

Congenital Cervical Cysts, Sinuses and Fistulae

Stephanie P. Acierno, MD, MPH,
John H.T. Waldhausen, MD*

*Department of Surgery, Children's Hospital and Regional Medical Center,
University of Washington School of Medicine, G0035,
4800 Sand Point Way, NE, Seattle, WA 98105, USA*

Congenital cervical cysts, sinuses, and fistulae must be considered in the diagnosis of head and neck masses in children and adults. These include, in descending order of frequency, thyroglossal duct cysts, branchial cleft anomalies, dermoid cysts, and median cervical clefts. A thorough understanding of the embryology and anatomy of each of these lesions is necessary to provide accurate preoperative diagnosis and appropriate surgical therapy, which are essential to prevent recurrence. The following sections review each lesion, its embryology, anatomy, common presentation, evaluation, and the key points in surgical management.

Thyroglossal duct anomalies

Thyroglossal duct anomalies are the second most common pediatric neck mass, behind adenopathy in frequency [1]. Thyroglossal duct remnants occur in approximately 7% of the population, although only a minority of these is ever symptomatic [1].

Embryology

The thyroid gland forms from a diverticulum (median thyroid anlage) located between the anterior and posterior muscle complexes of the tongue at week 3 of gestation. As the embryo grows, the diverticulum is displaced caudally into the neck and fuses with components from the fourth and fifth

* Corresponding author.

E-mail address: john.waldhausen@seattlechildrens.org (J.H.T. Waldhausen).

branchial pouches (lateral thyroid anlagen). The descent continues anterior to or through the hyoid bone with the median anlage elongating into the thyroglossal duct (Fig. 1) [2]. By weeks 5 to 8 of gestation, the thyroglossal duct obliterates, leaving a proximal remnant, the foramen cecum, at the base of the tongue and a distal remnant, the pyramidal lobe of the thyroid [1,2]. If the duct fails to obliterate before the formation of the mesodermal anlage of the hyoid bone, it persists as a cyst [2].

Clinical presentation and diagnosis

Two thirds of thyroglossal duct anomalies are diagnosed within the first 3 decades of life, with more than half being identified before age 10 years [1]. The most common presentation is that of a painless cystic neck mass near the hyoid bone in the midline (Figs. 2 and 3) [2]. Although they are most commonly found immediately adjacent to the hyoid (66%), they can also be located between the tongue and hyoid, between the hyoid and pyramidal lobe, within the tongue, or within the thyroid [2,3]. The mass usually moves with swallowing or protrusion of the tongue. Approximately one third present with a concurrent or prior infection, which is the more common presentation in adults [2,4]. One fourth of patients present with a draining sinus that results from spontaneous drainage or surgical drainage of an abscess [2]. This drainage can result in a foul taste in the mouth if the spontaneous drainage occurred by way of the foramen cecum. These lesions also fluctuate in size. Other rare presentations can be severe respiratory distress or sudden infant death syndrome from lesions at the base of the tongue, a lateral cystic neck mass, an anterior tongue fistula, or coexistence with branchial anomalies [2].

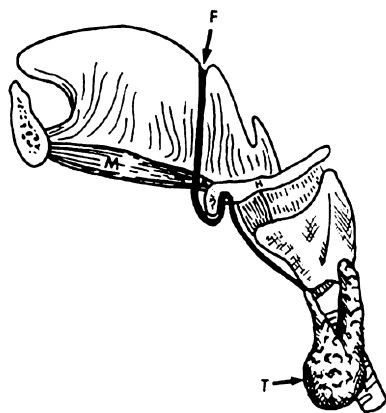


Fig. 1. The course of the thyroglossal duct extending from the foramen cecum (F) to the thyroid (T). (From Som PM, Smoker WRK, Curtin HD, et al. Congenital lesions in head and neck imaging. In: Som PM, Curtin HD, editors. Head and neck surgery. St. Louis: Mosby; 2003. p. 121–5; with permission.)



Fig. 2. Thyroglossal duct cyst, uncomplicated. (*From* Foley DS, Fallat ME. Thyroglossal duct and other congenital midline cervical anomalies. *Semin Pediatr Surg* 2006;15: 70–5; with permission.)

The preoperative evaluation for a patient who has a suspected thyroglossal duct cyst includes a complete history and physical examination, preoperative ultrasound, and a screening thyroid stimulating hormone (TSH) level. Patients who have history, examination findings, or elevated TSH levels suggesting hypothyroidism or a solid mass should undergo scintiscanning to rule out a median ectopic thyroid [2]. When median ectopic thyroid is present, all of the patient's functional thyroid tissue can be located within the cyst, and its removal would render the patient permanently dependent on thyroid replacement. The management of median ectopic thyroid is controversial. Some investigators believe these patients can be treated with exogenous thyroid hormone to suppress the gland, whereas others advocate for resection for reasons that are discussed later [2]. Although median ectopic thyroid only occurs in 1% to 2% of thyroglossal duct cysts, some authors advocate for scintiscans in all patients [5].

Treatment

Elective surgical excision is the treatment of choice for uncomplicated thyroglossal duct cysts to prevent infection of the cyst. The Sistrunk

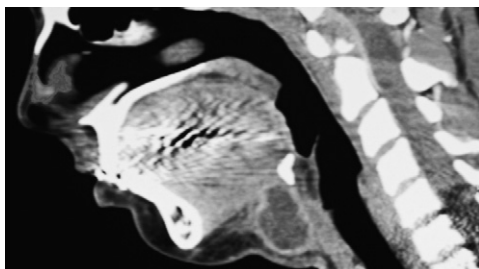


Fig. 3. Sagittal CT reconstruction of a thyroglossal duct cyst demonstrating the close relationship to the hyoid bone. (*Courtesy of* Glenn Isaacson, MD, Philadelphia, PA.)

procedure is performed, rather than simple excision, to reduce recurrence risk [2]. With the patient in supine position and the neck extended, a transverse incision is made over the mass. The dissection is carried down to the cyst, then caudally to identify the tract to the pyramidal lobe. If present, it is excised en bloc with the cyst. The surgeon then dissects cranially toward the hyoid bone and a block of tissue around the proximal tract is also excised. The central portion of the hyoid bone is also excised and the tract is further dissected with a core of tissue from the muscle at the base of the tongue to the foramen cecum (Fig. 4) [2]. After confirming adequate proximal dissection by pressure on the base of the tongue from the mouth, the tract is ligated and transected. Intrathyroidal thyroglossal duct cysts should also undergo a Sistrunk procedure if there is a transhyoid fistulous tract, but can be treated with hemi-thyroidectomy if no tract can be identified [3].

Infected cysts or sinuses are first managed by relieving the infection. The cysts are usually infected by way of the mouth, thus the most common organisms are *Haemophilus influenza*, *Staphylococcus aureus*, and *Staphylococcus epidermidis* [2]. Antibiotics directed toward those organisms should be started. Needle aspiration may allow for decompression and identification of the organism. Formal incision and drainage should be avoided, if possible, to prevent seeding of ductal cells outside the cyst, which increases recurrence [2]. If incision and drainage is necessary, the incision should be placed so it can be completely excised with an ellipse at the time of definitive resection. Once the infection clears and the incision heals, the patient may undergo an elective Sistrunk procedure [2].

If a solid mass is encountered during excision of a suspected thyroglossal duct cyst, it should be sent for frozen section to rule out a median ectopic thyroid. If the biopsy returns as normal thyroid tissue and the patient has functional thyroid tissue in the normal location, it should be excised by

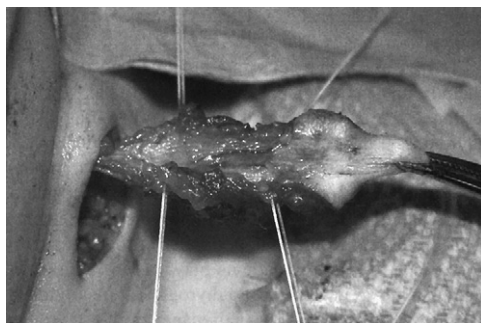


Fig. 4. Sistrunk procedure resection of a thyroglossal duct cyst; note that the specimen includes the cyst, hyoid bone, and proximal tract en bloc. (From Foley DS, Fallat ME. Thyroglossal duct and other congenital midline cervical anomalies. *Semin Pediatr Surg* 2006;15:70–5; with permission.)

the Sistrunk procedure [2]. If the mass is possibly the patient's only functional thyroid tissue, the management becomes controversial. One option involves leaving the ectopic thyroid, either in situ or repositioning it laterally below the strap muscles or into the rectus abdominus or quadriceps muscles. This option aims to not render the patient permanently hypothyroid; however, most patients still require long-term thyroid hormone therapy to treat hypothyroidism or control the size of the ectopic thyroid tissue for cosmetic or functional reasons. This need for long-term therapy and the possibility of malignant degeneration have led some to recommend excision of the median ectopic thyroid regardless of the presence of additional thyroid tissue [2].

Thyroglossal duct cysts are lined with ductal epithelium or contain solid thyroid tissue. Less than 1.0% have malignant tissue, usually well-differentiated thyroid carcinoma. This malignancy occurs more often in adults, but has been reported in children as young as 6 years old [6]. It is usually identified incidentally at the time of surgery for a suspected thyroglossal duct cyst. Papillary carcinoma is seen most often, although all types of thyroid carcinoma except medullary carcinoma have been reported [2,4]. If there is no evidence of capsular invasion or distant or regional metastasis, the Sistrunk procedure has been associated with a 95% cure rate, although careful follow-up is necessary [2]. Other investigators recommend completion thyroidectomy regardless of capsular invasion citing the benefits of full pathologic examination of the gland, facilitation of radioactive iodine ablation, and increased sensitivity of radioisotope screening for recurrence [1]. If capsular invasion is present, completion thyroidectomy, nodal dissection, and radioiodine ablation should be pursued as indicated by type and stage of disease [2].

Recurrence of thyroglossal duct cyst after complete excision using the Sistrunk procedure is reported to be 2.6% to 5% [1,4]. Several factors have been identified predisposing patients to increased risk for recurrence. Failure to completely excise the cyst (especially simple excision alone) can result in recurrence rates of 38% to 70% [1,4]. In children less than 2 years old, intraoperative cyst rupture and presence of a cutaneous component increases the risk for recurrence. Preoperative or concurrent infection of the cyst has been historically reported as a risk factor because of the increased difficulty of complete resection, although a recent review found that postoperative infections rather than preoperative infections were associated with increased recurrence [2,7]. Recurrent thyroglossal duct excision has a higher risk for recurrence (20%–35%) and requires a wider en bloc resection [2].

Branchial cleft anomalies

Branchial anomalies compose approximately 30% of congenital neck masses and can present as cysts, sinuses, or fistulae [1,8]. They are equally common in males and females and usually present in childhood or early adulthood.

Embryology

By the end of the fourth week of gestation, there are four well-defined pairs of arches and two rudimentary arches. These are lined externally by ectoderm, internally by endoderm, with mesoderm in between. The mesoderm contains the dominant artery, nerve, cartilage rod, and muscle for each arch. Each arch is separated by clefts externally and pouches internally. In fish these structures form gills, but in humans the clefts and pouches are gradually obliterated by mesenchyme to form the mature head and neck structures (Fig. 5) [8]. Branchial anomalies result from incomplete obliteration of the clefts and pouches.

Each arch transforms throughout gestation into a defined anatomic pattern. Knowledge of this pattern of transformation and its relationship to normal structures in the neck is essential in the diagnosis and treatment of these anomalies. Branchial anomalies are classified by the cleft or pouch of origin and this is determined by the internal opening of the sinus and its relationship to nerves, arteries, and muscles. A thorough understanding of these relationships is needed to prevent injury to surrounding structures and ensure complete resection [8]. The essential embryology and anatomy is described for each cleft in later discussion.

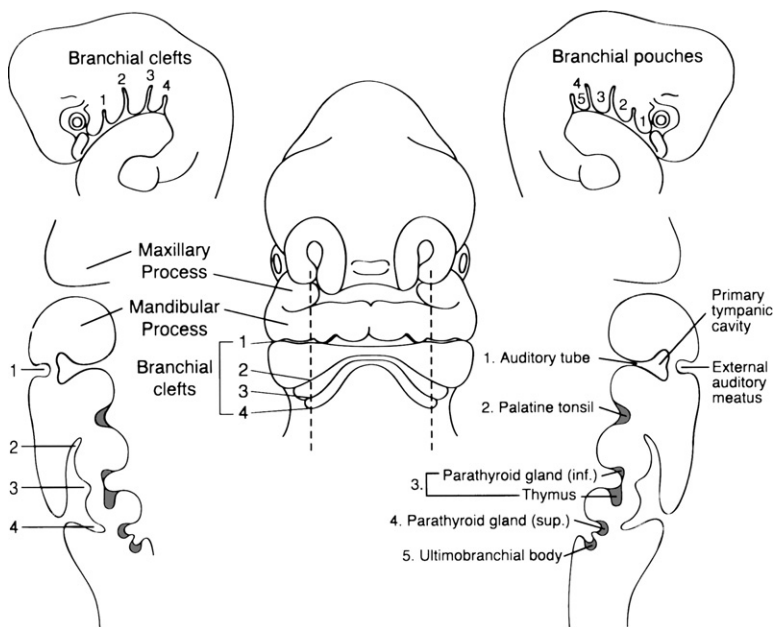


Fig. 5. Branchial embryology at the fifth week of gestation. Sagittal sections demonstrate the anatomic relationship of the external clefts and internal pouches and the derivation of important head and neck structures. (From Waldhausen JHT. Branchial cleft and arch anomalies in children. *Semin Pediatr Surg* 2006;15:64-9; with permission.)

Pathology

Branchial anomalies can be lined with either respiratory or squamous epithelium. Cysts are often lined by squamous epithelium, whereas sinuses and fistulae are more likely to be lined with ciliated, columnar epithelium [8]. Lymphoid tissue, sebaceous glands, salivary tissue, or cholesterol crystals in mucoid fluid can also be seen. Squamous cell cancer can be found within branchial lesions in adults, although it is rare. It is difficult to distinguish between a primary lesion arising from within an anomaly and a metastatic lesion from an occult primary [1].

Diagnosis

Branchial anomalies can present as cysts, sinuses, or fistulae. Cysts are remnants of the cervical sinus without an external opening. Sinuses are the persistence of the cervical sinus with its external opening, whereas a fistula also involves persistence of the branchial groove with breakdown of the branchial membrane resulting in a pharyngocutaneous fistula [1]. The specific presentation for each cleft is described in later discussion.

The evaluation of these lesions begins with a complete history and physical, which may include upper airway endoscopy to locate the pharyngeal opening. The pyriform sinus and the tonsillar fossa should be carefully examined. In adults, fine needle aspiration should be performed to rule out metastatic carcinoma or clarify the diagnosis [8]. This clarification is not necessary in children and incisional biopsy should not be performed because this makes the resection more difficult. Ultrasound, CT, and MRI can be used to help define the lesion and its course, but CT is the current study of choice. Current tomography is able to demonstrate the fistula in up to 64% of cases [9]. Barium esophagram can also be helpful with a 50% to 80% sensitivity for third and fourth branchial fistulae [10].

Treatment

The definitive treatment of all branchial anomalies is complete surgical excision. Unresected cysts and sinuses have a high risk for infection and incomplete resection results in high rates of recurrence [8]. Timing of resection is controversial with some advocating for early resection to prevent infection whereas others support waiting until age 2 to 3 years [8,11,12]. Twenty percent of lesions have been infected at least once before the time of surgery [11]. As with thyroglossal duct cysts, acute infections should first be treated with antibiotics, needle aspiration, and, if necessary, incision and drainage, followed by complete resection after resolution of the infection. Specific considerations for the resection of each type of anomaly are discussed later.

First cleft anomalies

First branchial cleft anomalies account for only 1% of branchial cleft malformations [8]. The first arch, or mandibular arch, forms the mandible, part of the maxillary process of the upper jaw, and portions of the inner ear. The first cleft and pouch form the external auditory canal, eustachian tube, middle ear cavity, and mastoid air cells. First cleft anomalies may involve either the external auditory canal or, occasionally, the middle ear [8]. First cleft anomalies course close to the parotid gland, especially the superficial lobe, traveling above, between, or below the facial nerve branches. First cleft anomalies are classified as Type I or Type II (Figs. 6 and 7). Type I lesions are duplications of the membranous external auditory canal, are composed of ectoderm only, course lateral to the facial nerve, and present as swellings near the ear. Type II lesions have ectoderm and mesoderm, can contain cartilage, pass medial to the facial nerve, and present as preauricular, infra-auricular, or postauricular swellings inferior to the angle of the mandible or anterior to the sternocleidomastoid muscle [1,8].

First cleft anomalies can present as cysts, sinuses or fistulae located between the external auditory canal and the submandibular area. They are more common in females than males and are often misdiagnosed leading to a delay in excision [13,14]. Ten percent have an asymptomatic membranous attachment from the floor of the external auditory canal to the

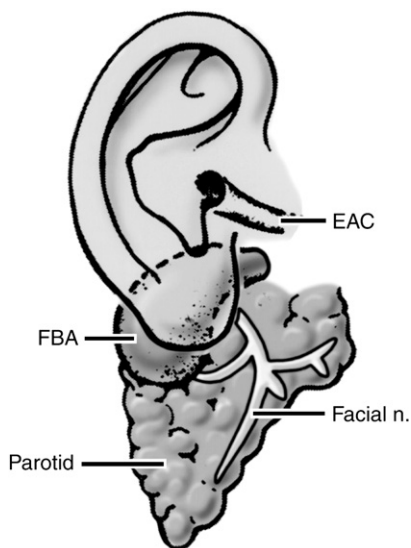


Fig. 6. Type I first branchial cleft anomaly. Note that the cyst (FBA) is located within the parotid gland and does not connect to the external auditory canal (EAC). (From Mukherji SK, Fatterpekar G, Castillo M, et al. Imaging of congenital anomalies of the branchial apparatus. *Neuroimaging Clin N Am* 2000;10:75–93; with permission.)

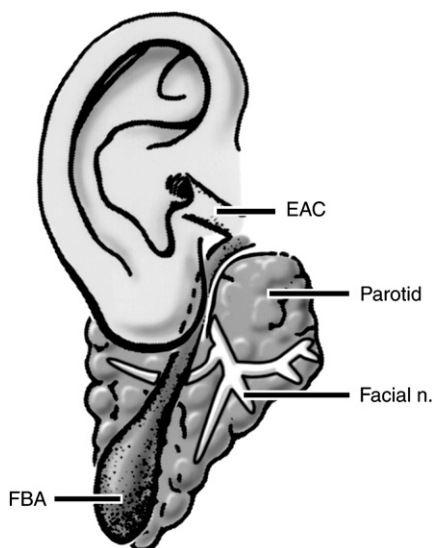


Fig. 7. Type II first branchial cleft anomaly. Note that the cyst (FBA) communicates with the external auditory canal (EAC) and extends into the deep lobe of the parotid gland. (From Mukherji SK, Fatterpekar G, Castillo M, et al. Imaging of congenital anomalies of the branchial apparatus. *Neuroimaging Clin N Am* 2000;10:75–93; with permission.)

tympanic membrane [8,13]. Presentations vary, but include cervical, parotid, or auricular signs. Cervical signs consist of drainage from a pit-like depression at the angle of the mandible, which can be purulent if infected. Parotid involvement is likely if there is rapid enlargement because of inflammation. Auricular signs include swelling or otorrhea.

The surgical resection of first arch anomalies often requires at least partial facial nerve dissection and superficial parotidectomy. It is also necessary to excise any involved skin or cartilage of the external auditory canal. If the tract extends medial to the tympanic membrane, it may be necessary to transect the tract and remove the medial portion during a second procedure. Compared with tracts that go to the external auditory canal, tracts going to the middle ear tend to lie deep to the facial nerve [9]; however, tracts can split around the nerve [13]. Recurrence is common, with the average number of procedures required to achieve complete resection being 2.4 per patient [15]. Each repeat surgery has an increased risk for injury to the facial nerve because of previous scarring, indicating the importance of complete resection at the first attempt when possible [8].

Second cleft anomalies

Second branchial cleft anomalies are the most common, representing 95% of all brachial cleft malformations. The second arch, or hyoid arch,

forms the hyoid bone and adjacent areas of the neck. The second pouch gives rise to the tonsillar and supratoronsillar fossae. Second cleft anomalies thus enter the supratoronsillar fossa [8]. These anomalies pass close to the glossopharyngeal and hypoglossal nerves on their course to the fossa. Second arch anomalies are classified into four types as demonstrated in Fig. 8. Type I lesions lie anterior to the sternocleidomastoid muscle (SCM) and do not contact the carotid sheath. Type II lesions are the most common and pass deep to the SCM and either anterior or posterior to the carotid sheath. Type III lesions pass between the internal and external carotid arteries and are adjacent to the pharynx. Type IV lesions lie medial to the carotid sheath close to the pharynx adjacent to the tonsillar fossa.

Second brachial cleft anomalies present as a fistula or cyst in the lower, anterolateral neck. Cysts are most commonly diagnosed in adults during the third and fifth decades as a nontender mass that can acutely increase in size after an upper respiratory infection. The enlargement can lead to

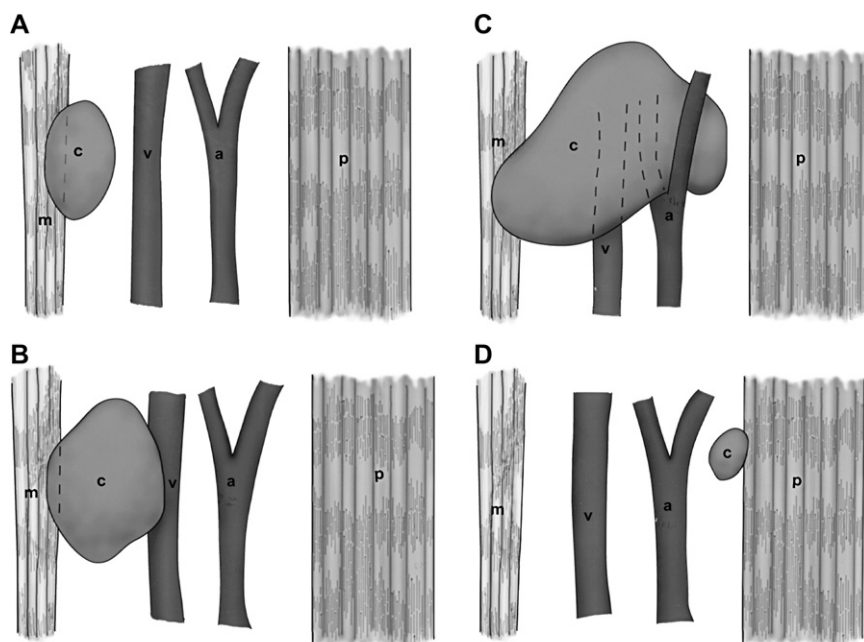


Fig. 8. Types I through IV second branchial cleft anomalies. (A) Type I: the cyst (C) is superficial to the anterior border of the sternocleidomastoid muscle (M). (B) Type II: the cyst is adjacent to the carotid sheath. (C) Type III: the cyst passes between the internal and external carotid arteries and extends to the lateral wall of the pharynx (P). (D) Type IV: the cyst is deep to the carotid sheath abutting the pharynx. (From Mukherji SK, Fatterpekar G, Castillo M, et al. Imaging of congenital anomalies of the branchial apparatus. *Neuroimaging Clin N Am* 2000;10:75–93; with permission.)

respiratory compromise, torticollis, or dysphagia. Fistulae, however, are usually diagnosed in infancy or childhood and present as chronic drainage from an opening along the anterior border of the SCM in the lower third of the neck [1,8].

Surgical resection of second cleft anomalies can be approached by way of a transverse cervical incision placed within a natural skin fold. Cysts can be located either superficially or deep to the cervical fascia. A careful exploration for an associated fistula tract must be performed with a complete excision of the entire tract if one is found. Fistula excision can be facilitated by cannulating the tract with a 2-0 or 3-0 monofilament suture or probe. The tract can also be injected with methylene blue; however this may stain the surrounding tissues making dissection difficult [8]. As the tract is followed, the skin incision may have to be extended to allow adequate exposure, although step-ladder incisions may provide improved visualization of the tract near the pharynx. The spinal accessory, hypoglossal, and vagus nerves must be protected from injury during the dissection. A finger or bougie in the oropharynx can help identify the opening in the tonsillar fossa. The thin tract must be carefully ligated and divided at its entry into the fossa. If the excision is complete, no drain is needed.

Third and fourth cleft anomalies

Third and fourth branchial anomalies are rare. The third and fourth pouches form the pharynx below the hyoid bone, thus these sinuses and fistulae enter into the pyriform sinus. Third and fourth branchial anomalies normally contain thymic tissue as do cysts and sinuses that result from thymic or parathyroid rests, but only branchial anomalies have the connection to the pyriform sinus. Third arch anomalies present as cystic structures located at the lower, anterior border of the SCM, at the level of the superior pole of the thyroid [1,8]. They pass deep to the internal carotid artery and the glossopharyngeal nerve, entering the thyroid membrane above the internal branch of the superior laryngeal nerve, then entering the pyriform sinus of the pharynx (Fig. 9). Third arch cysts can cause hypoglossal nerve palsy if infected. The course of fourth arch anomalies depends on the side (Fig. 10). On the right, the lesion loops around the subclavian artery, passes deep to the internal carotid artery, ascending to the level of the hypoglossal nerve, then descends along the anterior border of the SCM to enter the pharynx at the pyriform apex or cervical esophagus. On the left, the tract descends into the mediastinum, looping around the aortic arch, medial to the ligamentum arteriosus, then ascends in a similar course to the right side. Fourth arch lesions present as lateral cysts in the lower third of the neck [1,8]. Both third and fourth cleft lesions can present at any age. Either can also present with tracheal compression and airway compromise in the neonate because of rapid enlargement in size. Third and fourth branchial cleft cysts can also present as cold nodules in the thyroid leading to confusion with thyroglossal

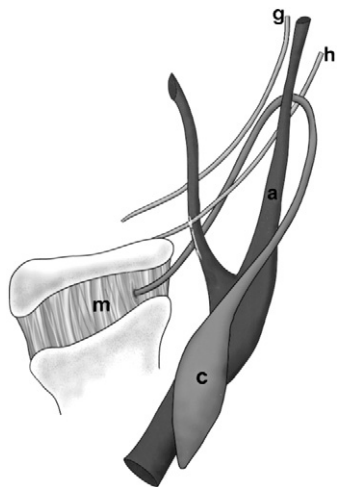


Fig. 9. Third branchial cleft anomaly. The cyst (C) is posterior to the sternocleidomastoid muscle, and the tract ascends posterior to the internal carotid artery. It then passes medially to pass between the hypoglossal (H) and glossopharyngeal (G) nerves. It pierces the thyroid membrane to enter the pyriform sinus. (From Mukherji SK, Fatterpekar G, Castillo M, et al. Imaging of congenital anomalies of the branchial apparatus. *Neuroimaging Clin N Am* 2000;10:75–93; with permission.)

duct cysts. Other possible presentations include recurrent upper respiratory tract infections, neck or thyroid pain, or thyroid abscess.

Surgical therapy of third and fourth arch anomalies is similar to that of second arch anomalies, with the following exceptions. Endoscopy should be used to identify the pyriform sinus entry point. This identification can allow cannulation or injection of the tract to aid with dissection. There are some reports of chemical cauterization of these tracts; however, there are no long-term results for this approach [16]. Fourth arch anomaly resections require ipsilateral hemithyroidectomy to completely excise the tract and possible partial resection of the thyroid cartilage to provide adequate exposure of the pyriform sinus [17].

Branchiootorenal syndrome

Branchiootorenal syndrome (BOR) or Melnick-Fraser syndrome is an autosomal dominant disorder with coinheritance of branchial arch anomalies. It occurs in approximately 2% of profoundly deaf students, with an estimated 1:40,000 to 1:700,000 prevalence [18]. It has been mapped to chromosome 8q 13.3, the human homolog of the *Drosophila* eyes absent gene that has roles in cochlear and vestibular development and renal morphogenesis [18]. The typical phenotype consists of cup-shaped pinnae; preauricular pits; branchial fistulae; conductive, sensorineural, or mixed



Fig. 10. Fourth branchial cleft anomaly. The cysts (C) are located anterior to the aortic arch on either side. The tract hooks either the subclavian artery or the aortic arch, depending on the side, and ascends to loop over the hypoglossal nerve (H). (From Mukherji SK, Fatterpekar G, Castillo M, et al. Imaging of congenital anomalies of the branchial apparatus. *Neuroimaging Clin N Am* 2000;10:75–93; with permission.)

hearing impairment; and renal anomalies ranging from mild hypoplasia to complete absence. Other findings may include preauricular tags, lacrimal duct stenosis, a constricted palate, a deep overbite, and a long, narrow face. Hearing loss and preauricular pits are most common, with branchial cleft fistulae occurring in approximately 50% of individuals [18].

Dermoid cysts and teratomas

Dermoid cysts result from entrapment of epithelial elements along embryonic lines of fusion (median and paramedian) and contain ectodermal and endodermal elements [1]. Dermoids are lined by epithelium but contain epithelial appendages, such as hair, hair follicles, or sebaceous glands [2]. Cervical dermoid cysts represent only 20% of head and neck dermoids [1]. Cervical dermoids present as painless superficial subcutaneous masses in the anterior neck and usually move with the skin. They can be close to the hyoid and move with swallowing or tongue protrusion leading to confusion with thyroglossal duct cysts. They gradually increase in size over time because of accumulation of sebum. Infection is rare, but the cysts can rupture and present with granulomatous inflammation. They are often diagnosed before the patient is 3 years old [1].

Evaluation begins with history and physical examination, but can be augmented with ultrasonography to delineate the depth of the lesion and its

relationship to the hyoid bone. If the lesion is inflamed, fine needle aspirate may be helpful to distinguish between a ruptured dermoid cyst and an infected thyroglossal duct cyst. If the lesion is symptomatic, enlarging, or has ruptured, surgical excision is recommended. Complete simple excision is usually adequate, but if it is attached to the hyoid bone a Sistrunk procedure should be performed to prevent inadequate excision of an atypical thyroglossal duct cyst [19]. Rate of recurrence is increased by incomplete resection or intraoperative rupture [2].

Teratomas differ from dermoid cysts in that they contain all three germ layers. Head and neck lesions compose less than 2% of teratomas, with the most common sites being the nasopharynx and neck. They develop during the second trimester and present as rapidly expanding lateral or midline neck masses. They may be diagnosed by prenatal ultrasonography, with 30% accompanied by polyhydramnios because of esophageal obstruction [1]. If the diagnosis is known before delivery, cesarean section is recommended. Although the lesions may initially be asymptomatic, rapid growth may eventually lead to dysphagia and respiratory distress. Eighty percent of infants who have neonatal teratomas may die if untreated [1]. Ultrasound, CT, or MRI may be helpful in evaluating these lesions. Some neonates may require intubation or even extracorporeal membrane oxygenation if the lesion has caused pulmonary hypoplasia. Complete surgical excision is the treatment of choice once the airway has been stabilized. Malignancy has not been reported in pediatric cervical teratomas, so all critical structures in the neck should be spared [1]. Malignant cervical teratomas have



Fig. 11. Congenital midline cervical cleft. (From Foley DS, Fallat ME. Thyroglossal duct and other congenital midline cervical anomalies. *Semin Pediatr Surg* 2006;15:70–5; with permission.)

been found in adults and require aggressive treatment because they can spread by hematogenous and lymphatic routes and carry a poor prognosis.

Midline cervical clefts

Midline cervical clefts are rare congenital cervical anomalies. They are present at birth as a cutaneous ulceration with overhanging skin or cartilaginous tag in the anterior lower midline of the neck (Fig. 11). There is often a sinus tract that extends downward from the skin and may connect to the sternum or mandible or end in a blind pouch. The embryologic origin is unknown but is believed to be a “mesodermal fusion abnormality involving the paired branchial arches during gestational weeks 3 and 4” [2]. Fibrous tissue with interwoven skeletal muscle is present. Most cases are sporadic, but can be associated with other cleft abnormalities of the tongue, lower lip, or mandible. If untreated, some clefts can result in neck contractures or growth deformities of the mandible or sternum. Early surgical excision at the time of diagnosis is recommended, therefore, with complete excision of the skin lesion and the subcutaneous sinus to reduce the rate of recurrence. This excision can usually be accomplished by stair-step incisions, but if more complicated may require a series of Z-plasty incisions to improve the cosmetic and functional result [2].

References

- [1] Enepekides DJ. Management of congenital anomalies of the neck. *Facial Plast Surg Clin North Am* 2001;9:131–45.
- [2] Foley DS, Fallat ME. Thyroglossal duct and other congenital midline cervical anomalies. *Semin Pediatr Surg* 2006;15:70–5.
- [3] Perez-Martinez A, Bento-Bravo L, Martinez-Bermejo MA, et al. An intra-thyroid thyroglossal duct cyst. *Eur J Pediatr Surg* 2005;15:428–30.
- [4] Mohan PS, Chokshi RA, Moser RL, et al. Thyroglossal duct cysts: a consideration in adults. *Am Surg* 2005;71(6):508–11.
- [5] Pinczower E, Crockett DM, Atkinson JB, et al. Preoperative thyroid scanning in presumed thyroglossal duct cysts. *Arch Otolaryngol Head Neck Surg* 1992;118:985–8.
- [6] Peretz A, Lieberman E, Kapelsuhnik J, et al. Thyroglossal duct carcinoma in children: case presentation and review of the literature. *Thyroid* 2004;14:777–85.
- [7] Ostlie DJ, Burjonrappa SC, Synder CI, et al. Thyroglossal duct infections and surgical outcomes. *J Pediatr Surg* 2004;39:396–9.
- [8] Waldhausen JHT. Branchial cleft and arch anomalies in children. *Semin Pediatr Surg* 2006;15:64–9.
- [9] D'Souza AR, Uppal HS, De R, et al. Updating concepts of first brachial cleft defects: a literature review. *Int J Pediatr Otolaryngol* 2002;62:103–9.
- [10] Shrimel M, Kacker A, Bent J, et al. Fourth branchial complex anomalies: a case series. *Int J Pediatr Otolaryngol* 2003;67:1227–33.
- [11] Roback SA, Telander RL. Thyroglossal duct cysts and branchial cleft anomalies. *Semin Pediatr Surg* 1994;3:142–6.
- [12] O'Mara W, Amedee R. Anomalies of the branchial apparatus. *J La State Med Soc* 1998;150:570–3.

- [13] Triglia JM, Nicollas R, Ducroz V, et al. First branchial cleft anomalies. *Arch Otolaryngol Head Neck Surg* 1998;124:291–5.
- [14] Liberman M, Kay S, Emil S, et al. Ten years experience with third and fourth branchial remnants. *J Pediatr Surg* 2002;37:685–90.
- [15] Ford GR, Balakrishnan A, Evans JN, et al. Branchial cleft and pouch anomalies. *J Laryngol Otol* 1992;106:137–43.
- [16] Park SW, Han MH, Sung MH, et al. Neck infection associated with pyriform sinus fistula: imaging findings. *AJNR Am J Neuroradiol* 2000;21:817–22.
- [17] Nicollas R, Ducroz V, Garabedian EN, et al. Fourth branchial pouch anomalies: a study of six cases and a review of the literature. *Int J Pediatr Otorhinolaryngol* 1998;44:5–10.
- [18] Smith RJH, Schwartz C. Branchio-oto-renal syndrome. *J Commun Disord* 1998;31:411–21.
- [19] Turkyilmaz Z, Sonmez K, Karabulut R, et al. Management of thyroglossal duct cysts in children. *Pediatr Int* 2004;46:77–80.