Evaluation of Pediatric Neck Masses

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Introduction

Neck masses can occur quite commonly in children for a variety of causes. Pediatric neck masses can broadly be classified as inflammatory, congenital, or neoplastic in origin. Evaluation of neck masses should be organized and preliminarily focus on the most common causes. However, it is important to consider malignancy as it is the second leading cause of death in children after the first year of life and head and neck malignancy accounts for 12% of childhood cancers.[1]

When evaluating neck masses, the severity of presentation will guide the initial investigation. Complete histories regarding the neck mass, coupled with the age of the child and a complete physical examination often give enough information to begin treatment. As imaging modalities improve they may play a larger role to complement or guide treatment decisions. A complete knowledge of anatomy, embryology, and common clinical presentations is required.

History

A thorough history of the neck mass is paramount to the diagnostician. The patient's age, duration, and character of symptoms can yield beneficial information. Often, congenital or benign neoplastic lesions can be present at birth, while malignant neoplasms in neonates are rare. However, some congenital lesions such as branchial cleft and thyroglossal duct anomalies may present later in childhood when these lesions become secondarily infected. Likewise, other inflammatory lesions of the neck commonly present between the ages of 1 and 4 years.[2] Rare neonatal inflammatory neck masses include maternally acquired infections such as syphilis or human immunodeficiency virus (HIV).

While age at presentation can be quite helpful, often the time-course of symptoms can provide additional important information. Rapidly progressive lesions commonly indicate an inflammatory or neoplastic cause. Adenopathy of short duration associated with recent upper respiratory infection may warrant conservative treatment with antibiotics, while masses associated with more concerning signs of deep inflammatory involvement (torticollis, dysphagia) may require imaging to rule out abscess formation. Slowly progressive lesions over the course of several months suggest a congenital lesion or benign neoplasm.

Involvement of other organ systems can be revealed in a complete review of systems. Associated fevers, weight loss, or night sweats are concerning for neoplastic causes. Pain in other parts of the body such as joint, muscles, or organs support the diagnosis of a systemic inflammatory condition.

History regarding the child's environment can be obtained from the family. If an infectious etiology is suspected, cat and other animal exposure can be elucidated. Insect (particularly tick) exposure can also be queried. Although rare in children, ionizing radiation exposure can also contribute to neoplastic growth.[3] Recent travel or exposure to people with tuberculosis can also aid in diagnosis.

Family history is helpful as congenital neck masses may be associated with syndromes such at branchial-oto-renal syndrome. Some malignancies occurring in the head and neck in children may be associated with syndromes such as Multiple Endocrine Neoplasia (MEN) and neuroblastoma.

Physical Examination

A complete discussion of pediatric head and neck examination technique is beyond the scope of this chapter, but certain features can help distinguish the etiology of a neck mass. A thorough examination of the mass itself as well as the nodal regions of the neck is required.

The mass should be bimanually palpated to elucidate the consistency and character of the lesion. Tenderness of the mass points to an inflammatory cause, though rapidly progressive tumors or hemorrhage into neoplasms may present with pain and rapid growth of the lesion. Fluctuance can indicate abscess in acutely inflamed lesions or cystic congenital lesions. Firmness or fixation to the skin or other structures is concerning for neoplasm. Vascular lesions are often readily distinguished by their characteristic color and growth patterns.

Examination of the chest, abdomen, groin, and extremities cannot be overlooked. Skin changes such as café-au lait spots or target lesions can suggest congenital or inflammatory etiologies, respectively. Splenic or liver enlargement may also be detected in inflammatory conditions such as Ebstein-Barr virus (EBV) infection.

Associated signs on physical exam should be noted. Dysphagia, torticollis, trismus or voice change can indicate deep neck space inflammatory changes. Bilateral lymphadenopathy also often indicates inflammatory etiologies. Epistaxis or history thereof can indicate nasal or nasopharyngeal involvement often of neoplastic nature.

Drainage from the neck may be associated with a fistulous congenital lesion or an inflammatory condition resulting in suppurative lymphadenitis. Certain maneuvers in a cooperative patient may also aid in diagnosis such as vertical movement of the mass with tongue protrusion in thyroglossal duct cysts.

Mass location may yield some of the most important diagnostic information. Midline neck masses can be thyroglossal duct anomalies, lymph nodes, lipomas, dermoid cysts, and thyroid tissue or thyroid neoplasms. Multilevel lymphadenopathy may point to inflammatory causes while longstanding enlarging masses occupying the posterior triangle or supraclavicular region should raise suspicion for malignancy. Branchial cleft cysts are commonly seen as painless masses anterior to the ear (Type I) or below the angle of the mandible along the anterior border of the sternocleidomastoid muscle (Type II or III). Similarly, branchial cleft anomalies may drain through a fistula in this location.

Imaging

Imaging modalities such as computed tomography (CT), magnetic resonance imaging (MRI), and ultrasound are not required in the diagnosis of many neck masses, but are quite valuable in select situations. Concern for exposure to ionizing radiation and the need for sedation in uncooperative patients are some challenges to obtaining imaging in the pediatric population.

Plain x-ray films lack specificity for routine use in neck masses, but may be helpful in selecting treatment in some acute cases. For example, plain films may indicate airway impingement associated with a neck mass that requires urgent otolaryngology intervention. Lateral neck films may also identify prevertebral edema necessitating further imaging or subcutaneous or deep neck air suggestive of necrotizing infection.

Ultrasonography is increasingly employed for diagnosis of neck masses. The relative ease of obtaining imaging, low cost, and lack of radiation are desirable factors in pediatric diagnosis. Ultrasonography is particularly useful for evaluation of fluid-tissue interfaces, as in cystic lesions and superficial abscess formation. It is the test of choice in thyroid imaging and should be conducted in all patients diagnosed with a thyroglossal duct cyst prior to surgical intervention to confirm native thyroid tissue not associated with the cyst. Ultrasound is helpful in delineating the relationship between neck masses and surrounding neck structures including vessels. Addition of doppler-flow imaging provides information regarding mass vascularity and relation to surrounding neck vasculature.

CT and MRI both provide exceptional detail of neck anatomy. However, the risks and benefits of each should be considered. CT involves ionizing radiation and requires a cooperative patient though conventional scans allow for more rapid acquisition of multiplanar images. Contrast enhanced CT is the standard of care for the diagnosis of deep neck space abscesses and provides superior soft tissue resolution to ultrasound.[4] CT is also the imaging modality of choice when osseous involvement is suspected.

MRI provides the best characterization of soft tissue, but requires a cooperative patient or sedation to obtain adequate images. Certain soft tissue lesions such as hemangiomas can be diagnosed based upon MRI appearance alone, obviating biopsy. Other lesions may require both CT and MRI to reconcile soft tissue and bony tissue relations.

Laboratory evaluation

Laboratory analyses may be helpful in pediatric neck mass work-up. A complete blood count (CBC) may indicate infectious etiology or presence of a hematologic malignancy and is inexpensive and easily obtained in most patients. Other more specific tests should be dictated by clinical suspicion. Patients with a history of tuberculosis exposure may undergo tuberculin skin test. Those with concern for inflammatory neck lesions not responsive to conservative measures can be evaluated serologically for Ebstein-Barr Virus, cytomegalovirus, toxoplasmosis, bartonella, or HIV.

Fine needle aspiration may be helpful in instances of suspected neoplasm or atypical infection.[5, 6]

Common Lesions

Inflammatory

Palpable lymphadenopathy occurs in up to 50% of otherwise normal children.[7] Bilateral acute cervical adenopathy is most commonly caused by viral upper respiratory pathogens or streptococcal pharyngitis while unilateral cervical lymphadenitis is the result of streptococcal or staphylococcal pharyngitis in up to 80% of cases.[8] Virally induced lymphadenopathy often occurs in relation to common upper respiratory viruses such as rhinovirus, parainfluenza virus, respiratory syncytial virus, CMV or EBV. Rarely mumps, measles, rubella, and coxsackie viruses can be causative. Virally induced lymphadenopathy rarely suppurates and generally resolves spontaneously.[8] Failure to improve spontaneously or with oral antibiotics should prompt additional diagnostic methods such as ultrasound, serology or aspiration for gram stain, culture, and cytology where appropriate.[5]

Atypical mycobacterial infections usually present with nontender, indurated and suppurative nodes. Tuberculin skin test is variable positive and some cases may respond to antibiotic therapy although surgical resection is the definitive therapy.[9] However, lymphadenopathy associated with *Mycobacterium tuberculosis* is treated with anti-tuberculin medications and surgery is reserved for advanced cases.[10]

Fungal diseases endemic to some areas such as *Histoplasmosis capsulatum*, *Blastomyces dermatitidis,* and *Coccidioides immitis* may present with cervical lymphadenopathy. Primary fungal infection often involves the mediastinum or lungs and diagnosis can be obtained via serology. It is important to consider fungal causes in unusual cases of lymphadenopathy, immunocompromised patients, or in endemic areas.

Mass location may help determine the diagnosis in cases of acutely infected thyroglossal duct or branchial cleft cysts. These may fluctuate in size and cause a sensation of fullness or rarely airway compromise. Posterior triangle apparent inflammatory lesions should prompt investigation for scalp or upper airway inflammatory conditions with a low threshold for radiologic and cytological analysis to rule out malignancy.

Children with neck masses and signs and symptoms suspicious for deep neck space abscesses such as dysphagia, drooling, stridor, torticollis or trismus should undergo airway stabilization if necessary followed by imaging to characterize the lesion location for treatment.

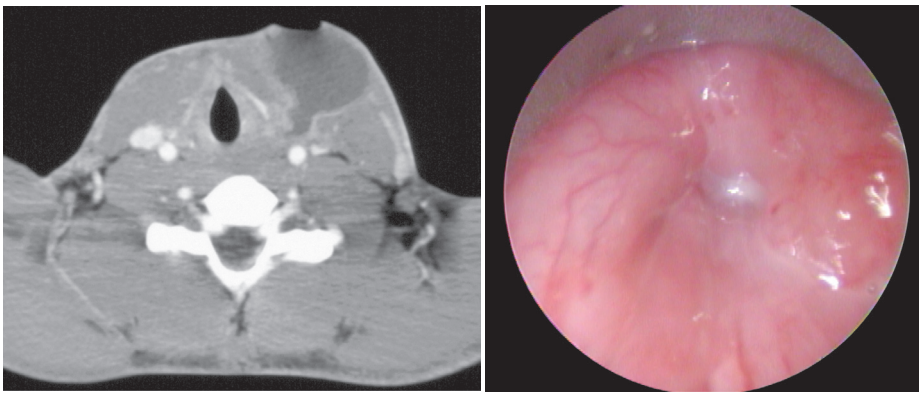
Congenital

Common congenital lesions include thyroglossal duct cysts, brachial cleft anomalies, and vascular lesions. As discussed, thyroglossal duct cysts commonly present when they become acutely infected as a tender mass in the midline neck mass near the level of the hyoid that moves in a vertical plane with tongue protrusion. During development, the thyroid descends from the base of the tongue to its adult paratracheal location and a thyroglossal duct anomaly represents aberrancy in this development. Dermoid cysts should be included in the differential diagnosis of midline neck masses and tend to be attached to the skin.

Local resection of dermoids is sufficient, while a Sistrunk procedure is required in management of thyroglossal duct lesions.

Branchial cleft anomalies are second to thyroglossal duct cysts as the most common congenital neck lesions in children. Branchial cleft anomalies arise from incomplete obliteration of the pharyngeal pouches and clefts during embryogenesis and may manifest in several ways. Most involve the first and second cleft and pouch derivatives. Commonly, they present as a painless mass located below the angle of the mandible along the anterior border of the sternocleidomastoid (SCM). They can drain through sinus or fistulous tracts and complete excision including the entire is necessary **(Figure 1)**.

Figure 1: CT scan demonstrates neck abscess. The endoscopic view of purulence coming from the apex of the piriform sinus confirms a type III branchial cleft sinus with cyst.



Vascular anomalies likewise can be found in several forms. Vascular malformations are congenital lesions arising from arteries, veins, lymphatic vessels, capillaries, or a combination thereof. Vascular tumors form by endothelial hyperplasia.

Hemangiomas represent the majority of vascular tumors. Hemangiomas present as red or bluish soft multilocular mass, which usually involves the skin and appears 2-4 weeks following birth. Hemangiomas exhibit an early rapid growth phase followed by slow regression with up to 50% of patients exhibiting complete resolution by 5 years of age. Thus, many hemangiomas are observed, yet surgical and medical therapies have a role in treatment. Necessary for the otolaryngologist to remember is the potential for airway hemangiomas associated with those of the head and neck. Up to 30% of patients with hemangiomas in a V3 or "beard distribution" may have a concurrent airway hemangioma.[11] Untreated, the mortality rate due to obstruction of airway hemangiomas is between 40 and 70%.[12]

Common vascular malformations include arteriovenous malformations, venous malformations and lymphatic malformations. Lymphatic malformations

may be microcystic or macrocystic and involve several layers of the neck. They are usually diagnosed prenatally or soon thereafter based upon physical appearance of a soft, diffuse, and painless neck mass. Treatment is usually surgical if possible. Arteriovenous malformations are relatively uncommon in childhood and may present anytime from birth to adulthood. Doppler ultrasound imaging may confirm the presence of arteriovenous shunting.[13] Venous malformations are commonly seen in the skin and subcutaneous tissue and enlarge in proportion to the individual. Compression of the jugular vein or a Valsalva maneuver may make the lesion more evident. Both medical and surgical therapies can be employed in treatment.[13]

Neoplastic

Malignant

Lymphoma is the most common head and neck malignancy in children comprising almost 50% of childhood malignancies of the head and neck and 12% of all childhood malignancies until age 14. [14] 60% are classified as Non-Hodgkin's lymphoma while the other 40% are Hodgkin's lymphoma. Despite this percentage, pediatric otolaryngologist may encounter Hodgkin's lymphoma more frequently as it commonly presents in the neck. Patients usually present with painless supraclavicular or cervical lymphadenopathy that is usually firmer than inflammatory lymph nodes and characterized as rubbery and nontender. Persistent lymphadenopathy greater than 4 weeks, or nodes greater than 2 cm in size should raise suspicion for neoplasm.[15] 30% of patients may present with nonspecific symptoms of fever, night sweats, weight loss and/or anorexia.[16] Definitive diagnosis necessitates biopsy of the affected nodes with adequate tissue for immunohistochemistry and cytogenetic testing. Therefore needle aspiration and frozen sections are inadequate. Important to remember in planning operative biopsy is that up to 65% of patients with Hodgkin's and 38% with non-Hodgkin's lymphoma can have mediastinal involvement which may cause airway compression during induction of general anesthesia.[17]

Non-Hodgkin's lymphoma refers to a group of malignancies that affect the head and neck region 10% of the time. It usually presents as a widespread disease and also contrary to Hodgkin's lymphoma, non-Hodgkin's frequently presents in extranodal sites.[17] It has a marked male predominance in all age groups. Similar diagnostic requirements exist as for Hodgkin's lymphoma. Surgical intervention for both Hodgkin's and non-Hodgkin's lymphomas is generally limited to diagnostic biopsy.

Soft tissue sarcomas of the head and neck in pediatric patients are commonly rhabdomyosarcomas and undifferentiated sarcomas. Rhabdomyosarcoma is the most common soft tissue malignancy in children and the head and neck is the most frequent site of origin with tumors commonly diagnosed in the orbit, nasopharynx, middle-ear/mastoid, and sinonasal cavities.[18] Presentations are often nonspecific based upon the variety of locations and tumors may spread by direct extension to adjacent structures or metastatic spread. Diagnosis is by biopsy and treatment is multimodality and can involve surgical intervention if complete resection is possible.

Other sarcomas occurring in the head and neck include fibrosarcoma, neurofibrosarcoma, synovial sarcoma, hemangiopericytoma, and chondrosarcoma.[19] These often present as painless masses and diagnosis is based upon surgical biopsy. Therapy is multimodality as in rhabdomyosarcoma.

Thyroid carcinoma and salivary gland malignancies are also seen in pediatric neck mass evaluation but represent less than 5% of cases. Thyroid carcinomas can be found in up to 20% of solitary thyroid nodules in children. Salivary carcinoma can be found in 23% of firm salivary gland masses in children.[20] Although data is limited, FNA can aid diagnosis in both thyroid and salivary gland disease. Treatment of both is surgical resection, though extent of resection is debatable.[21]

Benign

Several benign neoplasms of the head and neck deserve mention. Pilomatrixomas are solitary, firm, intradermal or subcutaneous nodules often mistaken for lymphadenopathy.[22] Histologically, they represent hamartomas of follicular origin and their firm consistency is due to a high incidence of calcium deposition. Surgical excision including the overlying skin is curative.[23]

Benign tumors of neural origin such as schwannomas and neurofibromas may occur in the head and neck in children. Commonly, imaging such as MRI can confirm diagnosis.[24] Schwannomas generally are found along the course of cranial or sympathetic nerves and surgical resection is indicated. Neurofibromas commonly present as part of one of two genetic subtypes of neurofibromatosis. Surgical excision is pursued if there is disfigurement or risk of functional compromise, though complete excision is difficult.[24]

Unique to neonates is the occurrence of a benign cervical mass referred to as sternocleidomastoid tumor of infancy (STI). This condition presents as a firm mass within the SCM at birth or soon thereafter. Birth trauma is thought to contribute to appearance of the lesion, though exact etiology is still controversial. Torticollis may be present and ultrasound or CT is usually adequate for diagnosis. Recovery can be spontaneous or with conservative measures such as physical therapy.[25] Surgical intervention in the form of distal SCM release is indicated if the condition persists beyond 6-8 months.[26]

Conclusions

The various etiologies of a neck mass in children highlight the importance of a logical approach to diagnosis. Commonly, pediatric neck masses are due to benign conditions that are easily diagnoses and treated. The pediatric otolaryngologist should facilitate prompt and accurate diagnosis.

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