Chapter 28 Cervical Lymphadenopathy

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Cervical lymphadenopathy is common in children. The condition frequently results in a child's referral to a pediatric surgeon for further evaluation, and surgical intervention is often required. The majority of these masses represent benign disease, but the possibility of a malignancy exists. Parents often experience a significant amount of anxiety, and so it is important that pediatric surgeons are comfortable with the evaluation and management of these common lesions.

In general, neck masses in children can be congenital, neoplastic or inflammatory. Not all of these lesions cause cervical lymphadenopathy. Congenital lesions, including thyroglossal duct cysts, branchial cleft cysts, dermoid cysts, hemangiomas and lymphangiomas, are a part of the differential diagnosis of a neck mass in a child. Neoplastic causes of neck masses include relatively uncommon primary tumors such as neuroblastoma and rhabdomyosarcoma. Neoplasia that results in cervical lymphadenopathy is much more common and includes lymphoma and metastatic disease (most commonly thyroid cancer).

Inflammatory lesions are the most common etiology of cervical lymphadenopathy. Acute lymphadenitis, either viral or bacterial, is most often seen. Pediatric surgeons, however, will also encounter cases of subacute or chronic lymphadenitis, and the management of these can differ significantly. Some of the causes of these more indolent infections include atypical mycobacteria, tuberculosis, Bartonella henselae (cat scratch disease), and rarer fungal, parasitic or opportunistic infections. Finally, there are several miscellaneous conditions that can cause cervical lymphadenopathy in children as well.

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Acute Cervical Lymphadenitis

Acute cervical lymphadenitis in children is most commonly associated with a viral respiratory tract infection. The lymph nodes generally undergo reactive hyperplasia due to the viral infection and are usually bilateral, multiple, and small. Erythema is uncommon, and suppuration rarely occurs. Viral agents frequently associated with a respiratory tract illness include adenovirus, coronavirus, influenza virus, parainfluenza virus, reovirus, respiratory syncytial virus, and rhinovirus. Other common viruses that cause cervical lymphadenitis include Ebstein-Barr virus and cytomegalovirus. Less common causes include measles, mumps, rubella, and varicella. When bilateral enlarged lymph nodes appear in conjunction with typical upper respiratory tract infection symptoms, further work-up is not immediately necessary. The lymph node enlargement generally subsides spontaneously within 2-3 weeks. Enlarged nodes that persist beyond this time or continue to enlarge will likely require further investigation.

Acute bacterial lymphadenitis in children usually occurs due to an infection by Staphylococcus aureus or Streptococcus pyogenes. In infants less than 1 year of age, Group B Streptococcus, Haemophilus influenza type B, and anaerobes (Bacteroides, Peptococcus, Peptostreptococcus species) are possible causative agents. The lymph node groups that are affected, in decreasing order of frequency, include the submandibular, upper cervical, submental, occipital, and lower cervical nodes. The adenopathy is occasionally bilateral but more often unilateral. The involved node is usually solitary, large, and tender. Erythema and suppuration are common. Other associated findings include fever, pharyngitis, malaise, otitis, tonsillitis, dental caries, or periodontal disease.

Initial treatment is with antibiotics unless there is obvious suppuration requiring incision and drainage. Antibiotic therapy should be directed at the most likely organism. In general, an antibiotic with broad spectrum coverage, particularly against beta-lactamase producing organisms, is instituted first. However, coverage against methicillin-resistant S. aureus might be necessary given the increasing prevalence of this organism in the community. In about 25% of cases, one finds that an initially firm and tender lymph node, initially associated with mild overlying erythema, will suppurate after the institution of antibiotics. Incision and drainage or aspiration may then be required.

Subacute and Chronic Cervical Lymphadenitis

Lymphadenitis that persists beyond approximately 2 weeks is considered to be subacute or chronic. These localized infections can be caused by a variety of organisms. Atypical mycobacteria, specifically Mycobacterium avium intracellulare and M. scrofulaceum, are the most common cause of subacute lymphadenitis. Other less common strains include M. kansasii, M. fortuitum, and M. hemophilum. Patients with atypical mycobacterial lymphadenitis commonly present with a rapid onset of unilateral lymph node enlargement near the angle of the mandible. Typically, the nodes are only mildly tender and gradually increase in size over the course of 2-3 weeks. Erythema, induration, and fluctuance are often present. The skin overlying the nodes often becomes dry and flaky, and can develop a pink or purple hue. Patients rarely have other symptomatology, and a tuberculin skin test is at most mildly reactive. Antibiotic therapy is generally ineffective, and excision of the involved nodes is indicated. Simple drainage can sometimes lead to formation of a chronic draining fistula or simply a recurrence and therefore should be avoided.

Chronic lymphadenitis due to *M. tuberculosis* has a similar appearance to the other atypical infections. However patients may have constitutional symptoms. There is usually systemic disease as evidenced by an abnormal chest radiograph. A tuberculin skin test is positive, and there is a history of contact with an infected individual. Multi-agent antituberculous antibiotic therapy for 12–18 months is indicated.

Cat scratch disease is a lymphocutaneous disorder in which regional lymphadenitis occurs after infection with the bacterium B. henselae. There is usually a skin lesion in the area of inoculation. Over the course of days to weeks after inoculation, regional adenopathy occurs. The neck is the second most commonly affected area after the axilla. Patients sometimes have mild constitutional symptoms. Typically, there is a single enlarged lymph node in the chain that drains the area of inoculation. The lymph node is usually tender and firm, and suppuration can occur. If cat scratch disease is suspected, the diagnosis can be confirmed by serologic testing. The infection usually is self-limited, but antibiotic therapy with a macrolide antibiotic is often helpful in facilitating resolution of the adenopathy. Once the diagnosis has been confirmed, surgical intervention is not necessary unless purulence develops, in which case incision and drainage may be necessary.

Fungal infections is occasionally the cause of cervical lymphadenopathy in children. Histoplasmosis, blastomycosis, and coccidiomycosis are examples of these infections and are caused by *Histoplasma capsulatum*, *Blastomyces dermatitidis* and *Coccidioides immitis*, respectively. These organisms, which are endemic to certain regions of the country, usually cause a pulmonary infection that subsequently leads to involvement of cervical lymph nodes. Most cases are self-limited, but severe infections require systemic anti-fungal therapy.

Toxoplasmosis is caused by the consumption of tainted meat or milk products. The intracellular protozoan *Toxoplasma gondii* is the causative organism. Lymphadenopathy, which can be only mildly symptomatic, is the presenting symptom in 10% of patients. The diagnosis is confirmed by serologic testing. Severe cases should be treated with 4–6 weeks of antibiotics.

Opportunistic infections in immunocompromised children can also be a cause of chronic cervical lymphadenopathy. For instance, Nocardia species are ubiquitous pathogens found in the environment that only cause infections in immunosuppressed hosts. The infections are acquired through the skin or by way of the respiratory tract. Direct skin contact may result in a localized pustule, which can be cultured to establish a diagnosis. Nocardia infections can also cause significant adenopathy, in which case biopsy and culture of the node itself establishes the diagnosis. Sulfonamides are the treatment of choice. Actinomyces species are oral commensal organisms in humans. However, in hosts with compromised defense barriers, local invasion results in craniofacial actinomycosis and cervical nodal involvement. The diagnosis is difficult to make, but sulfur granules may be seen on histologic examination of an involved lymph node. Human immunodeficiency virus in children is usually acquired by vertical transmission from mother to child. Adenopathy is often a prominent manifestation and is sometimes the presenting sign. The diagnosis is made by serology, and the treatment is medical.

Malignancy

Cervical lymphadenopathy can also be the result of neoplasia in children, although statistically this is much less common than an inflammatory cause. By far the most common etiology of neoplastic lymphadenopathy in the neck is lymphoma. The cervical lymph node chains may be the prominent lymph node basin harboring the systemic disease, or they may be associated with a mediastinal mass. Lymphomas generally fall into two histologic subtypes: Hodgkin's disease and non-Hodgkin's lymphoma.

Hodgkin's disease (HD) accounts for approximately 40% of childhood lymphomas. In the pediatric age group, HD generally occurs in adolescents, and is rare in children less than 10 years of age. It is characterized histologically by the pathognomonic Reed–Sternberg cells. The four classic subtypes include nodular sclerosing, mixed cellularity, lymphocyte predominant, and lymphocyte depleted. HD arises in

the lymph node itself. Patients will often have constitutional ("B") symptoms such as fever, night sweats, or unintentional 10% or greater weight loss in the preceding 6 months. The nodes are generally nontender, firm, and rubbery. Solitary nodes are usually mobile, whereas aggregates of nodes may be bulky and fixed to the underlying tissue.

Non-Hodgkin's lymphoma (NHL) accounts for approximately 60% of childhood lymphomas. It most commonly occurs in children 7–11 years of age, and there is a 3:1 male to female predominance. NHL are divided into small-cell noncleaved (Burkitt's and non-Burkitt's), lymphoblastic, and large cell lymphomas (anaplastic and diffuse large B cell). Ten percent of patients with NHL have head and neck involvement. The neoplasm itself may or may not arise in nodal tissue. Often there is an aggressively enlarging mass causing local symptoms due to invasion of bone, nerves or soft tissue, and constitutional symptoms may be present.

Cervical lymphadenopathy in children may also be caused by metastatic disease. Metastasis from a thyroid carcinoma sometimes present as unilateral lymph node enlargement. When this occurs, it is important not to disregard the mass as ectopic thyroid tissue, and a search for a thyroid mass should be undertaken. Patients with stage 4 neuroblastoma sometimes present with cervical lymphadenopathy, often bilateral. In these cases, the diagnosis is made on biopsy of the enlarged lymph node.

Miscellaneous Causes of Cervical Lymphadenopathy

There are numerous other causes of cervical lymphadenopathy in children that a pediatric surgeon should be familiar with. When the more common inflammatory and neoplastic causes have been ruled out, one must consider some of these esoteric conditions. In general, a biopsy of an involved node is required to make the diagnosis of one of these diseases.

Uncommon infections can lead to lymph node enlargement. An infection due to *Francisella tularensis* causes tularemia ("rabbit fever"), a serious infectious disease that occurs in humans after contact with infected rodents. *Yersinia pestis* is the causative organism of the plague. The vector of infection is a flea, and bites in the head and neck region can cause regional adenopathy. *Pasteurella multocida*, an organism transmitted from animal bites, is another unusual cause of cervical adenopathy.

Sarcoidosis is a chronic granulomatous disease that can affect children. Pulmonary involvement is common, but peripheral lymphadenopathy also readily occurs. The involved lymph nodes are usually bilateral, firm, and rubbery. Children can have cervical lymphadenopathy from sarcoidosis, and the diagnosis is made by biopsy of one of the affected nodes. The treatment is medical.

Kawasaki disease, or mucocutaneous lymph node syndrome, is an acute vasculitis in which there is inflammation of small and medium-sized blood vessels throughout the body. The peak age is between 1 and 2 years and 80% of cases occur before the age of 4. The etiology of the condition is unknown. Inflammation may occur in cervical lymph nodes early in the course of the disease. The involved nodes are usually confined to the anterior triangle of the neck on one side. The nodes are sometimes large (>1.5 cm) and are tender and non-fluctuant. The disease and the adenopathy are self-limited.

Kikuchi–Fujimoto disease, also known as histiocytic necrotizing lymphadenitis, is a rare condition of unknown etiology. Patients present with bilateral, painful, enlarged lymph nodes in the posterior triangle of the neck. Constitutional symptoms are present, and children can develop splenomegaly as well as a skin rash. The diagnosis is confirmed by excisional biopsy of an affected lymph node. The treatment is supportive as the disease rarely causes significant morbidity and is self-limited.

Rosai–Dorfman disease, also known as sinus histiocytosis and massive lymphadenopathy, is a rare disease of unknown etiology that occurs in young children. Cervical lymphadenopathy commonly occurs as proliferating histiocytes accumulate in lymph nodes. The lymph nodes are initially mobile and discrete. However, as the condition progresses, there is massive enlargement of the cervical lymph nodes as well as other nodal regions. The disease may resolve spontaneously, however progression of the disease requires chemotherapy to control associated histiocytosis and plasmacytosis.

Castleman's disease, or giant lymph node hyperplasia, may cause a unicentric or multicentric adenopathy. The disease is caused by the hypersecretion of the cytokine IL-6. Excision of the involved node can be curative.

Periodic fever, aphthous stomatitis, pharyngitis and cervical adenitis (PFAPA) syndrome is a disease of unknown etiology that occurs in young children. Patients have cyclic recurrences of the above symptoms every 3–5 weeks, and are healthy in between episodes. Corticosteroids have been used to alleviate symptoms during flare-ups but the episodes generally abate with time.

Preoperative Preparation

The most important aspect of the work-up for cervical lymphadenopathy is a properly obtained history and a thorough physical examination. The history should elicit whether the adenopathy has occurred acutely or has become a chronic condition. It should be determined whether there has been a recent upper respiratory infection or if there has been contact with an individual with typical URI symptoms. Any recent

travel and any contact with animals, especially cats, should be noted. The presence of other symptoms in the patient is also important. For example, the acute onset of pain and swelling should raise the suspicion of acute lymphadenitis. Related constitutional symptoms such as fever, night sweats, and weight loss might indicate a disseminated process such as a lymphoma. Physical examination, especially serial exams by the same practitioner, is of particular importance. Pertinent findings to note on exam include the laterality, size, number, and mobility of the lymph nodes. In addition, the presence of tenderness, overlying skin changes, erythema, induration, or fluctuance should be determined.

Laboratory studies are also an important aspect of the work-up for cervical lymphadenopathy. Basic studies like a complete blood count with differential and a peripheral smear should be obtained. Serologic studies can be obtained to confirm infections due to CMV, EBV, or HIV. Serologies can also confirm toxoplasmosis and cat scratch disease. If fluid can be obtained from an infected lymph node it should be sent for gram stain and culture (aerobic, anaerobic, fungal, acid-fast bacilli). Finally a PPD skin test should be applied.

Imaging studies should include at least a chest radiograph to evaluate for mediastinal lymphadenopathy, as this has important implications if a biopsy under anesthesia is being considered. If further imaging of the neck or lymph node itself is necessary, an ultrasound should be obtained. The ultrasound can give information about the lymph nodes such as size, number, and whether there is normal or abnormal architecture. Additionally, the relationship of the node to adjacent structures can be determined. It is sometimes difficult to determine whether the palpable mass is in fact a lymph node as opposed to either the parotid or submandibular salivary glands. In these cases, ultrasound is useful. Occasionally, a CT or MRI can give further information regarding the relationship of a palpable node to adjacent structures, but most of the time these costly studies do not add much information.

Fine needle aspiration is a procedure to consider in helping to make a diagnosis. It is most useful for obtaining fluid from an abscessed lymph node, and can be useful for making the diagnosis of a malignancy. An FNA should also be considered when a family is very reluctant for their child to have an anesthetic and undergo an operation. However, there is the distinct possibility that the FNA will be non-diagnostic. An open biopsy will then be required to obtain an adequate amount of tissue to make a diagnosis, for confirmation of a diagnosis that was suggested on FNA or for other studies that may be necessary such as flow cytometry. It is our preference, therefore, to forego an FNA and to proceed with a single operative intervention, whether it is a drainage procedure or a biopsy, when the decision to obtain tissue has been made.

Another very important aspect of the evaluation and management of cervical lymphadenopathy is a proper discussion with the parents of the child, particularly if the child's adenopathy has become chronic. The parents are usually aware of the possibility of an infection or a malignancy, and so their anxiety level is already high. A thorough discussion of significant history and physical findings and the possible diagnoses is essential. Also, a logical explanation of the rationale for either a period of observation or proceeding with immediate operative intervention is necessary. In general, it is our practice to observe small (subcentimeter), mobile, bilateral cervical lymph nodes that have been present for less than 2–3 weeks, particularly in the presence of recent URI symptoms or a documented exposure to cats. This allows time for a proper workup, including laboratory/serology studies, a chest radiograph, and PPD. When unilateral, large (>1 cm), firm, fixed or matted nodes are present, particularly in the posterior triangle of the neck or in the supraclavicular area, we favor biopsy. It is especially important to expedite a biopsy if constitutional symptoms such as fever, night sweats, and weight loss have been present. The workup, particularly the laboratory studies and chest radiograph, also should be done expeditiously.

Once it has been decided to proceed with operative intervention, the procedure planned must be explained to the parents. Options include an FNA only, a simple incision and drainage procedure, an incisional biopsy for large, fixed lesions, or an excisional biopsy. Risks of the operation must be discussed. These include bleeding, infection, injury to adjacent structures (such as the facial nerve for lymph nodes near the angle of the jaw), and the possible need for further treatment, including another procedure for recurrent infection, to obtain more tissue, or excision of a persistent fistulous tract in the case of an atypical mycobacterial infection. Further medical management, such as antibiotics for an infection or chemotherapy for a malignancy, may be necessary and should be thoroughly discussed.

Surgical Technique

There are several operative options available, and the appropriate technique depends on the clinical scenario. For a suspected abscessed lymph node, a simple incision and drainage procedure, either with a local or general anesthetic, is all that is required. Fluid is sent for gram stain and culture. Gentle curettage followed by irrigation and packing of the cavity to prevent premature skin closure is useful. When the abscess has been caused by typical bacteria, the cavity fills and the wound generally heals without the need for any further surgical therapy. However, in cases where the abscess has been caused by an atypical mycobacterial infection, the wound

may persist and subsequently mature into a chronic draining fistula. In this case, antibiotics are generally ineffective and it is therefore necessary to reoperate for excision of the entire fistulous tract and any residual nodal tissue.

When biopsy is required, one must consider what is easiest and safest for making a diagnosis. For large or fixed lesions, an incisional biopsy may be the best choice. A nerve stimulator is useful especially when the node is located near important nerves such as the facial nerve. For smaller, easily accessible lesions that are not fixed to adjacent structures, an excisional biopsy is safe. In cases where a lymphoma is diagnosed, further excision is unnecessary, and systemic chemotherapy is instituted. This is also the case when cat-scratch disease is diagnosed, as the disease is usually self-limited.

A very important situation to consider is when a cervical lymph node biopsy is required in a patient with a large anterior mediastinal mass. This scenario is most often encountered in cases of lymphoblastic lymphoma and is one reason that a chest radiograph is an important part of the preoperative workup. In such cases, there is a significant possibility of life-threatening airway collapse upon induction of anesthesia. Once this occurs, there is little that can be done to reestablish the airway as the collapse is distal to the tip of a typical endotracheal tube. Therefore, in this situation, a CT scan is recommended preoperatively to evaluate the degree of either tracheal or bronchial compression. Some also suggest obtaining pulmonary function testing to evaluate peak expiratory flow rate (PEFR). It has been suggested that a decrease in either the tracheal cross-sectional area by one half or the PEFR by 50% of predicted for age places a patient at high risk for respiratory compromise during general anesthesia.

It is the responsibility of the surgeon and anesthesiologist to recognize this danger when a cervical lymph node biopsy is requested and to plan to perform the biopsy as an awake procedure under local anesthesia. Alternatively, if a pleural effusion is present, one can forego the lymph node biopsy and perform thoracentesis, whereupon analysis of the fluid will make the diagnosis.

Postoperative Care

The post-operative management is relatively straight forward. For incision and drainage procedures, the packing should be removed in 24–48 h. Local wound care with a topical antibiotic and gauze dressing along with frequent washing is all that is necessary. Otherwise, wounds that have been primarily closed generally heal without incident. Culture and biopsy results are shared with the parents and further therapy, if necessary, can be planned.

Cervical lymphadenopathy is common in children, and pediatric surgeons must be familiar with the evaluation and management of this condition. Lymphadenitis is the most common cause of lymph node enlargement in children. However, neoplasia and other uncommon disorders should also be considered. The patient's history, physical findings, laboratory tests and imaging studies are all important in helping to make the diagnosis and to formulate a plan of care. If acute viral cervical lymphadenitis is suspected, the enlarged lymph nodes should be closely observed for 2–3 weeks. If a severe bacterial infection, neoplasm or other unusual condition is suspected, or if the adenopathy has become chronic, then surgical intervention must be considered. The operative technique chosen is based on the characteristics of the lymph node enlargement, and one should avoid a general anesthetic when a large anterior mediastinal mass is associated with the adenopathy. Finally, it is essential to have a thorough discussion with the parents of the child regarding the rationale for the treatment plan that has been instituted.

Summary Points

- In general, neck masses in children can be congenital, inflammatory or neoplastic.
- Specifically, cervical lymphadenopathy is most commonly due to inflammation or infection and can be acute, subacute or chronic.
- Cervical lymphadenopathy due to neoplasia is most likely a lymphoma.
- Unusual causes of cervical lymphadenopathy should be considered once the more common inflammatory and neoplastic causes have been ruled out.

Editor's Comment

Few clinical issues create more anxiety for parents than an enlarged cervical lymph node. They need to know that it is not cancer and they need to know today. The experienced pediatric surgeon usually has a good feel for whether an

enlarged lymph node is something to be concerned about or can be safely observed and the parents reassured. Unfortunately the only option for sampling a lymph node in a child is a surgical procedure under general anesthesia, which is generally safe and usually straightforward, but entails a certain amount of risk and an obligatory scar. This means that the surgeon should have a high index of suspicion before recommending a biopsy. Fortunately, a period of observation is almost always safe, even in the case of a malignant process, so, when in doubt, a brief delay can help one to make the best recommendation.

In children, FNA is simply not a good option for the evaluation of cervical masses: pediatric pathologists have little experience with the technique, most children will not let you come near them with a needle, and, most importantly, the most common neoplastic processes seen in children (lymphoma and leukemia) cannot be reliably differentiated from an inflammatory process by FNA. Likewise, a neoplastic process cannot be excluded on the basis of blood tests, serologies, or medical imaging. What we are left with then is the history, the physical examination, and the growth pattern of the node. A lymph node that is larger than 1.5 cm and continues to grow over time, especially if it is located in an unusual location (supraclavicular), should be excised. Likewise, the patient with constitutional symptoms (the presence or absence of which should be specifically documented at the initial visit) should undergo biopsy.

A typical busy pediatric surgeon will see at least one or two children with an enlarged lymph node every week. Most can be simply observed with no further studies, but nearly all should be encouraged to return for at least one follow-up visit in 2–3 weeks. At the other extreme is the rare patient with systemic symptoms and a worrisome node that clearly needs to be excised for biopsy. These patients should be scheduled for surgery and at minimum have a CBC with differential and a chest X-ray to rule out a mediastinal mass. The remainder will have clearly pathologic lymphadenopathy but no clear indication that a neoplastic process is necessarily involved. These patients should be scheduled for follow up in no more than 2–3 weeks and should undergo a work up: CBC w/diff.; serologies for cat

scratch, toxoplasmosis, and mononucleosis, depending on what is endemic in the area; and a chest X-ray. If there are risk factors, a PPD might be prudent. In some cases in which a bacterial lymphadenitis is suspected, an empiric 7-day trial of antibiotics is reasonable, albeit controversial. A node involved with tumor almost never get smaller without treatment, so a node that shrinks can probably be observed. However, lymphoma can regress rapidly when the patient is given corticosteroids (for example for a coincidental asthma flare), in which case a biopsy becomes imperative.

Cervical lymph node biopsy is a delicate procedure not to be taken lightly. There is always the risk of nerve injury and attention should be paid to scar placement for cosmesis and comfort. A small transverse incision placed in a skin crease is preferred. Once the platysma has been breached, the remainder of the dissection should be by careful blunt dissection only. A curved hemostat should be used to gently push adjacent tissues away from the capsule of the lymph node and nothing should be cut or cauterized. With proper technique, the node will gently rise up to meet the incision and the vessels at the hilum can be ligated or cauterized with precision right at the capsule. The goal should be complete excision of the node, but this can be done in piece-meal fashion. Lymph nodes that surprise the surgeon by being excessively vascular can be assumed to represent metastatic thyroid carcinoma (or another even less common neoplasm). The incision should be closed only at the level of the platysma and the skin as deeper sutures are not necessary and increase the risk of nerve injury. Finally, the child with an enlarged lymph node that is highly suspicious for malignancy should be evaluated by a pediatric oncologist before surgery so that a proper work up can be initiated, including a bone marrow biopsy to be performed while the patient is under general anesthesia.

Differential Diagnosis

Acute cervical lymphadenitis

- Viral
- Bacterial

Subacute and chronic cervical lymphadenitis

- · Atypical mycobacterial
- Typical or tuberculous mycobacterial
 - · Cat scratch disease
 - Fungal
 - Parasitic
 - Opportunistic

Neoplasia as a cause of cervical lymphadenopathy

- Hodgkin's disease
- · Non-Hodgkin's lymphoma
- · Metastatic disease

Uncommon causes of cervical lymphadenopathy

- · Unusual infections
- Sarcoidosis
 - Kawasaki's disease
 - Kikuchi-Fujimoto disease
 - Castleman's disease
 - · Rosai-Dorfman disease
 - PFAPA syndrome

Diagnostic Studies

- Complete blood count with differential
- Peripheral blood smear
- Serology testing
- PPD
- Chest radiograph
- Ultrasound
- CT, MRI (rarely necessary)
- Consider FNA

Parental Preparation

- Frank discussion regarding possible etiologies.
- Discussion of treatment options:
 - Period of observation
 - Operative intervention
- Discussion of possible complications of surgical intervention:
 - Bleeding
 - Wound infection
 - Nerve injury
 - Recurrence
- Discussion of the possible need for further therapy:
 - Antibiotics for infection
 - Re-excision for recurrence or fistula
 - Chemotherapy for lymphoma

Preoperative Preparation

- □ Review results of laboratory and imaging studies
- □ Informed consent

Technical Points

- Consider using a nerve stimulator.
- Perform incision and drainage for an abscess.
- A large mass should be sampled by incisional biopsy.
- Perform excisional biopsy for smaller lymph nodes.
- Perform complete excision of nodes for suspected atypical mycobacterial infection.
- If a lymph node biopsy is being performed in a patient with a large mediastinal mass, strongly consider performing the biopsy awake under local anesthesia to avoid life-threatening airway compromise.

Suggested Reading

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