Chapter 86: Congenital Neck Masses

J. Oliver Donegan

Congenital anomalies must be considered in the differential diagnosis of any neck mass. They form an interesting group of lesions with diverse embryogenesis and clinical manifestations. The otolaryngologist-head and neck surgeon managing these masses must be thoroughly familiar with their embryology, anatomy, and clinical manifestations to establish an early diagnosis and institute appropriate therapy.

This chapter discusses the more common congenital abnormalities that appear as neck masses, including (1) branchial cleft cysts, (2) lymphangiomas, (3) hemangiomas, (4) teratomas and dermoid cysts, (5) laryngoceles, and (6) thymic cysts. Thyroglossal duct cysts, although falling within this group, are covered in Chapter 137.

Branchial Cleft Cysts

Embryology

Many structures in the head and neck develop from the branchial apparatus. This development occurs around the second week of fetal life and usually is complete by the sixth to seventh week. The branchial apparatus consists of a series of five mesodermal arches that appear in the lateral wall of the foregut and are separated externally by ectodermal-lined grooves or clefts and internally by endodermal-lined pouches (Fig. 86-1). A branchial plate that temporarily is devoid of mesoderm separates the clefts and pouches. The clefts do not communicate with the lumen of the foregut at any time, as they do in the gill apparatus of fish.

The five branchial arches are numbered in a cranial to a caudal direction. The fifth arch does not appear on the surface and by convention is called the sixth arch. As the branchial arches develop, they contain a cartilaginous skeleton, muscle rudiments, a nerve, and an artery. The mandible develops from the cartilage of the first arch, whereas the second and third arch cartilages give rise to the hyoid bone. The fourth and sixth arches are forerunners of the laryngeal skeleton. Table 86-1 shows structures derived from the branchial apparatus.

Table 86-1. Structures derived from branchial apparatus

Ectoderm
First cleft

External auditory canal
Tympanic membrane

Mesoderm
First arch

Mandible and teeth, Meckel's cartilage

Muscle of mastication, tensor tympani muscle, and anterior belly of digastric muscle
Malleus and incus; external maxillary artery
Fifth cranial (trigeminal) nerve
Middle layer of tympanic membrane

Endoderm
First pouch

Eustachian tube
Tympanic cavity
Mastoid antrum and cells
Tympanic membrane

Ectoderm
Second cleft

Cervical sinus of His (epidermis of anterior cervical triangle)

Mesoderm
Second arch

Platysma, muscles of facial expression, and posterior belly of digastric muscle
Stapedial artery
Body and lesser horns of hyoid
Stylohyoid ligament
Seventh (facial) and eight (acoustic) cranial nerves
Styloid process and stapes; stapedius muscle

Endoderm
Second pouch

Palatine tonsil
Supratonsillar fossa

Ectoderm
Third cleft

Cervical sinus of His

Mesoderm
Third arch

Superior constrictor muscles
Internal carotid artery
Ninth cranial (glossopharyngeal) nerve
Greater horns and body of hyoid

Endoderm

Third pouch

Inferior parathyroid
Thymus reticulum
Piriform fossa

Ectoderm
Fourth cleft

Cervical sinus of His and epidermis of lower neck

Mesoderm
Fourth arch

Part of epiglottis
Thyroid and cuneiform cartilages
Tenth cranial (vagus) nerve
Aortic arch and right subclavian artery
Inferior constrictor and laryngeal muscles

Endoderm
Fourth pouch

Superior parathyroid

Mesoderm
Sixth arch

Part of laryngeal muscles
Corniculate, arytenoid, and cricoid cartilage
Eleventh cranial (spinal accessory) nerve
Ductus arteriosus.&

The first and second arch arteries usually degenerate before the other arch arteries are formed. The second artery may persist as the stapedial artery coursing through the crura of the stapes; this rare finding may cause difficulties during surgery on the middle ear. The proximal part of the internal carotid artery is derived from the third arch artery, and the remaining portion of the internal carotid artery develops from the dorsal aorta. The external carotid and common carotid arteries are derived from the third arch artery. The fourth arch artery becomes the arch of the aorta and on the right side also contributes to the proximal portion of the subclavian artery. The sixth arch artery on the left persists as the ductus arteriosus and on the right side it forms a small pulmonary artery.

The mandibular division of the fifth cranial (trigeminal) nerve is the nerve of the first arch. This nerve innervates muscles derived from the first arch: the muscles of mastication, the anterior belly of the digastric and the mylohyoid, the tensor tympani, and the tensor veli palatini. The nerve of the second arch is the seventh cranial (facial) nerve (CN VII), which supplies muscles of second arch derivation. The ninth cranial (glossopharyngeal) nerve (CN

XI) supplies the third arch artery. This innervates the stylopharyngeal and the superior and middle constrictor muscles. The nerve of the fourth arch is the superior laryngeal branch of the tenth cranial (vagus) nerve; the recurrent laryngeal nerve, another branch of the vagus, supplies the sixth arch.

As the branchial apparatus develops, the first and second arches grow in a caudal direction. At the same time mesodermal growth forms the epipericardial ridge, which contains rudiments of the sternocleidomastoid, trapezius, infrahyoid, and lingual musculature. As a result of these proliferations, the third and fourth arches become recessed in a deepening ectodermal pit, the cervical sinus of His (Fig. 86-2). The increasing growth of surrounding structures reduces the opening of the cervical sinus to a narrow channel called the cervical duct. The cervical duct soon is obliterated, followed by obliteration of the ectodermal-lined cervical sinus. Branchial cysts other than first branchial cysts arise from retained portions of this cervical sinus. Branchial fistulas, epithelial-lined tracts connecting the skin to the lumen of the foregut, may arise because of persisting cervical sinus in addition to breakdown of the branchial plate or the closing membrane between the clefts and pouches. Branchial sinuses, which are epithelial-lined tracts opening onto the skin or the lumen of the foregut and ending blindly in the deeper tissues of the neck, may arise because of a persisting cervical duct or breakdown of a closing membrane. Cysts and sinuses frequently coexist and the sinus tract opens most commonly into the lumen of the pharynx.

As normal development proceeds, the branchial clefts and pouches are obliterated. The first branchial cleft is somewhat unique because only this cleft does not completely disappear. Portions of the first cleft deepen to form the external auditory meatus. Persistence of the cleft in this area or duplication of the external auditory meatus are the two main theories explaining the occurrence of first branchial cleft cysts, sinuses, and fistulas.

Classification

Branchial cleft cyusts, sinuses, and fistulas are classified as first, second, third, and fourth branchial cleft abnormalities. Second branchial clet cysts are by far the most common.

Much confusion exists in the literature concerning the subclassification of first branchial abnormalities. Arnot (1971) was the first to classify these anomalies. He proposed dividing these structures into two types. The type I defect was any cyst or sinus in the parotid gland that was lined by squamous epithelium and appeared in early or middle adult life. The type II defect was a sinus or a cyst in the anterior triangle of the neck having a tract communicating with the external auditory canal.

The classification that Work (1972) proposed now is most widely accepted. In this scheme a type I anomaly is of ectodermal origin and is considered a duplication of the membranous external auditory canal. Type II lesions are duplication anomalies of the membranous external auditory canal and pinna that contain skin (ectoderm) and cartilage (mesoderm). Both types may be related intimately to the parotid gland and CN VII. Middle ear involvement has occurred but is extremely rare.

Surgical anatomy

First branchial cleft cysts

The epithelial tracts of both types described here end close to the external auditory canal or may be attached to or communicate with the canal. First branchial cleft cysts account for 8% of branchial anomalies.

Type I. These lesions appear in the preauricular region, usually anterior or posterior to the pinna (Fig. 86-3). They may be cysts, sinuses, or fistulas with a fistulous tract connecting the skin and the external auditory canal. They tend to lie parallel to the external auditory canal and usually are lateral to CN CII. In the preauricular area they may be embedded in the parotid gland.

Type II. These anomalies usually are located immediately posterior or inferior to the angle of the mandible (Fig. 86-4). They are related intimately to the parotid gland and may lie lateral, medial, or between the branches of CN VII. Ensuring complete resection of these lesions frequently requires a superficial parotidectomy and exposure of the main trunk and branches of CN VII.

Second branchial cleft cysts

Second branchial cleft cysts account for the majority of branchial abnormalities. The cyst or external opening is found along the anterior border of the sternocleidomastoid muscle (Fig. 86-5). The epithelial tracts pass superiorly, lateral to the carotid arterial system and CN IX and CN XII. Coursing superior and lateral to CN IX, they turn in a medial direction between the internal and external carotid arteries. These tracts end close to the middle constrictor muscle or may have an internal opening in the region of the tonsillar fossa (Fig. 86-6).

Third branchial cleft cysts

Third branchial cleft cysts are rare. They also appear along the anterior border of the sternocleidomastoid muscle; however, the course of the fistulas and sinus tracts is quite different (Fig. 86-7). The tract ascends lateral to the common carotid artery, passes posterior to the internal carotid artery, superior to CN XII, and inferior to CN IX; and then courses medially, piercing the lateral aspect of the thyrohyoid membrane to open into the piriorm sinus. This opening usually is superior to the internal laryngeal nerve.

Fourth branchial cleft cysts

Until a few years ago fourth branchial cleft abnormalities were thought to be theoretical possibilities only. Such anomalies, when investigated, often proved to be third branchial derivatives if strict anatomic criteria were applied (Myers, 1988). However, recent reports are more convincing in proving the occurrence of these interesting lesions (Grodin, 1990). On the left side, fourth branchial cleft tracts begin at the apex o the piriform sinus (Fig. 86-8). They descend to exit the pharynx caudal to the superior laryngeal nerve, the cricothyroid muscle, and the thyroid cartilage. The tract then continues to course inferiorly,

lateral to the trachea and the recurrent laryngeal nerve. On the left side the fistula tract descends into the thorax and loops posteriorly around the aortic arch before ascending into the neck posterior to the common carotid artery. The tract then crosses CN XII before descending again to open onto the skin at the anterior aspect of the lower portion of the sternocleidomastoid muscle. On the right side the tracts pass anteriorly beneath the right subclavian artery before passing superiorly into the neck. Many authors believe that a complete fourth branchial fistula will never be surgically demonstrated because the approach and exposure of the cervical and mediastinal components would be unnecessarily aggressive and probably not indicated.

Cysts may occur in any location along the path of sinuses and fistulous tracts associated with all four branchial cleft anomalies.

Histopathology

Branchial cleft anomalies usually are lined with stratified squamous epithelium although a small number may be lined with ciliated columnar epithelium. Keratin, hair follicles, sweat and sebaceous glands, and cartilage also may be present. The presence of lymphoid aggregates in the walls of the lesions is a frequent finding.

The possibility of carcinoma arising in branchial remnants is controversial. Many authors doubt that such neoplastic changes occurs. Such lesions are indeed rare, and the physician must adhere to strict criteria when making such a diagnosis. Martin et al (1950) have outlined the criteria that must be met before the diagnosis is acceptable:

1. The tumor must be located on a line anterior to the tragus to the anterior border of the sternocleidomastoid muscle.

2. The histology should be consisted with an origin from tissue present in branchial vestigia.

3. The patient must be followed for at least 5 years with periodic examinations without developing another tumor that could be regarded as a primary lesion.

4. Ideally, it must be demonstrated that the cancer developed in the wall of an epithelial-lined cyst situated in the lateral aspect of the neck.

For any patient to fulfill all these criteria is extremely unusual, and the diagnosis remains tentative in most instances.

Clinical manifestations

Branchial cysts, sinuses, and fistulas may be seen at any age but generally become evident in childhood and early adulthood. No sex predilection is noted. On rare occasions a familial occurrence has been noted. Branchial cleft cysts present as nontender, fluctuant masses in the typical locations mentioned previously; 2% to 3% of these cases are bilateral. These cysts may become inflammed and tender and may develop abscesses. The inflammatory episodes frequently are associated with upper respiratory tract infections. Depending on their

size and location, associated symptoms may arise: dysphagia, dyspnea, and stridor. Spontaneous rupture of an abscess may occur, resulting in a draining sinus onto the skin or possibly the discharge of purulent secretion into the pharynx.

Sinuses and fistulas often are noted on routine examination, although a history of associated inflammation and discharge of mucoid and/or purulent secretions from the tract may exist. These inflammatory episodes occasionally are associated with upper respiratory tract infections.

First branchial anomalies may appear as parotid masses or parotid inflammation. If the associated tract communicates with the external auditory canal, otorrhea and otalgia also may be noted.

In recent years third and fourth branchial pouch abnormalities have been reported as being associated with recurrent acute suppurative thyroiditis in childhood. As third and fourth branchial abnormalities almost always present on the left side, these children may develop left paratracheal fullness and tenderness with associated fever. A thyroid scan may reveal reduced radioisotope uptake in the ipsilateral thyroid lobe. Cultures in these children demonstrate a variety of aerobic and anaerobic organisms such as *Escherichia coli, Klebsiella, Proteus,* and *Clostridium.* Retropharyngeal abscesses may also be associated with these abnormalities.

Differential diagnosis

The differential diagnosis of these abnormalities includes parotid tumor and infection, metastatic carcinoma in cervical lymph nodes, cystic hygroma, hemangioma, lymphoma, carotid body tumor, dermoid cyst, and laryngocele.

Management

The definitive therapy for branchial anomalies is complete surgical excision. The injection of sclerosing solutions and use of radiation therapy should not be done. Aspiration of these masses may be helpful in reaching a diagnosis, and incision and drainage sometimes are necessary if an abscess has developed. Surgical excision should await resolution of infection. Inadequate excision leads to recurrence and further inflammatory episodes, rendering subsequent excision extremely difficult.

Preoperatively the sinus tracts and fistulas may be outlined with radiopaque dyes. Other radiographic studies of potential diagnostic help in the management of congenital neck masses include ultrasonography, high-resolution computed tomography, and magnetic resonance imaging. Radiography may be used to distinguish cystic from solid masses, to localize lesions precisely, and to demonstrate compromise of adjacent structures. Probing these tracts may elicit vasovagal reactions. Intraoperatively probes may be placed in the tracts; this maneuver greatly facilitates dissection and complete removal.

First branchial anomalies are dissected free to their termination close to or at the external auditory canal. A superficial parotidectomy and CN VII dissection are required or those lesions intimately associated with the parotid gland. If the tract terminates close to the external auditory canal, a portion of the canal skin and cartilage should be excised. Work

(1972) suggest opening and packing the external auditory canal to marsupialize the tract and prevent recurrence.

Second, third, and fourth branchial anomalies are approached through a horizontal incision in the neck. Isolated cysts are always deep to the platysma muscle and must be carefully dissected from surrounding structures. Associated or isolated sinus tracts and fistulas are approached by making an elliptic incision around the tract opening and dissecting the tract to its termination near the tonsillar fossa or piriform sinus. Not infrequently these tracts are lengthy and a second incision higher in the neck, the so-called stepladder surgical approach, is required to complete the dissection.

A single incision parallel to the sternocleidomastoid muscle also gives the desired exposure but is much less acceptable from a cosmetic viewpoint. The surgeon must have a thorough anatomic knowledge of these anomalies to avoid injury to adjacent important structures. Recurring infection and previous surgery make this dissection more hazardous and difficult.

In cases of suppurative thyroiditis resulting from a third or fourth branchial pouch anomaly, hemithyroidectomy in addition to closure of the internal piriform sinus tract opening is recommended.

Lymphangioma (Cystic Hygroma)

Lymphangiomas are congenital malformations of lymphatic channels. The majority occur in the neck, presumably as developmental anomalies of the jugular lymphatic sac. These jugular lymphatic sacs eventually form communications with the venous system and become the terminal portions of the thoracic duct and the right lymphatic duct.

Lymphangiomas usually are present at birth; 80% to 90% are detected by the second year of life, with less than 10% occurring in adults. Males and females are equally affected. Lymphangiomas may occur in any region of the body, with most found in the head and neck areas.

Histopathology

Lymphangiomas have been classified into three groups: (1) *lymphangioma simplex,* consisting of capillary-sized, thin-walled lymphatics; (2) *cavernous lymphangioma,* composed of dilated lymphatic spaces; and (3) *cystic hygroma,* containing lymphatic cysts ranging from a few millimeters to several centimeters in diameter.

Bill and Sumner (1965) have drawn these groups together by proposing that the three types depend on anatomic location rather than histologic differences. Large cystic lesions may develop in areas with loose areolar tissue and distinct tissue planes, whereas smaller lesions occur in areas devoid of these features. Combinations of these three types are often seen in a single location.

The cavernous lymphangioma may contain buds of lymphangiomatous tissue that extend into neighboring structures, especially muscle. This type, although not thought to be

neoplastic, has definite invasive tendencies and is more likely to recur following treatment. A flattened endothelium lines all three types.

Clinical manifestations

Lymphangiomas appear as painless, soft, single or multiloculated masses (Fig. 86-9). They vary in size from 1 to 2 mL to very extensive lesions that can occupy one side of the neck and even extend to the skull base and into the thoracic cavity. Compression of adjacent structures leads to symptoms such as dysphagia, dyspnea, stridor, and pain. Inflammatory episodes may occur. Hemorrhage into the cystic spaces may cause rapid enlargement of these masses, and they may transilluminate. A CT and/or MRI scan assists in delineating their true extent.

Management

All three types of lymphangioma extend locally, surrounding blood vessels, nerves, muscles, and other structures in the neck. The treatment of choice is surgical excision. These lesions exhibit little tendency toward spontaneous regression, but in infants the physician should allow time for involution to occur if confident of the diagnosis and if no complications have arisen. If possible, surgery should be postponed until the child is 3 to 4 years old. Recurring infection, symptoms of compression, and a doubtful diagnosis may precipitate surgery at an earlier age.

The mass is approached through a horizontal incision in the neck. The surgeon locates the wall of the cyst and makes every effort to avoid rupture of the lesion because this leads to collapse and renders complete excision difficult. These masses usually are quite adherent to adjacent structures and resist being shelled out. Although the surgeon occasionally must leave remnants of the cyst behind, recurrence rates are low, ranging from 5% to 10%. Cavernous lymphangiomas defy complete surgical resection because of their tendency to insinutate into muscle planes.

Other treatment modalities used in the past for lymphangiomas included injection of sclerosing solutions and radiation therapy. These have not been successful; radiation therapy is highly undesirable because of the risk of neoplastic change developing in the surrounding tissue many years later.

Hemangiomas

Hemangiomas are developmental vascular abnormalities. The majority are noted in the immediate postnatal period, with 96% being evident by the age of 6 months. Many of these lesions involute spontaneously by the time the child is 7 years old.

Histopathology

Hemangiomas have been classified on histologic grounds as capillary, cavernous, mixed, and juvenile (proliferative). Batsakis (1979) includes a fifth type, infiltrative, which may be derived from any of the other forms. This infiltrative quality is characteristic of more deeply seated subcutaneous, intramuscular, and deep fascial hemangiomas, manifesting a

locally invasive growth pattern that poses therapeutic difficulties. This type of lesion is commonly capillary or cavernous and may be seen in children and adults.

Hemangiomas are the most common benign tumors of the head and neck in children. One of the most widely accepted theories for the development of congenital vascular lesions describes the normal embryologic progression of the endothelial stage, consisting of endothelial lakes, to the retiform stage, consisting of a coalescence of the endothelial lakes into communicating capillary channels. Additional maturation to the gross differentiation stage results in the formation of mature arteries and veins. Abnormalities in the normal progression of developmental stages leads to vascular malformations. Anomalies within the endothelial stage will produce cavernous or capillary hemangiomas. Arrest within the retiform stage will produce mixed hemangiomas or arteriovenous istulas (Persky, 1986).

Clinical manifestations

In the head and neck region hemangiomas appear as either diffuse skin lesions or soft cystic masses in the oral cavity, pharynx, parotid gland, and neck (Fig. 86-10). The skin lesions are not difficult to diagnose. In the deeper tissues of the neck other characteristics such as compressibility, bluish discoloration of the overlying skin, bruits, change in size during crying or straining, and associated hemangiomas elsewhere in the body assist the physician in reaching the correct diagnosis.

Radiology may be helpful in delineating these masses. Findings on plain radiographs and CT and MRI scans are not characteristic, although occasionally phleboliths may be seen in these lesions. Angiography may be helpful in demonstrating feeding vessels and outlining the exact extent of the lesion. Not all hemangiomas can be demonstrated by these studies; if the diagnosis remains in doubt, needle aspiration, and perhaps open biopsy is necessary to identify the hemangioma.

Management

The approach to management of hemangiomas in the neck should be conservative. In infants these lesions often undergo a phase of rapid growth followed by slow resolution over the next 3 to 4 years. Lesions appearing for the first time later in life are less likely to involute spontaneously. A period of observation is recommended for those lesions that are not causing functional impairment.

Hemangiomas undegroing rapid growth in the neck may interfere with the air and food passages, necessitating early intervention. Steroids may be worthwhile as an initial step in management because these drugs are capable of inducing cessation of growth and occasionally even resolution. Steroids have been demonstrated to have an occasional dramatic effect on the regression o hemangiomas. The mechanism of action of steroids is unclear, but may be related to either an increase in vascular response to endogenous vasoconstrictive mediators or increased arterial constriction and narrowing o precapillary sphincters. Tracheostomy and feeding tubes may be required if expansion of these lesions goes unchecked.

Surgery is indicated for those hemangiomas causing functional and cosmetic impairment. This surgery may be fraught with difficulties. Blood loss may be life-threatening,

and major vascular and neural structures may be at risk. Sacrifice of important structures is not indicated in the surgical management of hemangiomas. External carotid artery ligation and/or intraarterial embolization can be used preoperatively in attempts to decrease intraoperative blood loss. Staged excision may be the preferred method of management in some instances. Because many of these deep-seated lesions are iniltrative, complete excision is even more difficult. Sclerosing agents and radiation therapy have no role to play in the treatment of these anomalies; however, the argon laser has proved effective in excision of some cutaneous hemangiomas.

Teratomas and Dermoid Cysts

Teratomas and dermoid cysts are developmental anomalies involving pluripotential embryonal cells; they are alien to the site in which they arise. Several theories have been proposed to explain the existence of these lesions. Of the two most plausible, one suggests isolation of pluripotential cells occurring during embryogenesis and subsequent disorganized growth of these cells. The second theory holds that germ layers may be buried in deeper tissues at points of failed fusion lines.

Teratomas reportedly occur in 1:4000 births, with less than 10% affecting structures in the head and neck. They most commonly are found in the sacrococcygeal, mediastinal, retroperitoneal, and gonadal regions. Teratomas are usually evident in patients at an early age, but no age group is immune. In the head and neck the sites most commonly involved are the orbital region, nose, nasopharynx, oral cavity, and neck.

Teratomas and dermoid cysts are classified into four groups. This system recognies both germ layer of origin and the complexity of tissue organization. Regardless of site, neurogenic tissue is a prominent feature of the lesions, especially in the head and neck area.

1. *Dermoid cysts.* These are the most common form of teratoma and are composed of ectoderm and mesoderm. They are covered with skin and contain epidermal appendages such as hair follicles, sebaceous glands, and sweat glands. They may be cystic but more commonly have an adipose matrix. Most occur along lines of embryonic fusion.

2. *Teratoid cysts.* These are composed of ectoderm, mesoderm, and endoderm. The lining of the teratoid cyst may range from simple stratified squamous epithelium to ciliated respiratory epithelium. Tissues from the three germ layers are poorly differentiated in these lesions.

3. *Teratomas.* These are also composed of cells from all three germ layers. In contrast to teratoid cysts, however, cellular differentiation is such that recognizable organs may be found in these masses.

4. *Epignathi.* The highest form of differentiation is found in epignathi; development of fetal organs and limbs may occur. These anomalies rarely are compatible with life.

In the neck the majority of dermoid cysts occur in the submental region above the hyoid bone. They are always midline lesions. The most common symptom is a painless mass. Pain may be present if infection develops or if the mass enlarges significantly. Submental

dermoid cysts are situated below the myohyoid muscle. Others may appear superior to this muscle, in which case the bulk of the mass is seen in the anterior floor of the mouth. In this location dermoid cysts may simulate a ranula and, if large enough, will interfere with deglutition and breathing. On palpation, these cysts frequently have a rubbery or doughy consistency.

The treatment of choice for dermoid cysts is complete surgical excision. Residual tissue may result in recurrence.

Teratomas of the neck usually are present at birth and are rare in patients over the age of 1 year. Maternal polyhydramnios is reported in 18% o cases (Batsakis, 1979). Teratomas frequently are quite large and can cause severe breathing and feeding difficulties. The overlying skin is normal.

Teratomas present at birth may present major difficulties in management of the neonatal airway at delivery and in the perinatal period. Ultrasonography allows these lesions to be diagnosed prenatally. Under these circumstances steps may be taken to protect the infant airway at birth. This may be accomplished by a multidisciplinary approach that includes endoscopy to secure the airway while the neonate remains on fetal circulation. Subsequent resection of the teratoma may then be undertaken once the airway has been secured.

Radiographs of the soft tissues of the neck may be helpful in diagnosis because calcifications and dental elements may be present in these masses.

These lesions are frequently closely associated with the thyroid gland, leading some authors to divide them into thyroid and nonthyroid teratomas. Batsakis (1979) believes that this is an unnecessary categorization, at least given the present knowledge regarding these lesions.

Teratomas should be excised as expeditiously as possible; encroachment on the airway is an ever-present danger. Encapsulization is common, facilitating complete removal. Recurrence following apparent total excision is rare.

Malignant teratomas of the neck are not common; a small number of cases have been documented, most occurring in adults. However, there are isolated reports of neonatal malignant teratomas. The prognosis in this situation is poor, and combined surgery and radiation therapy should be considered.

Laryngoceles

Laryngoceles are abnormal dilatations of the laryngeal ventricle and saccule. They are not common but should be considered in the differential diagnosis of any neck mass. They may appear at any age and not infrequently cause difficulties with diagnosis and management. They may be life-threatening because an internal component places the airway at risk.

Galen first mentioned the laryngeal ventricle in 200 AD. In 1741 Morgagni described the ventricle in detail and Hilton in 1937 completed the anatomic outline of the laryngeal saccule. The saccule arises vertically off the anterior end of the ventricle; then this pouch

passes superiorly between the false vocal cord, the base of the epiglottis, and the inner surface of the thyroid cartilage. Normally the adult saccule can extend as high as the superior border of the thyroid cartilage. Approximately 25% of these appendages in the fetus extend beyond the upper border of the thyroid cartilage and terminate medial to the thyrohyoid membrane. Pseudostratified ciliated columnar epithelium line the structure, and on the surface of its mucous membrane are the openings of 60 to 70 mucous glands. Medial and lateral to the saccule are delicate muscles that compress it to suppress its secretions on the vocal cords.

Much confusion existed in the past concerning the terminology of laryngeal cysts. DeSanto (1974) clarified this: the term *saccular cyst* has replaced the previous term *congenital laryngeal cyst.* Both the laryngocele and the saccular cyst arise from the saccular appendage. They differ in that the laryngocele communicates freely with the laryngeal lumen and is filled with air, whereas the saccular cyst has no communication with the laryngeal lumen and is filled with fluid.

Two types of saccular cysts exist. The *lateral* saccular cyst extends posterosuperiorly into the false vocal cord and the aryepiglottic fold from its origin at the nonpatent orifice of the saccule. The *anterior* saccular cyst extends medially and posteriorly to protrude into the laryngeal lumen from between the true and false vocal cords.

Three types of laryngoceles are described. An *internal* laryngocele is confined to the interior of the larynx and extends posterosuperiorly into the false vocal cord and the aryepiglottic fold. An *external* laryngocele extends superiorly to appear laterally in the neck through the opening in the thyrohyoid membrane for the superior laryngeal nerve and vessels. The simultaneous existence of both features is termed a *combined laryngocele* (Fig. 86-11). By definition a laryngocele contains air only but may become infected, filled with fluid, resulting in a *laryngopyocele.* Fluid-filled internal laryngoceles are indistinguishable from saccular cysts.

Laryngoceles may arise because of congenital enlargement of the saccule or may be acquired. One factor leading to development of a laryngocele is prolonged increase in intralaryngeal pressure, such as occurs in coughing, straining, wind instrument playing, and glass blowing; the lesions are hazards of these two occupations. Partial obstruction of the neck of the saccule because of inflammatory and neoplastic changes may also accompany the development of laryngoceles.

Although seen in all age groups, laryngoceles are most common in the fifth and sixth decade of life. Males are affected more often than females.

Clinical manifestations

The external and the mixed types of laryngoceles appear as masses in the neck. Swelling may be noted only during periods of raised intralaryngeal pressure, and the patient may be able to demonstrate the lesion on request. The patient may be aware of a gurgling sensation in the neck as the sac empties of air. The persistence and increasing size of these lesions also may be noted. Patients with combined laryngoceles also may complain of hoarseness, cough, dyspnea, and dysphagia. Laryngopyoceles appear as acute cervical

inflammations. Indirect laryngoscopy reveals the endolaryngeal component of combined lesions. These appear as mucosally covered masses involving the aryepiglottic fold and the false cord.

Radiography will assist in confirming the diagnosis. High kilovoltage inspiratory and expiratory radiographs in both anteroposterior and lateral projections usually demonstrate the abnormality (Fig. 86-12). CT also outlines these lesions (Fig. 86-13).

Management

These cysts are best managed by excision via an external approach. Direct laryngoscopy should be performed initially to rule out underlying pathology such as carcinoma within the endolarynx.

A horizontal neck incision is made at the level of the thyroid cartilage. The laryngocele is dissected free from surrounding structures and is followed to the point where it exits the larynx through the thyrohyoid membrane. This membrane is incised, and the excision is completed by removing the internal component of the sac. Removing a portion of the thyroid ala may facilitate this part of the procedure (Fig. 86-14). Similarly, a laryngofissure is not indicated for removal of these lesions and should be avoided because of the risk of anterior commissure stenosis. This lateral approach results in low recurrence rates.

Thymic Cysts

The third and fourth branchial pouches give rise to several important organs, including the thymus and parathyroid glands (Fig. 86-15). The location of these organs changes as the head and neck grows away from the pericardium. Maintaining its relation to the pericardium, the thymus extends into the thorax. The inferior parathyroid glands derived from the third pouch are drawn inferiorly with the thymus and occasionally may become intrathoracic. As the thymus descends, it maintains a connection to the third pouch. This epithelial-lined connection is called the thymopharyngeal duct. By the eighth or ninth week of life, this duct disappears. One theory proposes that remnants of this structure account for cervical thymic cysts.

Tissue derived from the thymus is a rare cause of a cervical mass. These lesions may be associated with epithelial tracts communicating with the pharynx through the thyrohyoid membrane or maintaining a connection to the thymus gland in the thorax. They usually are found in the lateral neck anterior or deep to the sternocleidomastoid muscle. Thus they are quite similar to branchial cleft cysts of the second type; their only distinguishing feature appears to be the presence of thymic tissue in those derived from the third pouch. These cysts are also susceptible to inflammation and sinus drainage. Surgical excision is the best treatment.