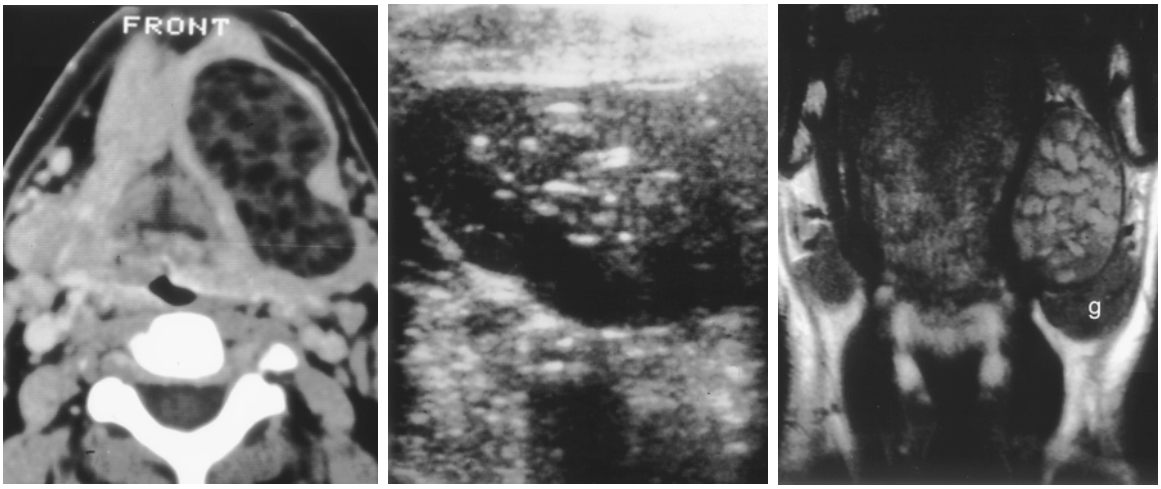
• Pathologic Characteristics

Dermoid cysts are circumscribed, encapsulated lesions. They are lined with ectodermally derived squamous epithelium that contains a variable number of skin appendages (sebaceous glands, hair follicles, and sweat glands). Consequently, the lumen of the cyst is filled with a mixture of keratin, sebaceous material, and occasionally hair (Fig 21) (59). Curiously, dermoid cysts of the floor of the mouth are remarkably free of hair compared with dermoid cysts elsewhere (57). In contrast, the "epidermal inclusion cyst" lacks the dermal appendages seen in dermoid cysts but is otherwise similar in histologic appearance (Fig 22).

• Radiologic Features

Dermoid cysts appear as moderately thin-walled, unilocular masses, located in the submandibular or sublingual space. On CT scans, the central cavity is usually filled with a homogeneous, hypoattenuating (0–18 HU) fluid material. It may appear to be filled with "marbles," due to the coalescence of fat into small nodules within the fluid matrix (55). This "sack-of-marbles" appearance is virtually pathognomonic for a dermoid cyst in this location (Figs 23, 24). Alternatively, the cyst may be heterogeneous on CT scans because of the various germinal components (37). Fluid-fluid levels with supernatant lipid are possible. The rim of these cysts often enhances following administration of contrast material (37).

Figure 24. Dermoid cyst. **(a)** Axial contrast-enhanced CT scan shows a well-defined mass in the submandibular space with a sack-of-marbles appearance. **(b)** US scan shows the mass with multiple echogenic foci and shadowing. **(c)** Coronal T1-weighted image shows the discrete intracystic foci, which have moderate hyperintensity. The mass displaces the left submandibular gland (*g*) inferiorly.



a. **b.** **c.**

MR imaging depicts the topographic relationship of these cysts to the mylohyoid muscle in the floor of the mouth and helps determine the surgical approach. Most dermoid cysts are located superior to the mylohyoid muscle and will be removed with an intraoral approach. Less commonly, the lesion is inferior to this muscle and will be removed with an external submandibular approach (37). The coronal plane is optimal for determining the location of the mass with respect to this muscle pair (25). Dermoid cysts have variable signal intensity on T1-weighted images. They may be hyperintense (because of the presence of sebaceous lipid) or isointense relative to muscle on T1-weighted images. They are usually hyperintense on T2-weighted images. The mass has a clearly demarcated rim but frequently has a heterogeneous internal appearance (Fig 24) (25,56).

Epidermoid cysts have fluid attenuation on CT scans and are hypointense on T1-weighted images and hyperintense on T2-weighted images, following the signal intensity of fluid (Fig 25). An epidermoid cyst located entirely within the sublingual space may be difficult to distinguish from other cystic lesions in the floor of mouth (eg, a simple ranula) on the basis of imaging criteria alone (60).

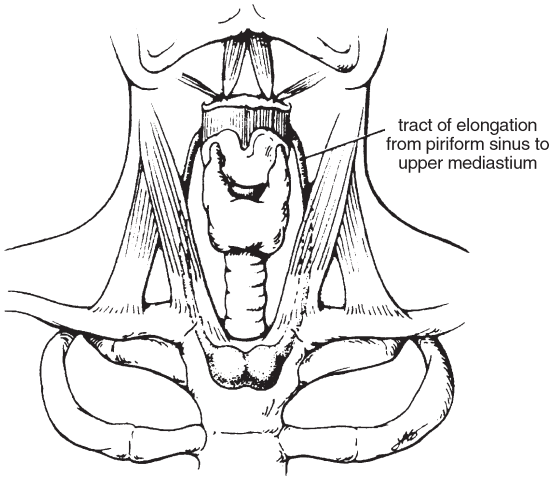
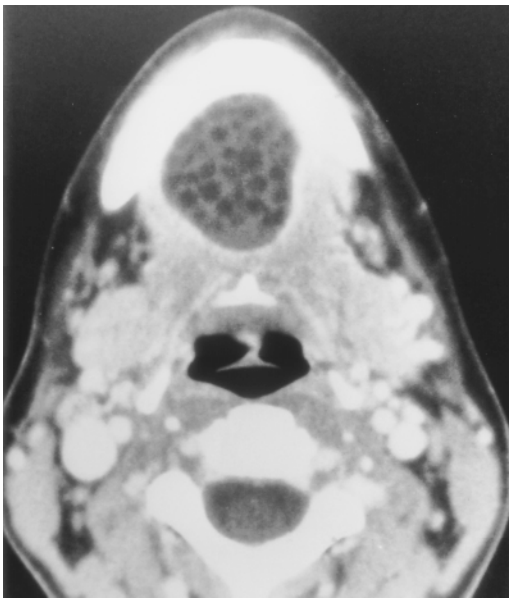


Figure 26. Pathway of the thymopharyngeal duct. Drawing shows a frontal view of the neck. The thymus and parathyroid glands arise from the third and fourth pharyngeal pouches. The thymopharyngeal duct arises from the developing pyriform sinus and descends into the mediastinum, traveling lateral to the thyroid gland. Cervical thymic cysts, among other congenital anomalies, arise along this tract. (Reprinted, with permission, from reference 26.)

■ THYMIC CYST

The pathogenesis of cervical thymic cysts remains controversial despite nearly 100 years of investigation. Among several theories proposed over the years, two mechanisms (congenital and acquired) remain as the most likely routes of

Figure 25. Epidermoid inclusion cyst in a 30-year-old woman. **(a)** Axial contrast-enhanced CT scan shows an encapsulated, hypoattenuated mass with partial adipose content. The coalescence of the lipid material produces a sack-of-marbles appearance. **(b)** Photograph of the cut specimen shows the mass is filled with homogeneous, sebaceous material.



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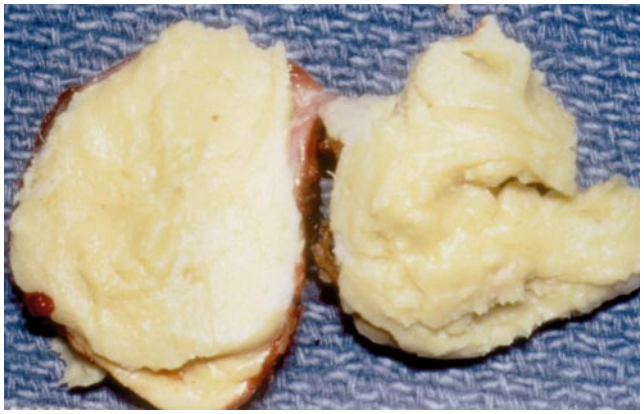


b.

pathogenesis. Most authorities favor the congenital persistence of the thymopharyngeal duct remnants as the cause of these lesions (Fig 26). Alternatively, other investigators believe that they result from acquired, progressive cystic degeneration of thymic (Hassall) corpuscles and the epithelium reticulum of the thymus (26,61). Whatever the cause, the presence of thymic tissue within the lesion is required for pathologic diagnosis.

• Clinical Characteristics

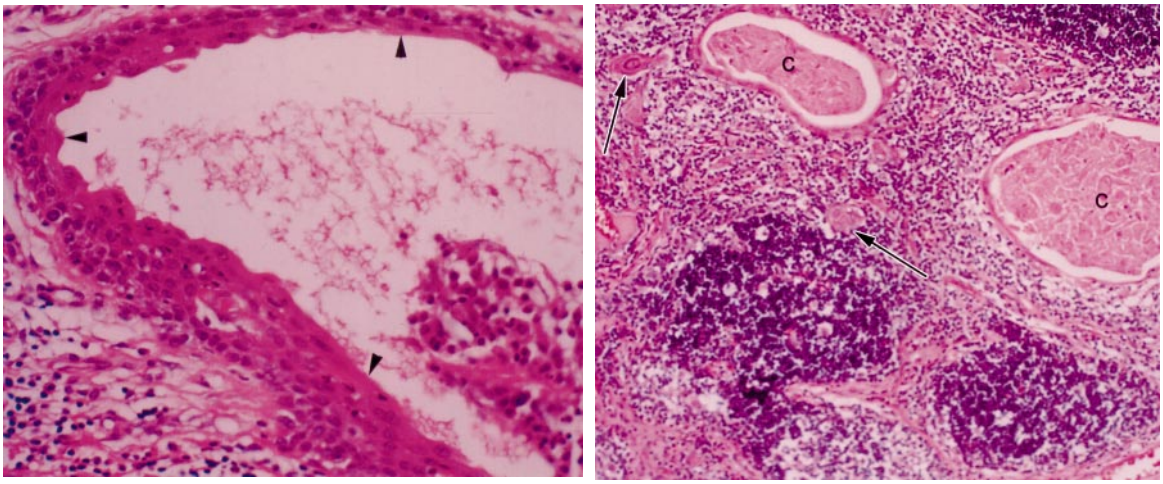
Cervical thymic cysts are very uncommon lesions, with two-thirds of the lesions detected in the 1st decade of life (the age of maximal thymic activity and size) and the remaining one-third in the 2nd and 3rd decades (5,21). The cysts are slightly more common in males than females (62,63). The vast majority of patients are asymptomatic, and their lesions are discovered incidentally (31,64). The diagnosis is seldom suspected preoperatively, most likely because of the rarity of the lesions (65,66).

Cervical thymic cysts can be found anywhere along the path of the thymopharyngeal duct, immediately adjacent to the carotid sheath from the angle of the mandible to the thoracic inlet. Patients characteristically present with a slowly enlarging, painless mass in the lateral portion of the neck near the thoracic inlet, either anterior or deep to the sternocleidomastoid muscle (61–63,66,67). Some investigators have found that the lesions occur more often on the left side (61,62). They vary in shape from a 1-cm round cyst to a variant, persistent thymopharyngeal duct cyst as long as 26 cm extending into the mediastinum (60,68).

Less commonly, they may cause hoarseness, dysphagia, stridor, and respiratory distress in newborns (67,69,70). Sudden enlargement may be seen when the Valsalva maneuver is applied (either due to increasing intrathoracic pressure which elevates a mediastinal thymic gland or vascular engorgement) or may be caused by hemorrhage or recent viral infection (21,67,69,71). There are no reports of myasthenia gravis or neoplasia associated with cervical thymic cysts, although tumors have occurred in solid cervical thymic tissue (71). This may reflect the lack of active thymic tissue in the cysts (64,67). About 2% of cervical thymic cysts recur in adults following surgical resection (67).

Connection of cervical thymic anomalies with mediastinal thymic tissue, either by direct extension from the main mediastinal gland or through a fibrous cord (remnant of the thymopharyngeal duct), occurs in 50% of cases (65,66,72). This connection often necessitates a more extensive surgical resection than might be expected from an innocuous clinical presentation. A fibrous strand may also connect cervical thymic cysts with the thyroid gland (61).

Figure 27. **(a)** Photomicrograph (original magnification, ×10; H-E stain) of a thymic cyst specimen shows its squamous epithelial lining (arrowheads). **(b)** Photomicrograph (original magnification, ×10; H-E stain) of a thymic cyst specimen shows thymic tissue in the cyst wall. These lobular aggregates of lymphoid tissue contain Hassall corpuscles (small, round, squamous islands) (arrows). Two large areas of cystic degeneration (*c*) are indicated.



a. **b.**

● Pathologic Characteristics

Most cervical thymic cysts are unilocular and thin walled, containing light amber to dark brown fluid. The cyst wall may be composed of a broad spectrum of epithelium, ranging from flattened squamous or cuboidal cells to multilayered stratified squamous epithelium to even primitive respiratory epithelium. Lobulated lymphoid tissue in the cyst wall contains Hassall corpuscles (Fig 27) (62,63). Occasionally, the wall contains foreign-body giant cell reaction along with cholesterol clefts and cholesterol granulomas (61,72). Both the cyst fluid and cyst wall may contain lymphoid aggregates and cholesterol crystals as nonspecific findings (62). Less commonly, small foci of thyroid or parathyroid tissue are noted and emphasize the embryologic association of the thymus with these structures (60,65).

Because the thymus arises from the third and fourth branchial clefts, thymic cysts may have extension through the thyrohyoid membrane into the pyriform sinus. This similarity with third and fourth branchial cleft cysts explains why previous authorities believed that cervical thymic cysts were actually a variant of branchial cleft cysts (63). The difference between these lesions is the presence of thymic tissue in thymic cysts. Mikal (62) used this distinction to classify three lesions: true thymic cysts (congenital persistence of the thymopharyngeal duct), mixed thymic cysts (a theoretical possibility of a fistula extending from the skin to the pharynx), and false thymic cysts (from acquired degeneration of Hassall corpuscles).

● Radiologic Features

At US, a thymic cyst appears as a large, usually unilocular cystic mass extending downward, parallel to the sternocleidomastoid muscle. The characteristic CT appearance is that of a uniloculated or multiloculated, hypoattenuated cystic mass adjacent to the carotid space (68,70,73,74). The mass may extend into the mediastinum (Fig 28) (21,74). The signal intensity of thymic cysts on MR images correlates with long T1 and T2 relaxation times (hypointense on T1-weighted images, hyperintense on T2-weighted images). One report described these lesions as "a T1- and T2-enhancing cystic mass," although it is not clear if a contrast agent was indeed administered (75). Because of its superior depiction of thoracic inlet anatomy and its lower cost compared with those of MR imaging, CT is considered the imaging modality of choice to direct an optimal surgical approach (73).

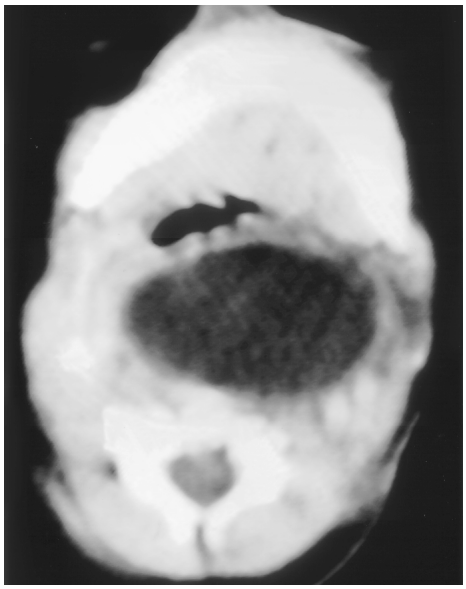
■ CERVICAL BRONCHOGENIC CYSTS

Cervical bronchogenic cysts are extremely rare. They result from an anomalous foregut development, but the reason why these cysts reach an aberrant position in the neck remains unclear. They have been reported in infants as well as in adults and occur in males about three times as often as in females (37).

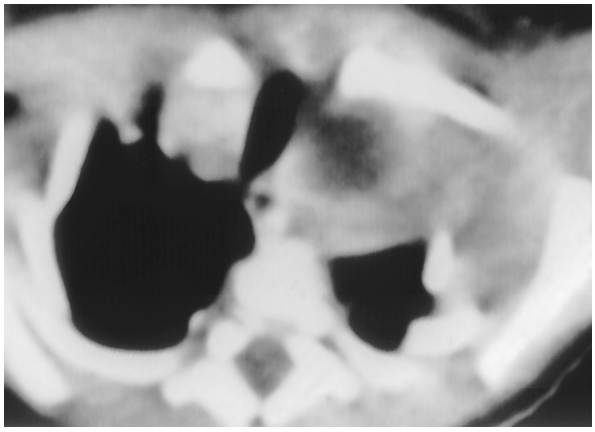
● Clinical Characteristics

Cervical bronchogenic cysts are seldom recognized clinically. They may manifest as a swelling or draining sinus, usually located in the suprasternal notch or supraclavicular area. The size of the cysts varies from less than 1 cm to more than 6 cm (37).

Figure 28. Thymic cyst in a 35-week gestation male infant in whom a left-sided neck mass was noted incidentally at discharge physical examination. During radiologic evaluation, the patient developed stridor secondary to enlargement of the mass. **(a)** Unenhanced CT scan obtained at the level of the mandible shows the largest width of the predominantly hypoattenuated mass. The airway is displaced and compressed by the retropharyngeal mass. **(b)** Unenhanced CT scan obtained at the thoracic inlet level shows the inferior extent of the mass into the mediastinum with expansion of the left hemithorax.



a.



b.

• Pathologic Characteristics

A cervical bronchogenic cyst has a columnar, ciliated, pseudostratified epithelial lining. Blood vessels, hyaline cartilage, smooth muscles, seromucinous glands, and elastic fibers are also variably present in the cyst wall. Squamous metaplasia is not uncommon in cysts that have been previously infected (76).

• Radiologic Features

Three of the four cases of cervical bronchogenic cyst reported in the literature occurred before the advent of CT. In the solitary case seen at CT, the cyst had a tubular conFiguration anterior to the trachea (77). This cyst was infected at the time of imaging, which accounted for the presence of air within the lesion.

■ LARYNGOCELE

While stationed in Egypt in 1829, Larrey, Napoleon's surgeon-in-chief, first reported a series of laryngoceles or "goîtres aeriennes" that he observed in men who would hourly chant the Koran from the minarets and in several drill instructors (78). He speculated that shouting and voice abuse led to the condition. His discovery and explanation were met with considerable skepticism by his medical contemporaries, given the rarity of the finding in other people with similar occupations (79). Even today, the cause of these lesions is disputed. A brief review of the laryngeal anatomy highlights the current thinking about the origin of these lesions.

The laryngeal ventricle (of Morgagni) is a slitlike cavity, the orifice of which is located between the false and true vocal cords. Along the anterior third of the roof of this ventricle arises a small, blind, mucosa-lined pouch known as the laryngeal saccule (of Hilton) or appendix (80,81). It extends superiorly between the false vocal cord and aryepiglottic fold medially and the thyroid cartilage laterally (82). The saccule is relatively large in newborns and children but starts to involute by the end of the 6th year of life. It is heavily invested with mucous glands, which has led some to speculate that the structure provides lubrication to the vocal cords (80).

Uniform agreement among authorities about the features that distinguish a laryngocele has been lacking (83). Early investigators (Burke and Golden) considered a saccule that extended beyond the superior border of the thyroid cartilage to qualify as a laryngocele (79). However, in an autopsy review of 100 "normal" larynxes, Broyles (81) demonstrated that 7% had saccules of sufficient length that they would be considered laryngoceles on the basis of the early criteria. Consequently, still others defined a laryngocele as a large saccule that is symptomatic and palpable (83). Today, most accept Burke and Golden's original description and recognize both symptomatic and asymptomatic laryngoceles (84).

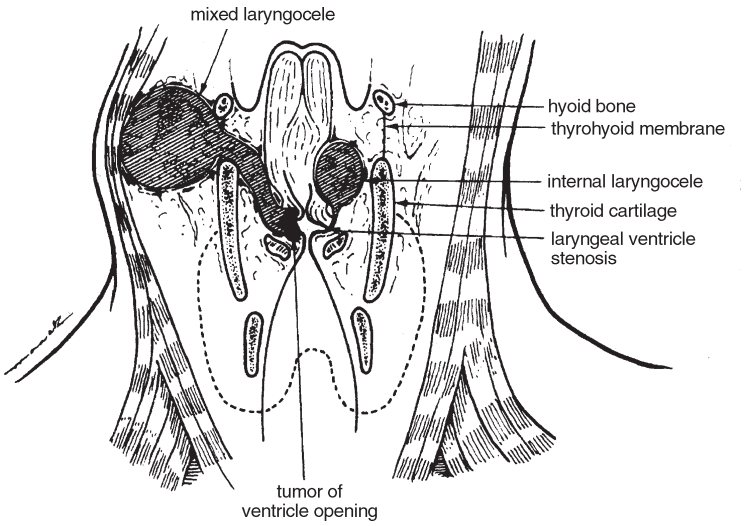


Figure 29. Drawing illustrates the three types of laryngocele. Internal laryngoceles are within the larynx itself and do not cross the thyrohyoid membrane. External laryngoceles penetrate the thyrohyoid membrane at the neurovascular bundle. The segment confined by the membrane is of normal size and connects normally with the laryngeal ventricle, whereas the portion outside the membrane is dilated. Mixed laryngoceles are dilated in both segments. (Reprinted, with permission, from reference 5.)

Simply stated, a laryngocele is a dilated laryngeal saccule, and there are three types: internal, external, and mixed. Approximately 40% of laryngoceles are internal; these laryngoceles are confined to the larynx and do not pierce the thyrohyoid membrane. External laryngoceles (26% of cases) extend through the thyrohyoid membrane at the point of insertion of the superior laryngeal nerve and vessels (neurovascular bundle). The component superficial to the thyrohyoid membrane is dilated, and the saccular portion inside the membrane is normal in size. Finally, mixed laryngoceles (44% of cases) have abnormal dilatation of the saccule on both sides of the thyrohyoid membrane (Fig 29) (85).

A laryngocele may develop due to an increase of supraglottic pressure, but this factor alone apparently does not produce a laryngocele. It is likely a combination of a long saccule and actions that result in increased supraglottic pressure (shouting, playing a wind instrument, coughing, etc) that lead to the development of these lesions (79). Congenital laryngoceles are rare and likely reflect the presence of an enlarged saccule during infancy and early childhood (78,86,87).

The prevalence of laryngoceles is uncertain given the prior difficulty in defining exactly what constitutes this entity. Stell and Maran (88) estimated a prevalence of one case per 2.5 million people in England. Acquired laryngoceles usually manifest clinically in middle-aged men. The lesions are bilateral in 23% of cases (85). Infection of laryngoceles occurs in 8%–10% of cases, and in that setting the lesion is called a laryngopyocele (88).

The association of laryngocele with laryngeal carcinoma is well-documented: Investigators worldwide have reported an increased frequency of laryngoceles in patients with laryngeal carcinoma (84,89,90). In about 15% of cases, laryngoceles coexist with laryngeal carcinoma (85). French investigators have also demonstrated a statistically significant difference in the lengths of laryngeal ventricles in patients with laryngeal carcinoma (longer ventricle) compared with those in patients with pharyngeal carcinoma (normal length) (91). Of importance to the radiologist, almost half of laryngoceles detected with plain radiography contain a laryngeal carcinoma (84). It appears that a laryngocele, when it occurs in association with a laryngeal carcinoma, is the result of the neoplasm. The tumor effectively occludes the orifice to the laryngeal ventricle (and saccule), and this partial airway obstruction leads to increased intralaryngeal pressure (84).

● Clinical Characteristics

Despite their congenital derivation, laryngoceles usually manifest in adults. A patient's symptoms depend on the size and location of the lesion. Hoarseness, dysphagia, and stridor may occur

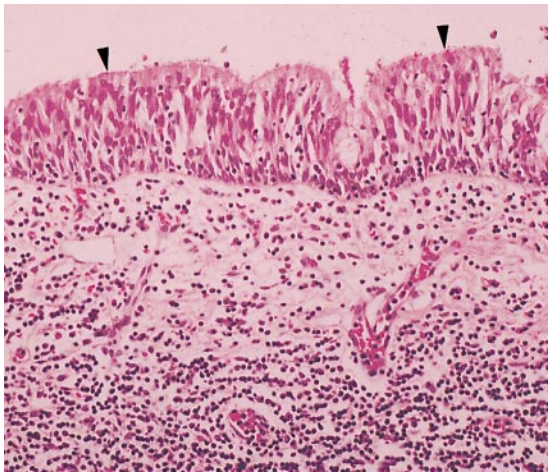


Figure 30. Photomicrograph (original magnification, ×10; H-E stain) of a laryngocele specimen shows respiratory epithelium (arrowheads) lining a cystic dilatation of the laryngeal saccule.

with internal laryngoceles, whereas a compressible soft-tissue mass is seen with the external type (88). This mass may expand when a Valsalva maneuver is performed (87). The Bryce sign (gurgling or hissing sound on compression of the mass) may also be noted in external laryngoceles (87). Most congenital laryngoceles are of the internal type, and patients have symptoms of airway obstruction, feeding difficulties, and a weak cry (87). Surgical resection is the recommended therapy of choice.

• Pathologic Features

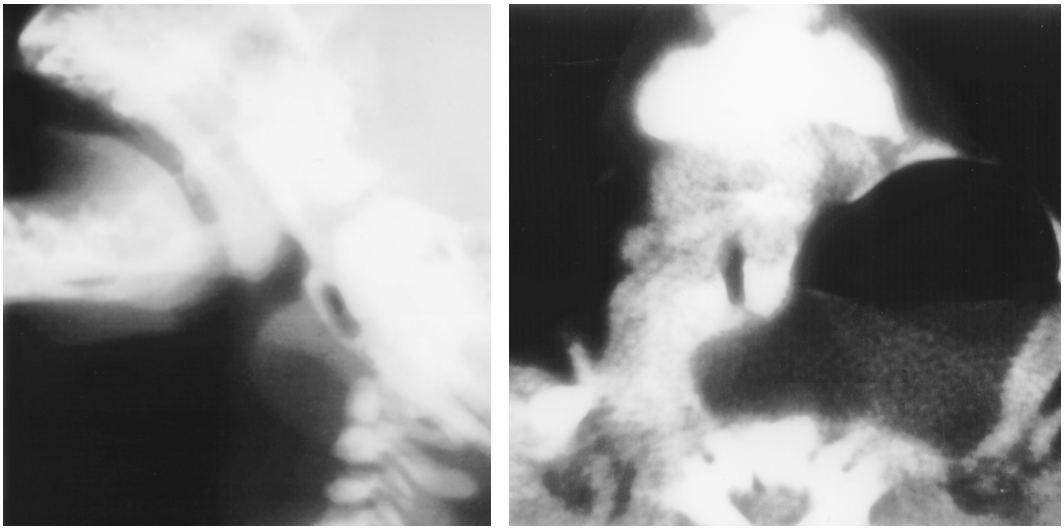
Laryngoceles are lined by pseudostratified, columnar, ciliated epithelium with occasional foci of stratified squamous epithelium and a mixture of submucosal serous and mucous glands (Fig 30) (88,92). This composition distinguishes these lesions from laryngeal cysts, which are lined entirely by squamous epithelium (88).

• Radiologic Features

The radiographic findings of a sharply defined, round or oval radiolucent area within the paralaryngeal soft tissues may be diagnostic of an external or mixed laryngocele (93). Internal laryngoceles, however, may be difficult to perceive on plain radiographs. Use of a barium swallow study in which the laryngocele does not fill with contrast material will aid in differentiating it from a lateral pharyngeal diverticulum (93).

On CT scans, a laryngocele appears as a well-defined, smooth mass in the lateral aspect of the superior paralaryngeal space. Internal laryngoceles will be limited by the thyrohyoid membrane. External and mixed laryngoceles lie superficial to the thyrohyoid membrane at the point of insertion of the superior laryngeal nerve and vessels (neurovascular bundle). The attenuation of these lesions may vary, depending on the amount of secretions, air, and soft tissue from an associated laryngeal neoplasm. A laryngocele may be filled entirely with air or contain air-fluid levels (Fig 31) (87,92,93). If it is obstructed, it may be completely filled with mucoid secretions. The presence of soft-tissue attenuation within a laryngocele suggests an underlying laryngeal neoplasm, the extent of which is accurately depicted by CT (94). Definitive diagnosis of a laryngocele at CT rests with establishing a connection between the air sac and the airway (95).

MR imaging with its superior contrast resolution and multiplanar capability appears superior to CT for distinguishing the laryngocele from an underlying laryngeal neoplasm in the presence of fluid or debris within the dilated sac. This is best appreciated on T2-weighted images on which the neoplasm is lower in signal intensity compared with that of the fluid within the laryngocele (82).



a. **b.**

Figure 31. Congenital laryngocele in a 2-day-old infant boy. **(a)** Lateral neck radiograph shows both a large retrotracheal soft-tissue mass and an air-containing mass anterior to the airway. **(b)** Axial CT scan shows a large retro- and paratracheal cystic lesion displacing and narrowing the airway lumen. The presence of an air-fluid level within the mass suggests communication with the airway.

■ SUMMARY

Congenital cystic lesions of the neck are uncommon. They are usually diagnosed in infancy or childhood, but detection may be delayed until adulthood. They often manifest as slow-growing masses and cause symptoms only after enlarging sufficiently or after infection. The clinical manifestations combined with knowledge of the embryology and spatial anatomy of the head and neck often provide clues for a correct diagnosis. US is frequently used initially to confirm the cystic nature of the lesion. CT and MR imaging provide essential information on the cyst location that allows optimal preoperative planning.

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