

TITLE: Evaluation and Management of the patient with a neck mass

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Introduction

A mass in the neck is a common clinical finding that presents in patients of all age groups. The differential diagnosis may be extremely broad, and although most masses are due to benign processes, malignant disease must not be overlooked. Therefore, it is important for physicians to develop a systematic approach for developing a working diagnosis and management plan for the patient.

Anatomy

The prominent landmarks of the neck are the hyoid bone, thyroid cartilage, cricoid cartilage, trachea, and sternocleidomastoid muscles. In females, the cricoid cartilage is often the most palpable laryngeal structure, whereas in men, the thyroid cartilage is most easily palpable. The SCM divides each side of the neck into two major triangles, anterior and posterior. The anterior triangle is delineated by the anterior border of the SCM laterally, the midline medially, and the lower border of the mandible superiorly. The anterior triangle can be further divided into the inferior carotid (muscular), superior carotid, submandibular and submental triangles. The borders of the posterior triangles are the posterior border of the SCM anteriorly, the clavicle inferiorly, and the anterior border of the trapezius muscle posteriorly. The omohyoid muscle divides this triangle into the subclavian and occipital triangles. The floor of the posterior triangle is formed by the splenius capitis, levator scapulae and scalene muscles.

The carotid bulb can be palpated near the anterior border of the SCM, near the level of the hyoid bone. In slender persons or those with significant atherosclerosis, it can be mistaken for an abnormal mass, especially when asymmetric.

The lymphatic flow in the normal neck follows a predictable pattern and the presence of a mass in a lymphatic nodal chain location may offer clues to identifying a primary tumor or site of infection. It is convenient to use the level system to describe the location of lymph node disease in the neck. Level I contains the submental and submandibular nodes. Levels

II through IV contain lymph nodes along the jugular vein (upper, middle and lower). Level V contains the lymph nodes located along the lower half of the spinal accessory nerve and the transverse cervical artery. The supraclavicular nodes are included in the posterior triangle group. The pre- and paratracheal nodes, precricoid (Delphian) node, and perithyroidal nodes are found within level VI.

General Considerations

The first consideration should be the patient's age group. Three main age groups need to be considered: pediatric (0 to 15 years), young adult (16-40 years) and late adult (>40 years). In general, neck masses in children are more commonly inflammatory than congenital or developmental and more commonly congenital than neoplastic. This distribution is similar in the young adult. However, the first consideration in the late adult should be neoplasia. The "rule of 80" is often applied, which states that 80% of non-thyroid neck masses in adults are neoplastic and that 80% of these masses are malignant. A neck mass in a child, on the other hand, has a 90% probability of being benign.

The next consideration should be the location of the mass. The location of the mass is particularly important with respect to congenital and developmental masses, because such lesions are consistent in location. For metastatic neck masses, their location may be the key to identification of the primary tumor. It is important despite the general considerations of age and location to treat each case individually.

Diagnostic Steps

Evaluation of the patient with a neck mass must begin with a careful and complete history and a thorough head and neck examination. A thorough review of the developmental time course of the mass, associated symptoms, personal habits, and prior trauma, irradiation or surgery is important. Inquiries about smoking and alcohol use, fever, pain, weight loss, night sweats, exposure to tuberculosis, foreign travel and occupational/sexual history should be made. In the late adult patient, symptoms of dysphagia, otalgia, and/or hoarseness with a smoking history most likely represent a neoplastic process.

The examiner should not pay undue attention to the neck mass neglecting the complete examination of the head and neck. All mucosal surfaces of the nasopharynx, oropharynx, larynx and nasal cavity should be visualized by direct examination or by indirect mirror or fiberoptic visualization. The oral and pharyngeal surfaces should be digitally palpated in addition to the neck mass. Emphasis on location, mobility and consistency of the neck mass can often place the mass within a general etiologic grouping, such as vascular, salivary, nodal/inflammatory, congenital or neoplastic.

A tender, mobile mass or a high suspicion of inflammatory adenopathy with an otherwise negative examination may warrant a clinical trial of antibiotics and observation not to exceed two weeks with close follow-up.

Diagnostic studies

Fine Needle Aspiration Biopsy (FNAB) -

Currently, FNAB is the standard of diagnosis for neck masses and is indicated in any neck mass that is not an obvious abscess and persists following prescribed antibiotic therapies. FNAB separates inflammatory and reactive processes that usually do not require surgery from neoplastic lesions, either benign or malignant. It also may allay patient fears for malignant disease and helps the clinician differentiate carcinoma from lymphoma, which can prevent unnecessary panendoscopy. There are no contraindications to FNAB. Pulsatile neck masses may represent a carotid body tumor, and although many clinicians prefer not to biopsy these lesions, the fine gauge of the needle reduces bleeding complications. Needle-track seeding of tumor is not a concern with the fine needles used today. FNAB can also be performed in children, however, in a child younger than 2 years of age, immobilization may be necessary. Children ages 2 to 7 can be sedated, and children above age 7 are fairly cooperative. With thyroid aspiration in the pediatric population, it is preferable to have the child asleep.

A skilled cytopathologist is critical to the efficacy of FNAB. On-site evaluation carries the advantage of assessing specimen adequacy, which reduces the number of unsatisfactory specimens. When a cytopathologist is not available on-site, the clinician may perform the FNAB and send the fixed slide to a regional specialist for review. In these circumstances, a minimum of four separate needle passes and preparation of smears of good quality produce optimal results.

Ultrasonography -

With the current accuracy of FNAB, this study has become less important in the work-up of the neck mass. However, it is sometimes useful in differentiating solid from cystic masses and congenital cysts from solid lymph nodes and glandular tumors.

Radionucleotide Scanning –

This imaging technique can differentiate a mass from within a gland from one outside a glandular structure and can also indicate the functionality of the mass. This is particularly important for salivary and suspected thyroid gland masses. The FNAB is now preferred in the evaluation of thyroid nodules and is currently indicated for solitary thyroid nodules, multinodular goiters with a new increasing nodule and patients with Hashimoto's who develop a new nodule.

Computed Tomography (CT) –

CT scanning of the neck has become a very helpful tool in diagnostically difficult cases. It can distinguish cystic from solid lesions, define the origin and full extent of deep, ill-defined masses, and when used with contrast can delineate vascularity or blood flow. In patients with metastatic SCCa to the neck from an unknown primary, CT should be obtained to detect an unknown primary lesion and to help with staging purposes. Lucent changes

within nodes, size larger than 1.5cm, and loss of sharpness of nodal borders are often signs of metastatic carcinoma. Contrast CT should be withheld in the suspected thyroid lesion to avoid administering iodinated material that could later interfere with radioactive-iodine imaging studies or therapy.

Magnetic Resonance Imaging –

MRI provides much of the same information as CT. It is currently better for upper neck and skull base masses due to motion artifact on CT. With contrast it is good for vascular delineation and may even substitute for arteriography in the pulsatile mass or mass with a bruit or thrill.

Nodal Mass Workup in the Adult

If the clinical evaluation of a neck mass does not lead to a definitive diagnosis, malignancy must be excluded. An asymptomatic cervical lump is the presenting symptom in about 12% of head and neck cancer cases. Of these cancers, approximately 80% are squamous cell carcinoma (SCCa). A history of alcohol and/or tobacco use should strengthen the clinician's suspicion. Also, ipsilateral otalgia with normal otoscopy should direct the examination to the tonsil, tongue base, supraglottis, and hypopharynx. Another ear-related sign of a head and neck primary is unilateral serous otitis in the adult patient. A careful nasopharyngeal exam is mandatory in this situation.

If careful repeated examination in the office does not identify a primary site, the entire aerodigestive tract must be examined endoscopically under anesthesia. Biopsies should be performed on any suspicious mucosal lesions observed or in any suspicious areas noted on CT or MRI. If no lesions or imaging abnormalities are noted, biopsies of the nasopharynx, tonsils (including an ipsilateral tonsillectomy for jugulodigastric nodal disease), base of tongue, and pyriform sinuses should be obtained. It is important at this time to examine thoroughly all areas of the aerodigestive mucosa, including the esophagus, to identify the primary or any synchronous lesions, which occur in between 10% and 20% of patients with head and neck malignancies.

In a recent study of SCCa of the neck from an unknown primary at the University of Florida, the primary tumor was able to be detected about 40% of the time. In approximately 80% of these cancers, the base of tongue or tonsillar fossa was the culprit. The ability to find the primary lesion greatly depended on suspicious findings during physical examination and on radiographic studies. If no suggestive findings on exam or CT were obtained the yield dropped to below 20%.

If a negative/equivocal FNAB is obtained yet suspicion for malignancy persists, an open excisional biopsy of the cervical lymph node may be performed. The patient and surgeon should be prepared to proceed with a complete neck dissection depending on results of frozen sections. If the biopsy shows only inflammatory or granulomatous changes, culture of the tissue is warranted. A result of adenocarcinoma or lymphoma dictates closure of the wound and further workup and staging procedures prior to further treatment decisions.

Primary Tumors

Thyroid Masses

Thyroid neoplasms, either benign or malignant, are a leading cause of anterior compartment neck masses in all age groups. In the adult, they have a female predominance and are mostly benign. Lymph node metastasis may be the initial symptom in 15% of papillary carcinomas and micrometastasis may be present in over 90% of cases. Fine-needle aspiration of thyroid masses has become the standard of care, replacing ultrasonography and radionuclide scanning in the assessment of thyroid nodules, although ultrasound may be performed to determine if the mass is cystic or to guide FNAB for small nodules. It has been shown that FNAB has decreased the number of patients being treated with surgery, increased the number of malignant tumors found at surgery, and doubled the number of cases being followed up. Unsatisfactory aspirates should be repeated, and negative aspirates should be followed up with a repeat examination and FNAB a month later.

Lymphoma

Lymphoma can occur in all age groups, but is a more common neoplastic process in pediatric and young adult groups. As many as 80% of children with Hodgkin's disease have a neck mass. Progressive enlargement of a lateral neck mass is often the only sign of disease in the head and neck. Systemic signs of fever, hepatosplenomegaly and diffuse adenopathy should be sought. The mass often appears discrete, rubbery and nontender. FNAB is the first line diagnostic test, and when it suggests lymphoma, an open biopsy with histologic examination is appropriate. Work-up of head and neck lymphoma includes CT scans of the chest, abdomen, head and neck and bone marrow biopsy.

Salivary Tumors

An enlarging mass anterior or inferior to the ear or at the angle of the mandible or submandibular triangle should raise suspicion for a primary salivary gland tumor. Benign tumors are usually asymptomatic, however rapid growth, skin fixation or cranial nerve palsies (VII, XII and lingual) suggest malignant disease. The diagnostic test of choice is open excisional biopsy-either submandibular gland excision or parotidectomy. However, FNAB has been shown to reduce the number of patients being treated with surgery by 33%. Some apparent neoplastic lesions on exam, may be intraparotid lymph nodes, localized sialadenitis, benign lymphoepithelial lesions or cysts. The accuracy of FNAB in salivary glands is greater than 90% and more exact in benign than malignant tumors. Its sensitivity is 90% with a specificity of approximately 80% for salivary glands. If signs of malignancy are noted, FNAB may facilitate surgical planning and patient counseling. In the case of the unknown primary, the surgeon and patient must be prepared for a total parotidectomy and facial nerve dissection with possible nerve sacrifice.

Carotid Body and Glomus Tumors

These tumors are rare in the pediatric patient. In adults, they classically appear in the upper anterior triangle at the carotid bifurcation as a pulsatile, compressible mass, which is mobile from side to side. A bruit or thrill is present and in glomus vagale tumors, the

ipsilateral tonsil may pulsate and/or deviate to the midline. The diagnosis is confirmed by angiography or CT scan. In the elderly patient, close observation or treatment with irradiation to arrest growth is adequate. In young patients, a small tumor should be resected under hypotensive anesthesia. Preoperative embolization is used routinely. Preoperative measurement of catecholamine release should precede removal.

Lipoma

Lipomas are ill-defined, soft masses usually appearing in patients over the age of 35 years. They are usually asymptomatic and diagnosed by clinical findings. Diagnosis is confirmed by excisional biopsy.

Neurogenic Tumors

Neurogenic tumors or peripheral nervous system tumors arise from neural crest derivatives including Schwann cells, perineurial cells, and fibroblasts. They include schwannomas (neurilemmoma and neurinoma), neurofibromas and malignant peripheral nerve sheath tumors (MPNST). An increased incidence is associated with heritable neurofibromatosis (NF) syndromes. Of the neurogenic tumors, schwannomas occur most commonly in the head and neck region. Most are sporadic cases, and when extracranial, 25 to 45% will occur in the head and neck. MPNST is uncommon in the head and neck region.

Schwannomas are benign, solid neurogenic tumors that can occur at any age, but are most common between the ages of 20 and 50 years. Those arising in the neck commonly occur in the parapharyngeal space. They usually present as a solitary, slowly enlarging and painless mass. On physical examination, they usually cause medial tonsillar displacement. Those originating in the vagus nerve may present with hoarseness and/or vocal cord paralysis, and when originating in the sympathetic chain may be associated with Horner's syndrome. MRI is helpful in diagnosis and shows displacement of the internal carotid artery anteriorly and laterally. For benign lesions, conservative surgical excision is the treatment of choice.

Congenital and Developmental Masses

Epidermal and Sebaceous Cysts

These are the most common of the congenital masses occurring most often in the older age group. Diagnosis is made clinically noting movement and elevation of the skin overlying the mass. Excisional biopsy confirms the diagnosis.

Branchial Cleft Cysts

A branchial cyst usually presents as a smooth, fluctuant mass underlying the SCM (second arch – most common) and often seems to appear rapidly after an upper respiratory tract infection. If infection is recent, skin erythema and tenderness may be present. Occasionally, purulent material may be expressed if a sinus tract is present. These masses most commonly occur in late childhood or early adulthood, but may occur later in life also.

The more common 2nd branchial cleft cyst often has a tract coursing medially over the 12th nerve between the internal and external carotids to the tonsillar fossa. The less common 1st branchial cyst occurs along the inferior or angle of the mandible or below the ear lobe. Close association with the facial nerve is possible, so excision of this type may necessitate a total parotidectomy with facial nerve dissection. In general, treatment is with initial control of infection, followed by surgical excision. Methylene blue injection into a draining tract prior to excision may facilitate removal.

Thyroglossal Duct Cysts

This is the most common congenital neck mass, which presents as a midline or near-midline mass that usually elevates on swallowing or protrusion of the tongue. This clinical finding distinguishes the TDC from other midline masses, such as lymph nodes, dermoids or ectopic thyroid tissue, which are included in the differential. Treatment is surgical removal including the midportion of the hyoid bone (Sistrunk procedure) after resolution of infection.

Vascular Tumors

Lymphangiomas and hemangiomas are considered congenital lesions almost always present within the first year of life. While lymphangiomas usually remain unchanged into adulthood, hemangiomas most often resolve spontaneously. The lymphangioma mass is soft, doughy, and ill-defined. CT or MRI may help define the extent of the neoplasm, and treatment includes excision for easily accessible lesions or those affecting vital functions. Often surgical treatment precipitates further disease and recurrence is common. Hemangiomas, on the other hand, often appear bluish and are compressible. CT or MRI again help the diagnosis and define the extent of disease. Surgical treatment is reserved for lesions with rapid growth, associated thrombocytopenia or involvement of vital structures that fail medical therapy (steroids, interferon).

Inflammatory Disorders

Lymphadenitis

Acute lymphadenitis is very common at some point in almost everyone's life, especially during the first decade. The presentation with marked tenderness, torticollis, trismus and dysphagia with systemic signs of infection is seldom a diagnostic challenge to the clinician and the source of the reactive lymphadenopathy is usually easily identified. Initial treatment with directed antibiotic therapy and follow up is the rule.

Inflammatory nodes generally regress in size. If the lesion does not respond to conventional antibiotics a biopsy is indicated after complete head and neck work-up. Other indications for FNAB of lymphadenopathy include progressively enlarging nodes, a solitary and asymmetric nodal mass, supraclavicular mass or persistent nodal masses without active infectious signs. Equivocal or suspicious FNAB in the pediatric nodal mass requires an open excisional biopsy to rule out lymphoma or granulomatous disease.

Granulomatous lymphadenitis

These infections usually develop over weeks and months, often with minimal systemic complaints or findings. They may be the result of typical or atypical mycobacteria, actinomycosis, sarcoidosis, or cat-scratch fever (*Bartonella*). The glands tend to be firm, with some degree of fixation and injection of the overlying skin. They may suppurate and drain only to reform. Tuberculosis is now rarely seen in our population and is more common in adults within the posterior triangle. Atypical mycobacteria and cat-scratch fever are more common and more prevalent in the pediatric age group. Atypical mycobacterial infection usually involves anterior triangle lymph nodes often with brawny skin, induration and pain, while cat-scratch commonly involves the preauricular or submandibular nodes. Typical TB lymphadenitis often responds to anti-tuberculosis medications. Cat-scratch often undergoes spontaneous resolution with or without antibiotic treatment. Atypical mycobacterial infection usually responds to complete surgical excision.

Summary

The differential diagnosis of neck masses is extensive and varies with the age of the patient at presentation. A thorough work-up including an accurate history and complete head and neck examination often narrows the diagnostic possibilities, thus obviating the need for excessive testing and invasive procedures. The fine needle aspiration biopsy has become an invaluable tool to aid clinicians in the evaluation of the neck mass and is safe, accurate, and cost-effective with minimal complications. The possibility of malignancy in any age group, especially in the late adult group, should never be overlooked. Close follow-up and aggressive pursuit of a diagnosis with appropriate work-up facilitates a timely and accurate treatment plan, which is essential to a favorable outcome.

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