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Management of common head and neck masses

Thomas F. Tracy Jr, MD, Christopher S. Muratore, MD

From the Division of Pediatric Surgery, Department of Surgery, Brown Medical School, Hasbro Children's Hospital, Providence, Rhode Island.

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Vascular malformation

Head and neck masses are a common clinical concern in infants, children, and adolescents. The differential diagnosis for a head or neck mass includes congenital, inflammatory, and neoplastic lesions. An orderly and thorough examination of the head and neck with an appropriate directed workup will facilitate the diagnosis. The most common entities occur repeatedly within the various age groups and can be differentiated with a clear understanding of embryology and anatomy of the region, and an understanding of the natural history of a specific lesion. Congenital lesions most commonly found in the pediatric population include the thyroglossal duct cyst and the branchial cleft and arch anomalies. The inflammatory masses are secondary to local or systemic infections. The most common etiology for cervical adenopathy in children is reactive lymphadenopathy following a viral or bacterial illness. Persistent adenopathy raises more concerns, especially enlarged lymph nodes within the posterior triangle or supraclavicular space, nodes that are painless, firm, and not mobile, or a single dominant node that persists for more than 6 weeks should all heighten concern for malignancy. In this review, we discuss the current principles of surgical management of the most common head and neck masses that present to pediatricians and pediatric surgeons.
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Head and neck masses are a common clinical concern in infants, children, and adolescents. The differential diagnosis for a head or neck mass across these age groups is broad and includes congenital, inflammatory, and neoplastic lesions (Table 1).[1,2] An orderly and thorough examination of the head and neck with an appropriate directed workup will facilitate the diagnosis. The most common entities occur repeatedly within the various age groups and can be differentiated with a clear understanding of embryology and anatomy of the region, and an understanding of the natural history of a specific lesion. In this review, we discuss the general approach to evaluate and manage the most common head and neck masses that present to pediatricians and pediatric surgeons.

Address reprint requests and correspondence: Thomas F. Tracy Jr, MD, Hasbro Children's Hospital, 593 Eddy Street, Providence, RI 02903.
E-mail: Thomas_Tracy@brown.edu.

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Overview of head and neck masses

Congenital lesions most commonly found in the pediatric population include the thyroglossal duct cyst and the branchial cleft and arch anomalies. Hemangiomas, lymphatic malformations, dermoid cysts, bronchogenic cysts, teratomas, and thymic cysts are other common congenital lesions.[3,4] The inflammatory masses are secondary to local or systemic infections. The most common etiology for cervical adenopathy in children is reactive lymphadenopathy following a viral or bacterial illness. Persistent unilateral adenopathy is concerning and can include acquired etiologies such as mycobacterium tuberculosis, the atypical mycobacterium spectrums such as mycobacterium avium intracellulare, and mycobacterium scrofulaceum, granulomatous processes, or cat scratch disease.[5] The midline lesions most commonly represented by thyroglossal duct sinus and cyst conditions and dermoid cyst are usually easily distinguished from the more lateral lesions represented by branchial cleft sinus and arch

Table 1 Differential diagnosis of pediatric head and neck masses*

Congenital masses	Inflammatory masses	Neoplastic masses
Thyroglossal duct cyst	Reactive lymphadenpathy	Benign (lipoma, fibroma, neurofibroma, thyroid nodule)
Branchial cleft cyst/sinus	Bacterial	Malignant (Hodgkin's lymphoma, non-Hodgkin's lymphoma, rhabdomyosarcoma, neuroblastoma, thyroid carcinoma, metastatic disease)
Vascular anomalies (hemangioma, lymphatic, capillary, venous, arterial, mixed)	Viral	
Dermoid cyst	Granulomatous	
Bronchogenic cyst	Mycobacterial (tuberculous, atypical)	
Teratoma	Histoplasmosis	
	Sarcoidosis	
	Cat scratch disease	

*Modified from Dickson and Davidoff.[1]

anomalies. Acute bilateral or diffuse cervical adenopathy is often the result of a recent viral infection and is usually a self-limited process. Acute unilateral adenopathy, particularly in infants and young children, may be associated with pyogenic sources such as Staphylococcus aureus or group B Streptococcal infections.[2,5,6] Persistent adenopathy raises more concerns but is usually still secondary to an infectious etiology. Enlarged lymph nodes within the posterior triangle or supraclavicular space, nodes that are painless, firm, and not mobile, or a single dominant node that persists for more than 6 weeks should all heighten concern for malignancy.[1] Malignant lesions such as non-Hodgkin's and Hodgkin's lymphoma, neuroblastoma, and thyroid carcinoma need to be differentiated from benign lesions. In the pediatric population, 80% to 90% of all head and neck masses represent benign conditions.[1] It is important, therefore, for surgeons to appreciate the relevant embryology, anatomy, and natural history of head and neck lesions and to be familiar with their appropriate evaluation and management.

Evaluation: History and physical examination

A detailed history and physical examination is the usual starting point. Historical information includes the patient's age, onset, and duration of symptoms, as well as any systemic signs of disease, such as fever, night sweats, fatigue, or weight loss. Although some congenital neck lesions, particularly cysts, may not present until later in childhood after the accumulation of secretions or becoming secondarily infected, many congenital lesions are present at birth or noted shortly thereafter.

Features from the history and examination should help to elicit and narrow the etiology. Specific questions to ask include whether the adenopathy is an acute or chronic process; whether the adenopathy was associated with a recent upper respiratory illness or following contact with an individual with a recent illness; whether the neck masses were associated with a systemic infection; if there had been any known animal bites or scratches; and whether there had been any recent changes in the character of the lesion. The physical examination should be directed at a systematic evaluation of each cervical lymph node region. The size, laterality, tenderness, overlying skin changes, and mobility should be noted. Finally, an examination of the chest, abdomen, groin, genitalia, and extremities must not be forgotten. A firm painless mass with fixation to underlying structures or overlying skin is always concerning for malignancy. Although most pediatric cervical adenopathy is of benign etiology, rapidly enlarging, nontender, or longstanding adenopathy, particularly within the supraclavicular space or posterior cervical triangle, are concerning for malignant disease.[1,2] The experienced clinician will seldom require laboratory evaluation for the classic midline or lateral congenital lesions associated with branchial arch anomalies; however, the workup for persistent adenopathy is more extensive and should include a complete blood count with differential, a chest x-ray, PPD skin test, and serological studies to investigate Epstein-Barr virus, cytomegalovirus, HIV, toxoplasmosis, or Bartonella.[5,6] Radiographic studies are usually unnecessary for evaluation of these lesions. However, persistent adenopathy suspicious for malignant disease warrants a chest x-ray. A plain chest x-ray might detect pulmonary or mediastinal lesions as a source for cervical or supraclavicular adenopathy. Ultrasonography has advantages, particularly in the pediatric population, because it does not involve ionizing radiation and is readily available. It can easily distinguish solid from cystic masses. It is helpful in evaluating the thyroid and parotid lesion and may be useful in diagnosing confusing congenital lesions. Ultrasound is also helpful in evaluating and char-

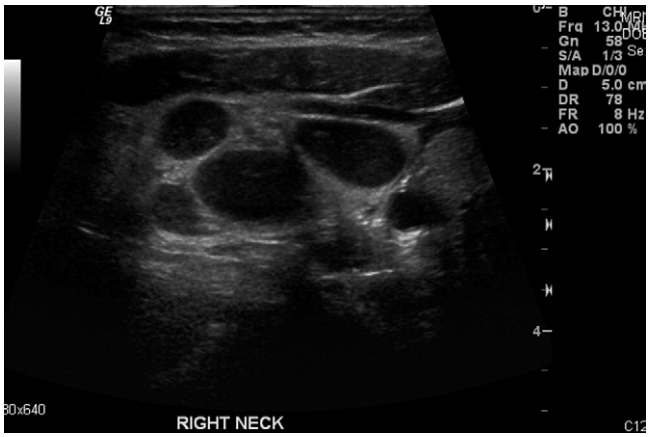

Figure 1 Ultrasonography of a large, bulky mass at the base of the neck. Differential diagnosis included lymphoma, cystic lymphatic malformation, or combined vascular malformation. Ultrasound demonstrated cystic structures surrounded by stroma.

acterizing the long-standing solitary lymph node (Figures 1 and 2).

Head and neck masses: Common etiologies

Inflammatory

Clinically palpable cervical lymphadenopathy occurs with a reported prevalence of 28% to 55% in otherwise normal infants and children.[6] Acute bilateral cervical lymphadenopathy is most commonly caused by viral respiratory tract infections or streptococcal pharyngitis, whereas unilateral cervical lymphadenitis is usually caused by streptococcal or staphylococcal infection in 40% to 80% of cases.[7] Acute suppurative lymphadenitis is typically caused by bacterial infections from penicillin-resistant Staphylococcal, group A Streptococcal infections, or both. Infants commonly have Staphylococcal lymphadenitis; anaerobic bacteria, group B streptococcal, and haemophilus influenza type B are less frequent. Local signs of inflammation, suppuration, erythema, fever, and malaise should preclude a search for primary infection around the oropharynx, head, and neck. Initial empiric treatment includes 5 to 10 days of an oral beta lacatmase-resistant antibiotic directed at the most likely organism. Failure to note improvement indicates the need for further diagnostic testing, including the use of serology, ultrasonography, fine needle aspiration with or without sedation, with a Gram stain evaluation of the collected material, and aerobic and anaerobic cultures. Bacterial infections, usually Staphylococcal aureus or Streptococcal pyogenes cause 40% to 80% of acute unilateral cervical lymphadenopathy in the 1- to 4-year-old age group.[5,8] Group B strep may cause unilateral facial or submandibular swelling, erythema, tenderness, and fever associated with poor feeding and irritability in the infant. Anaerobic bacteria can occur in the older child with dental caries or periodontal disease. Community-acquired methicillin-resistant staphylococcus aureus (MRSA) has become more prevalent as the etiology for pediatric suppurative adenitis.[7] If MRSA is identified, then clindamycin or Bactrim should be started.[5,6,9] Bactrim is effective for simple skin and soft tissue infections but is generally ineffective against group A and B Streptococcal infections. Clindamycin is highly effective treatment for pediatric MRSA, covers group A and B Streptococci, and clindamycin-resistant strains of MRSA remain uncommon. Since the spectrum of antibiotic resistance and the most common types of MRSA infections vary from one location to another, it is imperative to have a working knowledge of the local prevalence of clindamycin resistance to combat MRSA effectively.

Many cases of bilateral cervical adenitis are caused by upper respiratory tract infections of viral etiology including rhinovirus, parainfluenza virus, respiratory syncytial virus, cytomegalovirus, and Epstein-Barr virus. Less frequent etiologies include mumps, measles, rubella, herpes simplex, and human herpes simplex 6 (roseola) and coxsackie viruses. Virally induced adenopathy rarely suppurates and generally resolves spontaneously.[5]

Surgeons generally do not see children at the time of initial presentation for isolated cervical adenopathy. Most physicians would administer therapy for 10 days and cover for 5 days beyond the resolution of acute signs and symptoms. In most cases, symptomatic improvement should be noted after 2 to 3 days of therapy, although complete resolution may require several weeks. Failure to improve usually brings a patient to a pediatric surgeon. The question of surgical intervention becomes more controversial with respect to persistent lymphadenopathy of greater than 2 weeks duration on antibiotic therapy, or unilateral adenopathy in the supraclavicular or posterior triangle. Fluctuance develops in 25% of patients with acute bacterial adenitis and may be managed by additional antibiotics and or multiple needle aspirations. However, adequate drainage may not be obtained, particularly in the young and uncooperative child.

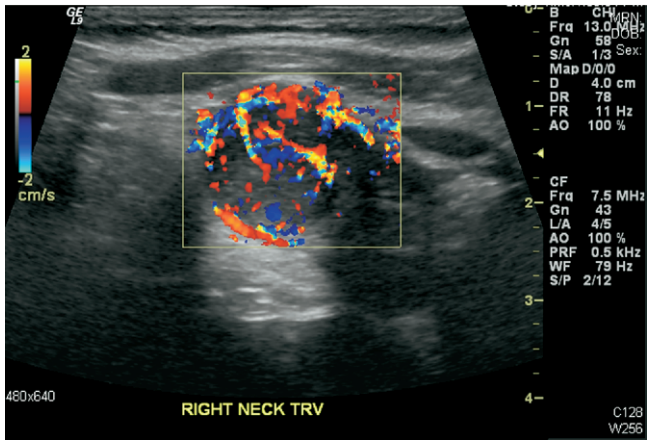

Figure 2 Duplex Doppler ultrasound evaluation of the same lesion reveals the vascular pattern characteristic of an infiltrative pattern involving a lymph node, raising the suspicion for lymphoma. (Color version of figure is available online.)

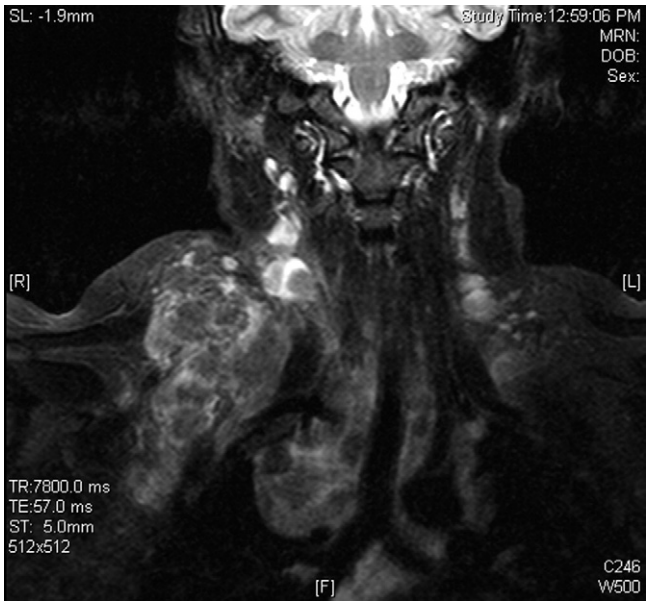

Figure 3 MRI evaluation of extensive Hodgkin's lymphoma presenting as a complex neck mass. Note the nodular appearance and surrounding fibrosis characteristic of nodular sclerosing Hodgkin's disease involving the mediastinum surrounding the airway and great vessels.

These patients are best managed with a formal operative incision and drainage under general anesthesia. This allows for proper drainage of all associated loculations and cavities not amenable to single-needle aspiration attempts. The abscess cavity is usually packed with gauze to facilitate continued drainage and to prevent early premature closure. The gauze pack is removed over a period of several days, either as an inpatient completing an intravenous antibiotic course or as an outpatient. Surgical intervention is indicated for atypical mycobacterial adenitis, suppurative inflammatory, or fistulous lymphadenopathy.

Atypical mycobacterial infections usually present in a subacute pattern, with relatively nontender, indurated, and suppurative nodes. PPD positivity is variable, and pulmonary involvement is absent. Although complete resection is the definitive therapy, macrolide antibiotics (such as clarithromycin) may have some utility.[10] In one recent series, 30/45 (67%) of children treated with cervicofacial atypical mycobacterial infections resolved with antibiotic therapy.[11] A trial of medical therapy may be worthwhile in these children.

Fungal disease

Histoplasma capsulatum, Blastomyces dermatitidis, and Coccidioides immitis are soil saprophytes endemic to certain geographic regions of the United States that cause fungal infections in humans. Most patients present with pulmonary or mediastinal involvement, with cervical lymphadenopathy secondary to the primary infection. Fungal disease must therefore be considered in the diagnosis in the child or adolescent with a mediastinal mass and cervical adenopathy, particularly in an endemic area or in an immunocompromised patient. Serological or skin testing is usually diagnostic, and most infections resolve spontaneously.[5]

Neoplastic

By far the most common head and neck malignancy in children is lymphoma. However, cervical rhabdomyosarcoma, neuroblastoma, and teratoma account for many pediatric neck lesions. Half of malignant neoplasms in the head and neck region are made up of lymphomas; 60% are non-Hodgkin's lymphoma, and Hodgkin's lymphoma make up the remaining 40%.[1] Commonly, the origin of lymphoma is from the lymph node. However, lymphomas of the head and neck may also arise from extra nodal sites and often are associated with extensive lesions within the mediastinum.[12] Neck lesions associated with mediastinal findings might represent nonmalignant disease, such as fungal or systemic inflammatory conditions, and must be distinguished from neoplastic lesions (Figures 3 and 4).

Hodgkin's disease

Hodgkin's disease has a bimodal age distribution,[12,13] with adolescents accounting for 15% of cases. Hodgkin's disease is rare in children under 10 years of age and is responsible for approximately 5% of all pediatric malignancies. The etiology is likely multifactorial, with a known association to Epstein-Barr viral (EBV) exposure. The precise role of EBV in the pathogenesis and biology in Hodgkin's lymphoma is not entirely clear. However, clinical studies indicate that

Figure 4 MRI evaluation of this neck mass revealed the extensive mediastinal involvement. The differential diagnosis included complex macro and micro cystic lymphatic malformation. Hodgkin's lymphoma was diagnosed by operative biopsy.

EBV infection precedes expansion of the tumor cell population.[12] Hodgkin's lymphoma is characterized by a small number of clonal tumor cells surrounded by a pleomorphic inflammatory cell population that constitutes the bulk of the tumor tissue. Only a small percentage of the cells (usually less than 10%) are represented by the malignant monoclonal expansion.[13] It is therefore imperative that Hodgkin's lymphoma be differentiated from the subtypes of non-Hodgkin's lymphoma that present with similar morphologic characteristics, and from other benign reactive lymphoid hyperplasias. The WHO classifications recognize two major classes of Hodgkin's lymphoma. Classic Hodgkin's lymphoma has four subtypes: nodular sclerosing, mixed cellularity, lymphocyte rich, and lymphocyte depleted. Nodular lymphocytic predominate Hodgkin's lymphoma (NLPHL) contains only rare Reed-Sternberg cells (in contrast to the classic Hodgkin's lymphoma). "Popcorn cells" are seen histologically. NLPHL is seen in 10% to 15% of all Hodgkin's lymphoma patients[1,12] and is more common among males under 10 years of age, presenting often as localized disease in an otherwise asymptomatic patient.

Clinical management

Patients usually present with painless supraclavicular or cervical lymphadenopathy. The affected lymph nodes are firmer than inflammatory lymph nodes and are usually characterized as firm, rubbery, and nontender.[1,12] Importantly, more than two-thirds of patients with cervical Hodgkin's lymphoma will have mediastinal involvement at the time of presentation.[12] When a mediastinal mass is confirmed by plain radiographs, it is advisable to evaluate the mediastinum more completely with a CT scan. In this setting, it is best to start with the assumption that airway compression exists in planning the approach to cervical Hodgkin's lymphoma.[12,14] All patients with Hodgkin's disease require a biopsy of the involved lymph node to establish the diagnosis and confirm a histological subtype.[12,13] Needle aspirations and frozen sections are inadequate; permanent hematoxylin/eosin sections must always be obtained and tissue procured for more detailed studies including immunohistochemistry and cytogenetics. Excisional lymph node biopsy or incisional biopsy of an enlarged or matted group of lymph nodes is essential to make an accurate diagnosis.[12] The contemporary therapy uses a risk-adapted approach that considers disease-related factors such as the presence of constitutional symptoms, stage, number of involved nodal regions, and the presence of tumor bulk.[1,12,13]

Non-Hodgkin's lymphoma

Non-Hodgkin's lymphomas in children and adolescents are a diverse group of neoplasms with 10% of these tumors arising in the head and neck region.[1,15] The etiology of non-Hodgkin's lymphoma (NHL) is unknown. There is a marked male predominance in all age groups, particularly in children younger than 15 years of age.[1] Compared with Hodgkin's disease (which is mostly of nodal origin), NHL often arises as a mass in extra nodal tissues. NHL's are classically divided into Burkett's and non-Burkett's lymphomas, lymphoblastic lymphomas, diffuse large B cell lymphomas, and anaplastic large cell lymphomas. The pathogenesis of NHL appears to relate to a malignant transformation of a single B or T cell of origin along its path of terminal differentiation.[12,15]

Clinical management

A contrast-enhanced CT scan or MRI of the head and neck is helpful to define the tumor in relation to other anatomic structures. Complete metastatic scanning is an important part of the staging process because NHLs are often diffuse at diagnosis and the treatment involves multiagent chemotherapy based on stage.[1,12,15] It is important to remember that childhood lymphoma is a systemic disease; the operative procedure should not delay institution of chemotherapy. Initial surgical management should include an incisional biopsy, followed by intense multi-agent chemotherapy once the diagnosis is confirmed, except in the cases of small and easily resectable isolated lesions.[12] Cervical primary tumors should undergo initial diagnostic biopsies with procurement of enough tissue to determine the histological subtype.

Mediastinal masses associated with neck masses

Because non-Hodgkin's and Hodgkin's lymphoma are often associated with lesions in the mediastinum, it is imperative for the pediatric surgeon to determine from the history and physical examination whether the possibility of airway compromise exists, particularly when contemplating a biopsy and the potential need for general anesthesia.[12,16] Respiratory collapse under general anesthesia is a recognized complication associated with anterior mediastinal masses. Identification of patients at risk has been a major challenge because the presence of specific respiratory symptoms has not been shown to correlate with the severity of airway compression.[14] Preoperative measures for identifying patients at risk for respiratory collapse on induction of general anesthesia have centered around measurement of the tracheal cross-sectional area by CT scans, in combination with pulmonary function tests.[16] Shamberger and coworkers' prospective study of the risk assessment in 31 children with mediastinal masses, using the combination of cross-sectional area of the trachea and peak expiratory flow rate, demonstrated that all patients with values greater than 50% for both predicted peak expiratory flow rate and tracheal area underwent administration of general anesthesia without respiratory complications. Conversely, all children with peak expiratory flow rate and tracheal areas that were less than 50% of predicted received local anesthetic for their biopsy without sequelae.[17]

All patients with cervical adenopathy and a significant mediastinal mass discovered by plain films should therefore

be evaluated by a CT scan of the chest to determine the cross-sectional area of the trachea, the burden of mediastinal disease, and also the possibility of pleural effusion. The peak expiratory flow rate has been determined to be the best predictor for evaluating the magnitude of an extra thoracic obstruction.[14,16,17]

Cervical neuroblastoma

Cervical neuroblastoma may occur as a primary tumor but is more commonly a site of metastatic disease from an abdominal or thoracic primary tumor. Primary cervical disease may occur as a mass in the lateral neck or retropharyngeal space. Metastatic disease may cause proptosis, periorbital swelling, ecchymoses, acute cerebellar ataxia characterized by opsoclonus–myoclonus, and chaotic nystagmus.[1,18] Primary cervical neuroblastoma has a favorable outcome. Localized lesions or low-stage disease has an excellent prognosis with complete surgical resection. Radiation therapy and/or chemotherapy and secondary surgeries are strategies to eradicate residual disease.

Cervical teratomas

Teratomas are tumors with elements derived from all three germ-cell lineages in various degrees of differentiation. Teratomas arise in a variety of locations throughout the body. Most are sacrococcygeal, with cervical teratomas representing less than 10%.[3,4] Most cervical teratomas in infants and children are benign lesions. The diagnosis is usually obvious at birth, but due to the widespread use of prenatal ultrasound, there has been an increase in the diagnosis of fetal neck masses, particularly airway-threatening lesions. The vast majority of cases of fetal airway obstruction are due to cervical teratomas or lymphatic malformations.[19] Fetal MRI provides better detail about the size and position of the mass and its anatomic relationship to the airway. A compromised airway secondary to cervical teratoma is an indication for the EXIT (ex utero intrapartum treatment) procedure (Figure 5).[19]

Figure 5 Antenatal diagnosis of a threatened airway by a cervical teratoma mandated the EXIT procedure. Neonatal tracheostomy was performed on placental support after bronchoscopy failed to secure the airway. (Color version of figure is available online.)

Congenital cystic lesions

Thyroglossal duct cyst

Thyroglossal duct cysts are the most common congenital midline cervical anomalies in children.[3,20-23] The thyroid gland originates in early gestation from a diverticulum between the anterior and posterior muscle complex of the tongue. This region represents the proximal remnant of the foramen cecum. As the embryo elongates and the thyroid gland descends, it does so in the vicinity of the eventual location of the hyoid bone. As this occurs, the median thyroid anlage elongates, with the descending gland forming the thyroglossal duct.[23] The thyroglossal duct generally obliterates by the 5th week of gestation, leaving behind a proximal remnant as the foramen cecum. A thyroglossal duct sinus persists when the thyroglossal duct fails to obliterate before the formation of the hyoid bone.

Based on this embryological descent, a thyroglossal duct cyst and sinus tract remnant can occur anywhere from the base of the tongue to the lower midline neck. The most frequent presentation is a midline painless cystic mass in the region of the hyoid bone. Most thyroglossal duct cysts present during the first 5 years of life. Clinically, the uncomplicated cyst typically moves cranially with swallowing and protrusion of the tongue because of its close relationship with the hyoid bone and the foramen cecum.[20,22] Dermoid cysts, the most likely item in the differential diagnosis, typically do not move with this maneuver. Infection is a complication seen in thyroglossal cysts because of the close anatomic association with the oral cavity. Many patients will present with a concurrent or recent history of an upper respiratory tract infection. The infected thyroglossal duct cyst usually resembles other abscesses, with intense erythema and fluctuance (Figure 6). The most common pathogens are Haemophilus influenza, Staphylococcus aureus, and Staphylococcus epidermidis.[23] It is unusual for a thy-

Figure 6 A 5-year-old girl with infected thyroglossal duct cyst initially treated with a first generation cephalosporin for 10 days without improvement. Surgical evaluation diagnosed a fluctuant abscess which subsequently spontaneously drained. (Color version of figure is available online.)

roglossal duct cyst to present as a communicating sinus tract to the skin, since the thyroglossal duct does not communicate with the ectoderm during the development. However, up to 25% of these lesions may present as a draining sinus tract in the midline, thought to represent a spontaneous rupture.[20,22,23]

Thyroglossal duct remnants can contain functional thyroid tissue and may have a solid component since they are lined by ductal epithelium.[20] A patient who presents with symptoms of hypothyroidism should be worked up for the possibility of median ectopic thyroid. In approximately 1% of patients with a thyroglossal duct cyst, the only functional thyroid tissue is located within the cystic mass.[20] These patients are frequently hypothyroid with elevated thyroid stimulating hormone (TSH) levels. The incidence of thyroid carcinoma in a thyroglossal duct cyst remnant is reported to be less than 1%. However, since the majority of thyroglossal duct cysts are removed in childhood, the absolute risk is unknown. With the exception of medullary carcinoma of thyroid, all types of thyroid malignancy have been reported, with the majority being papillary. Approximately 90% of cases of presumed thyroglossal duct carcinoma have presented in adulthood.[20,23]

The diagnosis of thyroglossal duct cyst is usually straightforward and almost never requires an extensive evaluation. The literature does, however, contain controversial claims regarding the need for preoperative thyroid scanning to identify those patients that have a median ectopic thyroid. Others argue that the cost of routine scanning is excessive given the overall incidence of 1% to 2%, and exposes many patients to unnecessary radiation.[23] A safe approach is to perform a thorough history looking for signs and symptoms consistent with hypothyroidism. If hypothyroidism is suggested, then TSH screening and preoperative ultrasound of the midline neck should provide the necessary information to select patients for preoperative thyroid scanning.[23]

Surgical management

In the uncomplicated thyroglossal duct cyst, an elective procedure described by Sistrunk is the operation of choice.[20,22,23] The patient is positioned supine, with the head of the table slightly elevated and the neck extended, and a transverse incision is used. Careful dissection is performed to identify the distal tract. Dissection around the cyst proceeds cranially toward the hyoid bone, which is facilitated by elevating the cyst out of the wound. Once the hyoid bone is reached, its central portion associated with the tract is resected. En bloc resection of the proximal tract is important to ensure complete removal of the lesion. The anesthesiologist or another surgeon may, if necessary, insert a finger into the mouth and press against the base of the tongue to ensure proximal dissection is complete. The tract is suture ligated at the most proximal end and the block is removed (Figure 7).

Occasionally, patients will present with an infected thyroglossal duct cyst unresponsive to initial antibiotic therapy. Intense erythema and fluctuance suggest an abscess, and formal incision and drainage may be necessary to control the infection. There is some concern that drainage procedures can "seed" the surrounding tissues, predisposing to recurrence. Occasionally, the thyroglossal duct cyst or abscess will spontaneously erupt. A Sistrunk procedure is performed once the infection has cleared (after incision and drainage or spontaneous drainage). A clinically palpable mass usually remains. It is important to excise an ellipse of

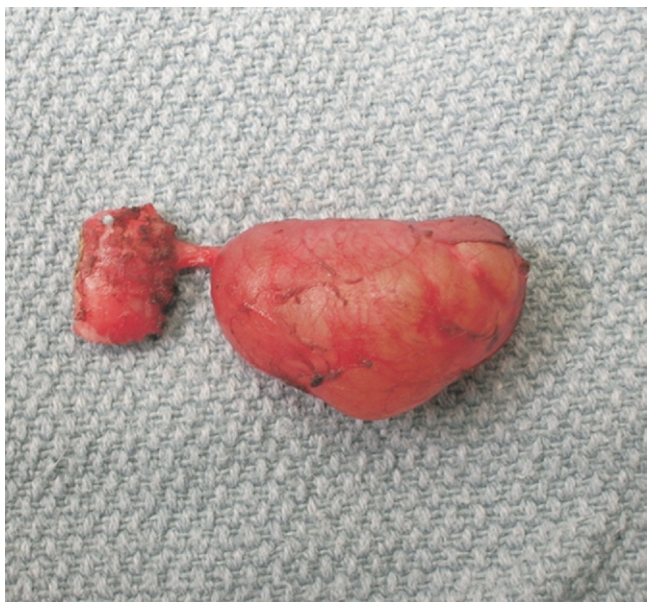

Figure 7 Thyroglossal duct specimen demonstrating the intact cyst, the fistulous tract, and the attached mid portion of the hyoid bone. Excision performed in the manner described by Sistrunk. (Color version of figure is available online.)

skin including the area of the drainage to prevent recurrence. The infection usually makes the otherwise elegant Sistrunk procedure more challenging secondary to the inflamed surrounding tissues. Although the presence of a preoperative or concurrent infection has historically been associated with increased recurrence rates, a recent review of 100 patients from a major pediatric hospital found no association between preoperative infection and increased recurrence rates.[23,24]

Branchial cleft cyst and sinus

Branchial cleft anomalies are the second most common congenital head and neck lesion found in children, after thyroglossal duct cysts.[3,20,21] These anomalies are composed of a heterogeneous group of congenital malformations that arise from incomplete obliteration of the pharyngeal clefts and pouches during embryogenesis. Most branchial cleft anomalies involve the first and second cleft and pouch complexes.[21,22] Normally, the first branchial arch forms the mandible and a portion of the maxillary process of the upper jaw. This arch is also involved in development of portions of the inner ear, whereas the cleft and pouch become part of the external auditory canal and mastoid ear cells. The second arch contributes to the hyoid bone and the adjacent area of the neck. This pouch becomes the palatine tonsil and supratonsillar fossa. Understanding this developmental process clarifies the clinical presentation of branchial anomalies. First branchial cleft and pouch anomalies enter the external auditory canal and occasionally the middle ear, while second arch anomalies enter the supratonsillar fossa. Third and fourth sinuses and fistula may appear similarly to the second cleft sinus externally. However, as a rule it is the internal opening of the sinus that is crucial in defining the cleft or pouch origin, and the exceedingly rare third and fourth derivatives enter the pharynx through the pyriform sinus.[21,22]

Presentation and diagnosis

First cleft remnants typically present as a cyst sinus or fistula somewhere between the external auditory canal and the submandibular area. These lesions have an intimate relationship with the parotid gland and the branches of the facial nerve. Second branchial cleft anomalies account for the vast majority of all branchial cleft disorders and usually present as a fistula or cyst found in the lower anterior lateral region of the neck.[3,20,21] Fistulas are usually diagnosed in infancy in childhood and typically have intermittent and chronic drainage from the opening along the anterior border of the sternocleidomastoid muscle. Cysts are more often diagnosed in adults as a nontender mass in the neck. The second branchial arch anomalies can be intimately associated with the glossopharyngeal and hypoglossal nerves, and enter the pharynx at the level of tonsillar fossa.[21] Third and fourth arch anomalies are uncommon. Branchial cleft cysts are lined by squamous epithelium but can contain respiratory epithelium as well.

Surgical management

The definitive treatment for all branchial cleft remnants is complete surgical excision. If incompletely resected, there is a high incidence of recurrence. Second branchial arch remnants are usually approached through a cosmetic transverse surgical incision. This cyst is meticulously dissected from the superficial tissue at fascia and dissected proximally, keeping the fistulous tract intact. It is possible to cannulate the tract with a small probe to aide in its dissection. Some surgeons prefer to inject the tract with a small amount of methylene blue. It is occasionally necessary to use a stepladder incision to gain better visualization of the upper portion of the tract as the dissection moves superiorly. Once the proximal opening near the pharynx has been identified, the tract is ligated and divided.

Preauricular cysts

Preauricular cysts do not represent true cysts or sinuses of the neck but need to be distinguished from first branchial cleft cysts. Unlike branchial cleft cysts, preauricular cysts are common, often bilateral, tend to be inherited, and are rarely complicated by infection. Furthermore, they are neither involved with the facial nerve nor do they enter the external auditory canal. They are thought to arise from abnormal formation of the external ear from the developing hillocks and are successfully excised by removing all the ductal epithelium through an inverted "L-shaped" incision.[22]

Vascular anomalies

Introduction

Vascular anomalies are best classified as vascular tumors or vascular malformations. The biological classification based on cellular kinetics and clinical behavior distinguishes vascular tumors as lesions that arise by endothelial hyperplasia. In contrast, vascular malformations are congenital lesions derived from capillaries, veins, lymphatic vessels, arteries, or a combination of these. They are lesions that arise by dysmorphogenesis but exhibit normal endothelial turnover.[25]

Vascular tumors of the head and neck: Hemangioma

Hemangioma is one of the most common tumors of infancy and childhood and is found in the head and neck region approximately 60% of the time.[25,26] Most cutaneous hem-

angiomas appear approximately 2 to 4 weeks after birth. Hemangiomas of infancy follow a predetermined course of proliferation followed by involution. The proliferative phase is characterized by rapid growth in the first 6 to 8 months of infancy. There is a variable period of quiescence followed by the involution phase. Maximum involution occurs in approximately 50% of children by age 5 years, and 90% of children by age 9 years.[25,26] Based on the anatomic depth, hemangiomas are characterized as superficial, deep, or combined. Superficial lesions tend to be soft, red, raised, and occasionally telangiectatic. Deep lesions may show a spectrum of appearances and consistency ranging from soft and subtle to raised and more firm with a bluish color. Combined lesions appear as red dermal tumors with epidermal and dermal components along with subcutaneous masses.

A variation of hemangioma is the congenital hemangioma; unlike the hemangiomas of infancy, these lesions are fully developed at birth and do not undergo additional postnatal proliferative growth. These congenital hemangiomas fall into two distinct subgroups: rapidly involuting congenital hemangiomas (RICHS) and noninvoluting congenital hemangiomas (NICHS). Both are high-flow lesions that can be misdiagnosed as arterial–venous malformations. The RICHS rapidly regress over the first year of life, whereas NICHS do not.[25,26]

Sixty-five percent of patients with hemangiomas have cervicofacial involvement.[25] Lesions that cover a beard distribution including the chin, jaw line, and preauricular areas may have associated airway involvement.[26] Infants with hemangiomas in the beard distribution area should be inspected for glottic and subglottic hemangiomas, the two most common locations in the airway. Treatment should be initiated if airway involvement is confirmed. Localized lesions are managed with laser therapy, intralesional steroids, or surgical resection. These lesions can proliferate for up to 12 to 16 months and require systemic treatment if the airway is compromised; tracheostomy should be reserved for patients who fail medical therapy.[26]

Treatment

Since many hemangiomas spontaneously involute, with little or no functional disability or cosmetic defect, reassurance and observation is usually all that is required. It is important to periodically see these patients so the lesions can be monitored for signs of ulceration, growth, and complications that may indicate additional therapy. The decision to intervene with a cervical facial hemangioma is based on the size and location of the lesion, presence of complications such as ulceration or bleeding, age of the patient, and the phase of growth at the time of the evaluation. Larger lesions that interfere with the function of vital structures (such as the eye or eyelid, mouth, or nares), are likely to require some form of treatment.[25] Additional therapies include corticosteroids (either systemically or via intralesional injection), laser therapy, and surgical excision. Intralesional corticosteroid should be considered for small localized cutaneous hemangiomas located on the nasal tip, cheek, or eyelid. Triamcinolone (25 mg/ml) is injected slowly at low pressure with a 3-mL syringe and a 25-gauge needle. A dosage of 3 to 5 mg/kg per injection is administered every 6 to 8 weeks for three to five injections.[25] Oral corticosteroids are the first-line treatment for problematic, endangering, or life-threatening hemangiomas. Prednisone or prednisolone is administered at 2 to 4 mg/kg per day for 2 weeks. The use of systemic corticosteroids accelerates the involutional phase. Interferon alpha 2A or 2B should be considered as a second-line drug for endangering or life-threatening hemangiomas.[25,26] Laser treatment for cervicofacial hemangioma remains controversial. Surgical excision is usually reserved for cervicofacial hemangiomas that present a threat to vital structures associated with complications such as ulceration, hemorrhage, or infection unresponsive to pharmacological therapy. Surgical excision may be indicated for the residual scar after complete involution, or when the emotional burden to the child or family is significant and potential for cosmetic deformity is quite low. The timing of surgery remains controversial, particularly in regard to the growth phase of the lesion and the age of the patient. Although lenticular excision has traditionally been commonplace, a useful approach with circular hemangioma is the technique of circular excision followed by a purse-string closure. This technique has been widely used with excellent results.[25]

Vascular malformations

The classifications of these anomalies are based on the clinical and histological appearance of the abnormal channels as resembling either capillaries, lymphatics, veins, arteries, or combinations thereof.[25,27] Cystic lymphatic malformations (cystic hygromas, lymphangiomas) are benign vascular lesions that arise from an embryological disturbance in lymphatic development. They are most commonly found in the head and neck region. They can be detected antenatally on a prenatal ultrasound or may be noted at birth; most present before the age of 2 years. Lymphatic malformations can be characterized as microcystic, macrocytic, or combined.[25] The prenatal diagnosis of anterior or posterior cervical lymphatic malformations may have significant clinical implications with potential neonatal airway obstruction. Prenatal consultation is usually obtained, and observation over the remainder of pregnancy occurs with follow-up level II ultrasounds and fetal MRI scans to judge the size and development of the lymphatic malformation and potential for airway obstruction. The EXIT procedure is a technique that occasionally needs to be employed by a multidisciplinary team to deliver the child and obtain control of the airway in a timely fashion.[19,28]

Postnatally, most cervicofacial lymphatic malformations are easily diagnosed by physical examination. All patients should have a chest x-ray to identify cervical extension into the mediastinum, which is common. Ultrasound is helpful to

determine the macrocystic and microcystic features. However, it is less valuable in showing deep extension into the structures of the neck and mediastinum. CT and/or MRI are superior modes to demonstrate the anatomical relationships.

Treatment depends on the clinical presentation, size, and complications of the lymphatic malformation. Small lesions may be amenable to complete surgical excision with excellent results, whereas other macrocytic lesions might be better served with intralesional sclerotherapy.[25] The microcystic or combined macro-microcystic lymphatic malformations, particularly ones that traverse different tissue planes of both the neck and mediastinum, require a careful and deliberate approach, usually in a staged fashion. Cervical cystic lymphatic malformations may become infected. General enlargement or swelling of the lymphatic malformation may be associated with cellulitis, and can occur following an upper respiratory tract infection.[25,27] Asymptomatic patients may develop respiratory distress secondary to the concurrent infection. Expectant management of the patient's airway is prudent in these cases, with administration of intravenous antibiotics. Another cause of rapid enlargement is hemorrhage into cyst. Other complications include lymphatic leakage and chylothorax. Because surgical excision is a large undertaking for complex and combined micro- and macrocytic lesions of the head and neck, sclerotherapy has frequently become an alternative initial approach, particularly for patients with macrocystic disease. A number of sclerosing agents have been used (Bleomycin, OK-432, fibrin glue, and others). However, ethanol is most commonly utilized in this country because of its efficacy and availability. The lymph is aspirated under ultrasound guidance and ethanol is injected into the cyst.[25,27] The procedure is usually done under general anesthesia. Fever, erythema, tenderness, and leakage are not uncommonly noted. As mentioned previously, swelling in the malformation is a dangerous side effect following sclerotherapy, potentially causing airway compromise when the lesion involves the cervicofacial and mediastinal locations. Good results (partial or complete regression) are obtained in slightly more than 50% of patients in the literature, dependent on the nature of the lesion, patient selection, and other variables.[29,30]

Summary

Head and neck lesions are some of the most common entities encountered in the pediatric population and can be distinguished as congenital, inflammatory, or neoplastic. The majority of these lesions are benign conditions that are readily diagnosed and have a predictable natural history. The role of the pediatric surgeon is to facilitate the diagnosis and provide definitive care of these lesions.

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