Paparella: Volume III: Head and Neck

Section 2: Disorders of the Head and Neck

Part 7: The Neck

Chapter 42: Congenital Neck Masses

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Except for benign cervical lymphadenopathy, the most common cause of cervical swelling in children is congenital neck masses. Their appearance is generally quite characteristic and reflects anomalies in the development of muscles, skin, blood vessels, lymphatics, and branchial apparatus. Not all congenital lesions are present at birth, and they may not present until a certain enlargement has occurred. Often, this follows an upper respiratory tract infection. Therapy must be individualized depending on the type of abnormality present (Telander and Deane, 1977).

Vascular Anomalies

Lymphangioma

Lymphangiomas, also commonly known as cystic hygromas, are soft, painless, compressible masses that represent areas of regional lymphatic dilatation. The majority of cervical lesions are though to arise from sequestration of tissue adjacent to the primitive jugular lymphatic sac. Three histologic groupings have been developed arbitrarily according to the size of the lymph channels present in the individual lesions. These histologic patterns - simple (capillary), cavernous, and cystic - may coexist in the same lesion. Because all types of histologic patterns may be found in any one lesion, the term *cystic hygroma* has been applied to all lymphangioma (Karmody et al, 1982).

Hemorrhage into a lymphangioma may result in the appearance of a hemangioma or may cause difficulty in distinguishing between these two types of lesions. Alternatively, components of a hemangioma may be present within a lymphangioma and result in a lymphangiohemangioma. Little clinical significance can be attached to the aforementioned histologic classification, and it does not influence the therapy of such lesions.

Lymphangiomas are relatively uncommon lesions that are most frequently seen shortly after birth. Approximately 60 per cent of the lesions are present before the age of 1 year, whereas 90 per cent are present clinically by the end of the second year of life. However, they have been reported as late as the fourth or fifth decade (Ravitch and Rush, 1979). It is speculated that local infection may precipitate rapid growth of previously unrecognized lesions. Thus, the initial presentation of many lymphangiomas is following an upper respiratory tract infection. It is unknown why lesions that do not present until middle age remain dormant for such a long time (Leipzig and Rabuzzi, 1978).

Except for their obvious cosmetic deformity, most cervical lymphangiomas do not cause symptoms. The differential diagnosis includes branchial cysts, thyroglossal duct cysts,

lymphomas, and true neoplasms. The lesions most commonly present in the posterior triangle of the neck. When they are located in the anterior triangle of the neck, there may be intraoral extension with pharyngeal compression and airway obstruction.

The initial evaluation of cervical lymphangiomas is frequently made by anteroposterior and lateral neck radiographs, a chest radiograph, and neck ultrasound. When lesions are quite extensive, a computed tomography (CT) scan can be helpful, especially if there is extensive mediastinal involvement or bony compression (Miller and Norman, 1979; Phillips and McGahan, 1981; Silverman et al, 1983). A lymphangiogram is rarely necessary.

Lymphangiomas are characterized radiographically as noncalcified neck masses of variable size. Mediastinal extension is present in approximately 5 per cent of patients. Careful examination of the airway is necessary to evaluate the presence of laryngeal or tracheal compression (Leonida et al, 1978; Pilla et al, 1982).

Neck sonography aids in the determination of the character and extent of a mass and its relationship to surrounding structures. The most common sonographic finding in a patient with a cervical cystic hygroma is a thin-walled, multiseptated cyst. However, solid or mixed echogenicity patterns can also occur, thus complicating the differential diagnosis (Leonida et al, 1978; Phillips and McGahan, 1981; Pilla et al, 1982). When laryngeal involvement is present, one should attempt to determine whether the lesion is primary within the larynx or is secondary to extension of a cervical lymphangioma. Although a tracheostomy will be necessary in almost all cases, therapeutic alternatives differ when laryngeal lesions are extensions of cervical lesions.

Since lymphangiomas rarely resolve spontaneously, observation is not a viable alternative for treatment. Surgical excision is the only acceptable mode of therapy for cervical lymphangiomas (Karmody et al, 1982). Controversy exists regarding the age at which excision should take place. Some experts argue for excision early in infancy, since a delay may allow multiple infections to occur and thus complicate removal. Others, however, favor waiting until a child is older if there are no complicating factors (Stal et al, 1986). Although complete excision should always be the goal, the surgeon must remember that he or she is dealing with a benign tumor. Normal structures should not be sacrificed in order to obtain a complete surgical resection. If all microscopic disease has been resected, recurrence is uncommon. Conversely, if gross tumor is left behind, a 15 per cent recurrence rate has been noted. In extremely large lesions, a planned staged resection may be preferable (Karmody et al, 1982).

Hemangioma

The most common head and neck neoplasms in children are hemangiomas. Although they are predominantly located on cutaneous surfaces, they may be found on mucosal surfaces as well. The scalp is the most frequently involved region, followed by the neck and face. Girls are more frequently affected than are boys, and the tumors are more often solitary than multifocal (Batsakis, 1979).

Even though less than 33 per cent of hemangiomas are present at birth, they are typically noted during the first month of life and progressively enlarge during the next year. In almost 90 per cent of cases, involution will occur and no therapy is necessary.

Hemangiomas can be defined as benign, nonreactive processes in which there is an increase in the number of normal- and abnormal-appearing blood vessels. In addition, these malformations can be divided into active and inactive hemodynamic categories, which are determined by the presence of arteriovenous fistulae, vascular proliferation, and overall clinical behavior. Several different types have been described (Stal et al, 1986).

The cutaneous birth mark (nevus flammeus) is an example of a capillary hemangioma that is located in the dermis. Capillary hemangiomas are composed of nests of capillary vascular channels that are lined by endothelial cells and surrounded by pericytes. In early lesions, the endothelial cells are quite plump, often obscuring the lumen of the capillaries. Thus, the lesion represents arrest in development of the mesenchymal primordia in the capillary network stage. When this occurs, the vascular nature of the lesion may not be readily apparent. The strawberry nevus is a subcutaneous mass that is actually a hypertrophic type of capillary hemangioma. This growth is marked by a period of slow evolution, which is then followed by a rapid growth phase and, possibly, involution, most often beginning by 10 months of age (Stal et al, 1986).

In contrast, cavernous hemangiomas are composed of tortuous, large vascular channels lined by endothelial cells. They are larger, more frequently involve deeper structures than the capillary hemangiomas, and are unlikely to regress spontaneously. Adventitial fibrosis is frequently present. Thrombosis may occur, with resultant dystrophic calcification and the development of phleboliths. Cavernous hemangiomas are found both in the skin and in the deeper tissues. If the lesions are not present at birth, involution is uncommon.

Arteriovenous hemangiomas frequently occur in the soft tissues of the neck and are referred to by some as arteriovenous malformations. In addition to many of the histologic features of a cavernous hemangioma, there is oftentimes intimal thickening in veins. Additionally, serial sections of such lesions demonstrates diverse arteriovenous connections. Although the histopathologic findings are characteristic, equally important to the diagnosis is the clinical presence of a pulsatile mass with the physiologic manifestations of an arteriovenous shunt (Garfinkle and Handler, 1980).

The radiographic evaluation of a cervical hemangioma is best done with a CT scan and angiography. The CT scan will demonstrate a deeply enhancing mass and graphically depict the soft tissue extent of the lesion. Angiographically, hemangiomas are well-circumscribed lobular masses that have a persistent dense tissue stain and are supplied by multiple slightly enlarged arteries. A proximal artery surrounds the lesion, and multiple smaller arteries enter the hemangioma at right angles. Arteriovenous shunting is usually not present. The angiographic appearance reflects the type of vessels that compose the hemangioma. Capillary venous malformations have dilatated, ectatic spaces that fill during the venous phase and demonstrate prolonged contrast pooling. Arteriovenous-lymphatic vascular malformations are high-flow communications with enlarged and more numerous vessels (Burrows et al, 1983).

Hemangiomas that are located in the deep subcutaneous tissues, fascia, and muscles of the neck tend to be infiltrating and difficult to treat. Although the lesions do not undergo malignant degeneration or metastasize, local control is difficult and is frequently not achieved. The intramuscular hemangioma is an example of such an invasive lesion. It usually presents

as a localized mass with a rubbery consistency and distinct margins. It is mobile and is not associated with a bruit, thrill, or pulsation. Cutaneous involvement may be present and there may be functional abnormality of the involved muscle. Patients often complain of pain secondary to compression (Batsakis, 1979).

The most common type of intramuscular hemangioma is a capillary hemangioma. In the neck, the scalene, trapezius, and sternocleidomastoid muscles are frequently involved (Stal et al, 1986). This lesion is associated with a 30 per cent recurrence rate following appropriate therapy because of its infiltrative nature. Cavernous intramuscular hemangiomas are the second most common type seen and they are associated with a 9 per cent recurrence rate. Mixed types are uncommonly found and are associated with a 25 per cent recurrence rate. Therapy for these intramuscular hemangiomas requires ligation of the feeding vessels and excision of the mass. As with lymphangiomas, the surgeon must remember that these are benign lesions and care must be taken to avoid injury to vital structures (Batsakis, 1979).

Because most congenital lesions involute spontaneously, conservative therapy is the rule for many hemangiomas. One must constantly reassure both the child and parent that involution is expected. If the tumor shows unusually rapid growth, hemorrhage, or recurrent infection, biopsy is indicated, and definitive therapy must be initiated. This obviously must be individualized based on several factors, including patient age, site of lesion, depth of extension, and the general characteristics of the mass (Batsakis, 1979). Steroids are often a helpful adjunct to surgical excision, but radiotherapy and sclerosing agents, though often recommended in the past, are generally avoided. The steroids are felt to interrupt proliferation for several possible reasons, including blockage of estradiol receptors or interference with the release of heparin or angiogenic factors from mast cells (Stal et al, 1986).

Teratoma

Teratomas are developmental lesions that contain tissue elements derived from all three germinal layers. The cells found in the lesion may be in any stage of differentiation, and when cells are quite immature, malignancy is suggested. Malignancy is unusual, however, and the histologic changes most likely represent immaturity of the tissue (Day and Arnold, 1971).

Cervical teratomas generally present as a mass in the neck that is covered at birth. It may be seen in stillborn children and rarely presents after the age of 1 year. An in utero diagnosis can be made on ultrasound when a cervical mass is demonstrated that is of mixed echogenicity and displaces the trachea posteriorly. Calcification may be present. There may be some confusion with cystic hygroma, but this mass typically presents as a multiloculated, noncalcified, cystic mass (Gundry et al, 1983).

These lesions are encapsulated and are usually partially cystic, having variegated appearance on cut section. Microscopically, the lesions are composed of a mixture of mature elements derived from ectoderm, mesoderm, and endoderm and of immature or embryonic tissue, including embryonic neuroectoderm. Consequently, most are classified as embryonal teratomas (Day and Arnold, 1971).

Cervical teratomas are sometimes referred to as teratomas of the thyroid gland. They cause symptoms secondary to pressure, and this frequently results in upper airway

compression and obstruction. Patients may present with stridor, cyanosis, and possible apnea. In addition, there may be dysphagia secondary to esophageal compression. Plain neck radiographs reveal a soft tissue mass that contains speckled calcification in approximately 50 per cent of cases; the trachea and the esophagus are displaced posteriorly, and there may be associated pulmonary atelectasis or collapse. On ultrasound, a teratoma is generally of mixed echogenicity and usually can be differentiated from a cystic hygroma, which appears as a multilocular cyst with possible mediastinal extension, or from a congenital goiter, which has a solid appearance (Frech and McAlister, 1969).

Patients do not seem too have an increased incidence of other congenital anomalies, but maternal hydramnios has been incriminated as a predisposing factor. The differential diagnosis is broad and includes cystic hygromas, branchial cysts, cavernous haemangiomas, thyroglossal duct cysts, laryngoceles, goiters, desmoid tumors, and lipomas. Cystic hygromas are generally differentiated by their more cystic appearance and ill-defined margins. Branchial cysts, in contrast, are distinguished on the basis of their size, location, and fluctuance (Day and Arnold, 1971).

Once the diagnosis of a cervical teratoma is made, surgical excision is mandatory to prevent upper airway obstruction or pulmonary compromise. Without intervention, most patients die. Even in those patients who do survive long enough to undergo surgery, there is a mortality rate associated with the condition (Batsakis, 1979).

Muscular Anomalies

Congenital muscular torticollis may be noted at birth or may become evident shortly thereafter. A child present with a nontender, fibrous mass that is palpable in the substance of the sternocleidomastoid muscle. The mass results in tilting of the head towards the side of the shortened muscle and rotation of the chin toward the opposite side.

The condition results from fibrosis within the sternocleidomastoid muscle with subsequent contraction and shortening of the muscle fibers. One study has suggested venous occlusion as the cause of the fibrosis.

Once the diagnosis is made, the muscle must be passively stretched at least four to six times daily in order to prevent developmental asymmetry of the face and ocular imbalance. If this conservative management fails, surgery is appropriate. Surgical management involves division or partial excision of the sternal and clavicular heads of the sternocleidomastoid muscle and postoperative splinting of the neck (Karmody, 1983).

Branchial Anomalies

In order to understand the concept of branchial cysts, sinuses, and fistulas, an appreciation of the embryologic development of the brachial apparatus is important. The branchial apparatus was described initially by von Baer in 1827. In 1832, cervical fistulas associated with abnormalities in embryologic development were reported by von Ascherson. The term *branchial fistulae* was coined in 1864, and at that time it was felt that all lateral cervical cysts were secondary to abnormalities in embryologic development. Wenglowski questioned this concept in 1912, but most experts now accept the branchiogenic origin of

lateral cervical cysts (Chandler and Mitchell, 1981).

There is a great deal of confusion surrounding the terminology of branchial abnormalities. The anomalies have been classified according to either their location (lateral cervical) or origin (branchial). In describing branchial anomalies, the terms *cleft* and *pouch* should be omitted when describing branchial cysts, sinuses, or fistulas. The branchial cyst is a mucosal or epithelial-line structure with no external or visceral opening. Swelling may occur secondary to retained secretions. A branchial sinus is a tract with or without a cyst, which has an internal or external opening. In comparison, a branchial fistula is a tract that connects the gut to the skin (Chandler and Mitchell, 1981).

Embryology

The branchial apparatus is a group of structures phylogenetically related to the gill slits, which develops between the third and seventh weeks of embryonic life. There are five mesodermic arches separated by invaginations of ectoderm (clefts) and endoderm (pouches). Each arch has its own unique arterial and neural supply and eventually develops into skeletal muscle and connective tissue structures. Additionally, each cleft and pouch differentiates into defined structures (Table 1).

Table 1. Derivatives of the Pharyngeal Apparatus

Arch
- Cranial
  - Muscles
    - Cartilage
      - Ligaments
        - Artery
          - Pouch

I (Mandibular)
- Trigeminal (V2, V3)
  - Muscles of mastication (temporalis, masseter, pterygoids)
    - Malleus, Incus
      - Anterior ligament of malleus, Sphenomandibular lig
        - Maxillary artery
          - D: Eustachian tube, Middle ear cavity
          - E: Obliterated by tongue
          - Site of origin of thyroglossal duct

II (Hyoid)
- Facial (VII)
  - Muscles of facial expression:
  - Posterior digastric
  - Stylohyoid
  - Stapedius
  - Auricular
    - Stapes

Styloid process
Lesser horn of hyoid
Upper body of hyoid
Stylohyoid ligament
Stapedial artery
D: Fossa of palatine tonsil
V: Obliterated by tongue

III
Glossopharyngeal (IX)
Stylopharyngeus
Upper pharyngeal
Greater horn of hyoid
Upper body of hyoid
-
Proximal third of internal carotid artery
D: Inferior parathyroid gland
V: Thymus

IV
Superior laryngeal of vagus (X)
Pharyngeal constrictors
Levator veli palatini
Cricothyroid
Upper thyroid cartilage
Arytenoid cartilages
Corniculate cartilages
Cuneiform cartilages
-
Left: aortic arch
Right: proximal subclavian artery
D: Superior parathyroid gland
V: Lateral thyroid gland (?)
Vestigial thymus gland (?)

V
-
See IV
-
-
-
Ultimobranchial body
Parafollicular cells of thyroid

VI
Recurrent laryngeal of vagus (X)
Laryngeal muscles (except cricothyroideus)
Lower thyroid cartilages

-

Bilateral: proximal pulmonary arteries
Left: ductus arteriosus
None

Postbranchial region
Spinal accessory (XI)(communicates with X)
Inferior pharyngeal constrictors
Trapezius
Sternocleidomastoid
Tracheal cartilages
-
-
-
-

D, definitive; V, vestigial.

The four pharyngeal clefts are separated by the pharyngeal arches in the 5-week embryo. The second arch grows caudally during development to cover the third and fourth arches and the second, third, and fourth pharyngeal clefts, eventually fusing with the lower neck. The buried clefts persist as cavities lined by ectoderm and generally disappear with development. If this does not occur, a branchial cyst, sinus, or fistula may develop. To summarize, a branchial sinus represents a vestigial branchial pouch or cleft; the branchial fistula contains remnants of both the pouch and the cleft, with rupture of their interposed branchial plate; the branchial cyst is formed from entrapped remnants of either branchial clefts or pouches without complete sinus tracts (Telander and Deane, 1977).

Most branchial arch cartilages disappear except for those parts that contribute to the bony structures or ligaments. In rare circumstances, however, remnants of the arch cartilages persist as small masses deep to the skin along the anterior border of the sternocleidomastoid muscle. Such growths present only a minor cosmetic problem and are easily removed surgically (Karmody, 1983).

Clinical Presentation

A branchial cyst presents as a smooth, round, nontender fluctuant mass between the level of the tragus and the clavicle along the anterior border of the sternocleidomastoid muscle in most cases. Males and females are affected equally, and patients with such anomalies usually do not present to the physician until the second to fourth decades of life. Secondary enlargement is common during periods of upper respiratory tract infection. Depending on the size of the mass, the cyst may produce dyspnea, dysphagia, dysphonia, and cosmetic deformity. In some situations, abscess formation may occur with either permanent sinus tracts or recurrent infections.

Branchial sinuses and fistulas present in the neonatal period as a skin pit in the lower third of the neck anterior to the sternocleidomastoid muscle. Boys and girls are affected equally, and the anomalies may be bilateral in up to 33 per cent of cases. The skin pits often

have a mucoid drainage that may become purulent during periods of upper respiratory tract infection. Many lesions contain salivary tissue, sebaceous glands, and cholesterol clefts with a foreign body reaction (Chandler and Mitchell, 1981).

Histopathological Features

Branchial anomalies are typically lined by respiratory or squamous epithelium. During episodes of infection, inflammatory cells are commonly seen. In addition, lymphoid tissue is often present beneath the epithelium. The differential diagnosis for branchial anomalies is extensive and includes cystic hygroma, benign lymphadenopathy, metastatic cervical lymphadenopathy, lymphoma, carotid body tumor, hemangioma, neurofibroma, thyroglossal duct cyst, plunging ranula, ectopic salivary tissue, and ectopic thyroid tissue (Chandler and Mitchell, 1981).

Special Considerations

As mentioned previously, knowledge of embryologic development and its related anatomy is essential for understanding branchial cysts, sinuses, and fistulas.

Patients with first branchial abnormalities usually present with unilateral painless swelling in the region of the parotid gland. The cysts have been classified by Work (1977) as types I and II and will be discussed within this framework. Type I branchial defects are duplication anomalies of the external auditory canal, which exist as fistulous tracts near the lower portion of the parotid gland in close association with the facial nerve. They commonly present as sinus tracts or localized areas of swelling near the postauricular sulcus or concha or anterior to the tragus. These anomalies commonly course through the infratemporal fossa parallel to the external auditory canal or middle ear space, though this is relatively uncommon. Complete surgical removal is curative. The cyst is commonly lined by squamous epithelium without skin appendages. If the lesion extends into the parotid gland, the facial nerve must be identified and preserved during surgical resection.

The less commonly encountered type II defect usually presents as a superficial cyst or sinus in the anterior triangle of the neck below the angle of the mandible. This represents an anomalous external auditory canal and a rudimentary pinna. In contrast to type I cysts, type II cysts are composed of both ectoderm (squamous epithelium, hair follicles, sweat glands, sebaceous glands) and mesoderm (cartilage). These anomalies usually course superiorly and posteriorly through the parotid gland in close approximation to the facial nerve and frequently terminate laterally in the region of the bony-cartilaginous junction of the external auditory canal. In some circumstances, the fistulous tract will end in the middle ear space. Abscess formation is common, and complete surgical excision should be attempted only during an asymptomatic period. Again, identification of the facial nerve is crucial during the surgical excision. In general, first branchial anomalies are relatively uncommon and, consequently, they may be mistaken for a second branchial anomaly. Such an error in diagnosis should have disastrous results if unrecognized, since identification of the facial nerve is imperative in the surgical resection of first branchial anomalies.

Second branchial anomalies usually present along the anterior border of the sternocleidomastoid muscle in its lower third. There is frequently a pinpoint external opening

from which there is a mucoid discharge. A tract commonly extends into the pharynx, entering anywhere from the nasopharynx to the hypopharynx but most commonly in the region of the tonsillar fossa. The tract usually passes between the second and third arch structures - that is, it passes medially between the internal and external carotid arteries above the glossopharyngeal nerve and below the stylohyoid ligament. The second branchial cyst is most commonly located at the level of the carotid bifurcation lateral to the internal jugular vein.

Similar to second branchial anomalies, third branchial anomalies present along the anterior border of the sternocleidomastoid muscle in the lower third of the neck. The tract of the third branchial anomaly passes behind the internal and external carotid arteries and the glossopharyngeal nerve while coursing over the hypoglossal and superior laryngeal nerves. It may enter the pharynx at the level of the thyrohyoid membrane or the pyriform sinus.

Fourth branchial anomalies are theoretically possible but have not been conclusively demonstrated. It is theorized that these anomalies present as sinus tracts in the anterior triangle in a fashion similar to that of second and third branchial anomalies. If they exist, their tract passes below the arteries of the fourth arches (aortic arch, right subclavian artery) into the mediastinum and then continues superiorly along the ascending portion of the recurrent laryngeal nerve to enter the upper part of the esophagus. This tract would cross the hypoglossal and spinal accessory nerves.

Management

Surgical excision is the treatment of choice for all branchial anomalies, though this is generally delayed until infants are at least 3 months of age. Infection should be controlled before surgery is attempted. When an abscess is present, incision and drainage will be necessary, as will antimicrobial agents. With extensive lesions, a "stepladder" incision may be employed to limit surgical scarring. An external approach should be used in all circumstances, and potential complications include injury to nearby vascular structures or neural structures. Recurrence is uncommon (3 per cent) unless there has been prior surgery or infection, in which case there is a 20 per cent recurrence rate. Radiographic studies are rarely necessary prior to initiating therapy (Chandler and Mitchell, 1981; Telander and Deane, 1977).

Thyroglossal Duct Cysts

Embryology

The thyroid gland originates from the foramen cecum at the floor of the pharyngeal gut on day 17 of gestational development. The thyroid gland then descends in front of the pharynx as a patent bilobed diverticulum, the thyroglossal duct. It reaches its normal position by about the seventh week of embryonic life and the thyroglossal duct usually disappears by the tenth week. Persistence of a portion of the duct with its epithelium will result in a cystic lesion filled with colloidlike material. The hyoid bone, which originates from the second and third arches, is intimately involved with the thyroglossal duct. As a result, the duct can be located anterior to, in the substance of, or behind the hyoid bone (Telander and Deane, 1977).

Clinical Presentation

The thyroglossal duct cyst is the most common benign cervical mass except for benign cervical lymphadenopathy. Most of these cysts are present before the patient is 5 years of age as a cystic midline neck mass that elevates with protrusion of the tongue. The cyst may be located anywhere along the course of the duct and commonly overlies the hyoid bone (67 per cent of cases). The lesions are usually in the midline but may be located laterally as well. There is no external sinus present unless the cyst has previously drained, either spontaneously or surgically. In some situations, the cyst may drain intraorally. Enlargement of the cyst commonly follows periods of upper respiratory tract infection. The differential diagnosis of such a mass should include cervical lymphadenopathy, dermoid cyst, lipoma, hemangioma, thyroid nodule, ectopic salivary tissue, plunging ranula, branchial cyst, ectopic thyroid tissue, and sebaceous cyst (Noyek and Friedberg, 1981; Telander and Deane, 1977).

Histopathologic Features

A thyroglossal duct cyst usually appears as a simple primary ductal structure connected to a cyst in passing up to the foramen cecum. It is in intimate association with the hyoid bone, and there may be several accessory ducts present. The ducts are lined with stratified squamous epithelium or ciliated pseudostratified epithelium, and there may be mucous glands or ectopic thyroid tissue present. In some patients, the thyroid acini may be found in the connective tissue outside the cyst wall or duct lining (Noyek and Friedberg, 1981).

Surgical Management

A high recurrence rate was noted with surgical excision of the thyroglossal duct when only local excision was employed. Schlange (1893) proposed inclusion of a central portion of the hyoid bone with the cystic specimen. When this was done, the recurrence rate dropped to 20 per cent. Sistrunk (1920) recommended inclusion of a block of tissue at the tongue base along with the specimen (including the hyoid bone). This technique, which is currently used by most surgeons, is associated with only a 3 per cent recurrence rate (Telander and Deane, 1977).

Special Considerations

There have been several reports of carcinomas that were thought to arise in a thyroglossal duct cyst. The majority were papillary thyroid carcinomas, but squamous cell and papillary-follicular thyroid carcinomas have also been reported. It is unclear whether they represent primary lesions or metastatic lesions from the thyroid gland. Surgical management must include a Sistrunk procedure as well as removal of any involved lymph nodes. Controversy exists regarding whether a thyroidectomy should be done in addition to the local excision. However, most physicians recommend adjunctive thyroid suppression in cases of papillary carcinoma.

Thymic Masses

A thymic mass must be considered in the evaluation of any cystic mass in the lower aspect of the neck of an infant. Patients usually present with a firm, round mass in the supraclavicular area with an associated cough and, possibly, respiratory distress.

A radiograph of the chest may be especially helpful, since it will commonly reveal a smooth density or mass filling the anterior mediastinum. Surgical extirpation is best accomplished with the assistance of a general surgeon who is capable of removing the anterior mediastinal component of the disease process. If the mass is rapidly growing, respiratory distress may be quite severe and prompt immediate surgical exploration. In addition to the thymic cyst, one must consider the possibility that such a mass represents hypoplasia of the thymus, a thymoma, or, rarely, thymic lymphosarcoma (Johnson and Cantrell, 1987).

Dermoid Cysts

A dermoid cyst often occurs as a submental mass or midline cervical mass that is usually mobile. In contrast to thyroglossal duct cysts, these cysts do not move with tongue protrusion. They are composed of elements from all three germinal layers, including appendages such as hair follicles, sweat glands, and sebaceous glands. Total excision is necessary to prevent recurrence (Work, 1977).

Laryngoceles

Laryngoceles are unusual disorders of the larynx that may present as masses in the neck. The concept of an air-containing tumor in the neck was introduced by Larrey in 1829, and the term *laryngocele* was coined by Virchow in 1867 (Donegan et al, 1980).

A laryngocele most likely represents an abnormal dilatation of the laryngeal saccule, which is in contiguity with the laryngeal lumen and is thus filled with air. As an extension of the laryngeal saccule, a laryngocele extends upward between the false vocal cord and the inner aspect of the thyroid cartilage. It is lined by normal respiratory epithelium, which contains mucus-secreting glands that are responsible for laryngeal lubrication. If the opening of the laryngocele becomes obstructed, the structure may temporarily become distended with mucus and possibly become infected, thus becoming a laryngopyocele (Rutka and Birt, 1983).

If a laryngocele extends posteriorly into the false vocal cord, the mass will be confined within the larynx, and no cervical swelling will be noted. However, if the laryngocele extends superiorly through the thyrohyoid membrane, an external laryngocele will be noted as a mass lateral to the thyroid ala. In some circumstances, both internal and external components will be present (Baker et al, 1982).

Although laryngoceles may not be evident until the fifth or sixth decade of life, they may be congenital or acquired. They are more commonly seen in males, and in the acquired type probably result from weakening of the laryngeal tissues through the aging process. Other factors that may precipitate development of laryngoceles includes circumstances that increase intralaryngeal pressure, such as coughing, straining, or blowing wind instruments. In children

the opening between the larynx and saccule may become obstructed, thus leading to the formation of a saccular cyst. Saccular cysts are mucus-filled and are actually more common in children than are laryngoceles (Baker et al, 1982).

Although many laryngoceles and saccular cysts are asymptomatic, the most common clinical manifestations are hoarseness, cough, and the sensation of a foreign body in the larynx. When there is a large cervical component present, the patients complain of a cervical mass or cervical emphysema. The diagnosis of an internal laryngocele or saccular cyst can be made with an endoscopic examination. This typically reveals a mucosally covered cystic mass filling the false vocal cord area or aryepiglottic fold, or both. Soft tissue radiographs of the neck or a CT scan may provide helpful information before surgical excision is attempted. Compression of a cervical mass will oftentimes produces a hissing sound in the larynx, as there is escape of air from the laryngocele. This should be avoided when there is an internal component present, since there may be compensatory enlargement of the laryngeal portion of the laryngocele and subsequent airway obstruction (Baker et al, 1982).

Some congenital laryngoceles require no therapy and may resolve spontaneously. Because of this, an affected patient should be monitored carefully for respiratory distress, abnormal cry, feeding difficulties, or cyanosis before deciding upon surgical intervention. As mentioned previously, saccular cysts are more common than laryngoceles in children. These lesions do not have an external opening and may not decompress spontaneously. As a result, surgical therapy is often required. If a lesion is confined within the larynx, an endoscopic approach may be used to excise smaller lesions and to marsupialize larger ones. However, external laryngoceles or saccular cysts should be removed via an external approach. A temporary tracheotomy may be necessary in some instances. One should attempt to preserve all neurovascular structures, including the superior laryngeal nerves and vessels. If a patient presents with a laryngopyocele, incision and drainage of the abscess as an initial step is appropriate, in addition to the administration of intravenous antimicrobial agents. A staged excision may be performed at a later date (Donegan et al, 1980).