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CASE REPORT

Enlarging Painful Neck Mass in a Child: Urgent or Elective?

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ABSTRACT

Four pediatric patients with enlarging, painful, and acutely infected lateral neck masses presented to the emergency department of a tertiary care Children's Hospital are described. These masses include suppurative lymphadenitis, brachial cleft anomaly, dermoid cyst, and an infected lymphatic malformation. Differences between these four disease processes in presentation, exam findings, and radiological features are highlighted to help determine the level of acuity in care. The immediate and subsequent management of the patients is described to help guide subspecialty consultations for intervention.

Keywords: Brachial cleft cysts, Dermoid cyst, Lymphatic malformation, Pediatric neck mass, Suppurative lymphadenitis.

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CASES

Case 1

An otherwise healthy 14-year-old female first presented to the emergency department (ED) with a tender left-sided neck mass that was progressively enlarging over the course of a few months. She was treated with a course of augmentin for lymphadenitis with slight improvement. Her pain had resolved, but the mass persisted. She did not have any overlying skin erythema, or systemic symptoms such as fever, chills, or preceding upper respiratory infections.

She subsequently presented to the pediatric otolaryngology outpatient clinic with a left-sided 3 2 cm globular level II neck mass that was mobile, discrete and nontender. No cervical lymphadenopathy was present. Computerized tomographic (CT) scanning was performed which demonstrated a cystic mass with well-defined boarders suggestive of a brachial cleft cyst (Fig. 1). She did not have any further infections, and underwent excision 3 months later without complications and had no recurrence.

Case 2

A healthy 2-year-old female presented to the ED with an enlarging right-sided neck mass of approximately 2 cm in size. It had a sudden onset 10 days prior and was associated with pain and rapid progression in size. On a diagnosis of acute lymphadenitis, she was admitted, treated with IV clindamycin for 2 days, improved rapidly and discharged on oral antibiotics. No radiological work up was obtained.

She was presented to the outpatient clinic 2 weeks after discharge with a recurrence of the swelling and pain. There was no history of fever, chills, or loss of appetite. Examination revealed an oval-shaped mass measuring 1 1.5 cm at level II that was mobile, discrete and tender. There was erythema of the skin over the swelling. Her antibiotic was changed to high dose augmentin. She returned to the clinic one week later with a slight decrease in the size of the mass, which was now firm with an area of fluctuance. A CT scan with contrast was obtained to rule an infected congenital anomaly. It demonstrated a ring-enhancing fluid collection overlying the sternocleidomastoid muscle with surrounding edema and slightly obliterated fascial planes (Fig. 2). The patient underwent

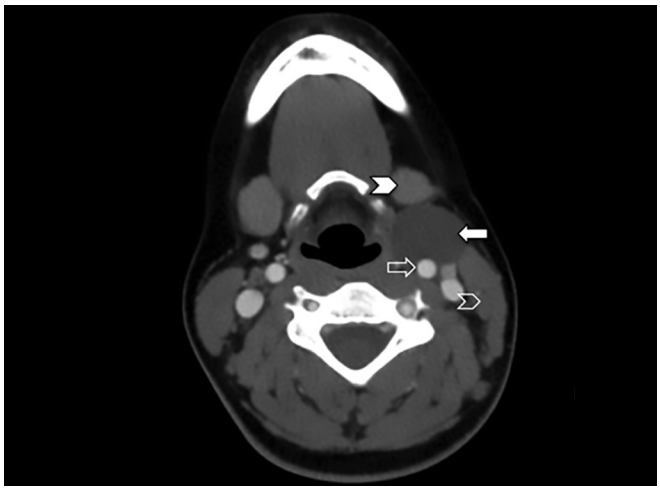

Fig. 1: Brachial clef cyst. Contrasted CT of the neck. Axial view. Image shows a left sided cystic mass (filled arrow) that is well-defined with homogenous content located posterior to the submandibular gland (arrow head), anterior to the carotid artery (empty arrow), and medial to the sternocleidomastoid muscle (empty arrow head).

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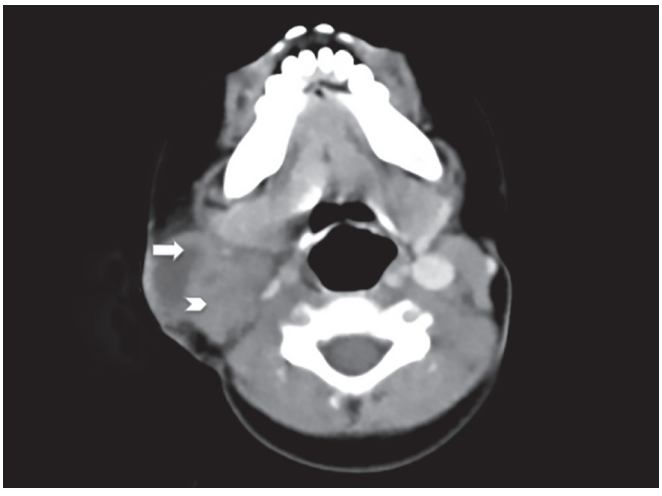

Fig. 2: Abscess. Contrasted CT of the neck. Axial view. Image shows a mass (arrow) with rim enhancement, low density, and surrounding fat stranding and loss of fasdal planes that is located superficial to the sternocleicioniastoid muscle (arrow head)

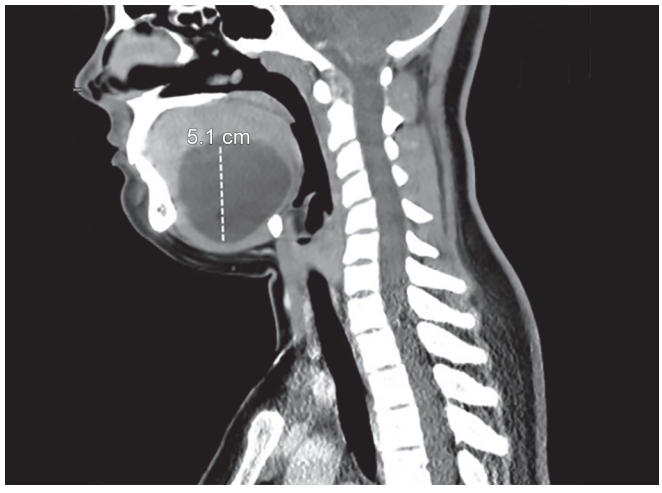

Fig. 3: Dermoid cyst. Contrasted CT of neck. Sagittal view. Image shows a mass measuring 5.1 cm

incision and drainage in the operating room. Purulent fluid and necrotic tissues were evacuated. Cultures had failed to grow any pathogens. Her recovery was uneventful with complete resolution of the mass on follow-up.

Case 3

A 17-year-old female presented to the ED with a progressively expanding right submandibular mass. It was first noticed 5 months ago and had grown increasingly tender over the previous 72 hours. A low-grade fever accompanied it. She was prescribed augmentin by her pediatrician and had four doses without relief of pain. She reported no difficulty in swallowing or breathing. She was otherwise doing well and had no other symptoms pertaining to the other systems except for those described above. Examination revealed a tender globular mass in the right submandibular space measuring approximately 4 3 cm. It was partially mobile, discrete, and did not appear attached to the mandible. It was bimanually palpable but did not displace the tongue. The skin over the swelling was erythematous but mobile. No significant clinical lymphadenopathy was present.

A CT scan of the neck showed a 5 5.5 cm mass in the right submandibular space abutting the midline and impinging on the lingual musculature (Fig 3). There was no evidence of abscess formation or compression of the airway. The pediatric otolaryngology service was consulted to rule out any airway impingement by the mass. A flexible laryngoscopy confirmed a normal patent airway with no displacement of the base of the tongue. She was asked to continue the antibiotic therapy and return if it got worse or difficulty in breathing or swallowing intervened. She had improved and was seen in the pediatric otolaryngology clinic electively after 2 weeks. The mass had decreased in size and was nontender. Elective surgery was performed, and a large cystic mass was completely excised from the submandibular space. Her postoperative course was uneventful. The final pathology diagnosis was that of an epidermoid cyst.

Case 4

A 13-year-old male with a diagnosis of right-sided cervical lymphangioma followed by the pediatric otolaryngology service presented to the ED with painful enlargement of the swelling in his neck of 48 hours duration. This was subsequent to an upper respiratory tract infection. The swelling had increased to three times its original size and was restricting neck movement. Examination revealed a tense, cystic, and painful swelling in the right neck below the mandible that was 6 4 cm in size. It had restricted mobility, and the skin over the swelling was inflamed. T1 with gadolinium magnetic resonance imaging (MRI) of the neck suggested an abscess (Fig. 4). He was admitted to the pediatric service and started on ampicillin-sulbactam and clindamycin. Later in the day, he was taken to the operating room where 30 cc of pus was drained. His postoperative recovery was uneventful, and he was discharged on the same combination of oral antibiotics. The patient later had sclerosing of the lesion by interventional radiology.

DISCUSSION

Painful enlarging neck masses are frequent complaints among children visiting the ED. Differential diagnosis of pediatric neck masses can be classified based on pathophysiology: congenital, inflammatory, traumatic or neoplastic, location: lateral vs midline neck masses or appearance: cystic vs solid.[1,2] This presentation to the ED of four pediatric patients with painful enlarging neck

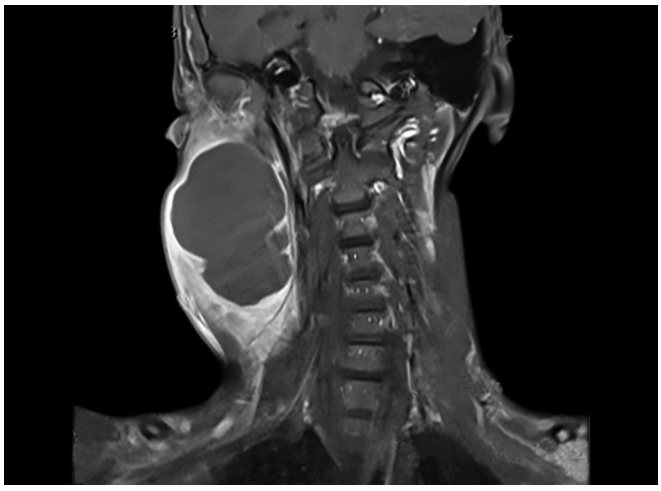

Fig. 4: Infected lymphatic malformation. T1-MRI with gadolinium

masses of different etiologies aims to guide consultation and intervention.

The most common congenital lateral neck masses are brachial cleft cysts.[1] They account for 30% of all congenital neck masses.[3] Second brachial cleft anomalies account for 95% of all of them.[3] These are further categorized into four different types based on their location relative to the sternocleidomastoid muscles (SCM) and carotid artery. Type II is the most commonly seen subtype located medial and deep to the SCM but anterior to the carotid artery as was seen in our patient.[3] Second cleft anomalies can present as a nontender lateral neck mass, which may acutely enlarge when infected, or after an upper respiratory tract infection. Broad spectrum antibiotic therapy should be initiated along with referral for definitive surgical care. Uncontrolled infection can lead to rupture and complicated by a chronically draining fistula that often opens along the anterior border of the SCM.[3] Recurrence after surgical intervention is higher in complicated fistulas.

Acute unilateral cervical lymphadenitis is the most common acute neck mass that presents to a pediatric ED. This infectious and inflammatory neck disease in children can have a wide spectrum of presentations. These can range from a painful neck swelling to fever, restriction of neck movements to a neck abscess or full-blown systemic infection.[4] They are more often bacterial in etiology when compared to bilateral lymphadenitis. Common pathogens, such as streptococcal or staphylococcal organisms, account for 40–80% of the infections.[4] While the initial stages can be successfully treated with oral antibiotics, those with restriction of neck movements would benefit from inpatient admission and intravenous antibiotics. An ultrasound in the ED will often help distinguish between early suppuration and an abscess that requires drainage. Needle aspiration under conscious sedation is an option for an uncomplicated superficial abscess. The patient can be discharged on a course of high dose amoxicillin-clavulanic acid or clindamycin. An initial parenteral does in the ED augments the later oral doses. Our 2-year-old patient described above probably had an antibioma that required surgical drainage. This was confirmed by the drainage of purulent fluid with negative bacterial cultures.

Dermoid cysts are less common and makeup only 10% of all congenital neck masses.[5] They are thought to originate from germ cell layers that failed to fuse during embryogenesis. Compared to teratoid cysts, dermoid cysts are composed of only two germ cell layers (ectoderm and mesoderm) with contributions from hair, hair follicles, or sebaceous glands.[6] Clinical presentation includes a midline or lateral, painless, cystic mass that is often misdiagnosed as a thyroglossal duct cyst.[2] Dermoid cysts most commonly involve the orbit, followed by the oral cavity (especially the floor of the mouth), but may also involve the neck. Infection can result in an increase in size as was seen in our patient and at times make prominent an otherwise unremarkable swelling. The patient was also concerned about the cosmetic deformity caused by the progressive enlargement of the lesion. In a retrospective study of 49 patients, the most common location for dermoid cyst was periorbital (61%) followed by the neck (18%).[7] Another study found that dermoid cysts located in the neck made up 7% of cases.[8] The age of distribution for dermoid cyst are typically between 9 months and 15 years (3.7 years mean).[5]

Lymphatic malformations are made up of dilated lymphatic channels, thought to be caused by an abnormal connection between the lymphatic and venous systems. Over 60% of lymphangiomas are congenital and the majority present within the first 2 years of life.[9,10] The age of onset ranges between 9 months and 15 years (3.6 years mean).[5] Lymphangiomas are classified as macrocystic, microcystic, or mixed depending on the size of the dilated vessel involved. There are four types of these anomalies: cystic hygroma, cavernous lymphangioma, capillary lymphangioma, and vasculolymphatic malformation.[7] Cystic hygromas are the most common form of lymphangioma with 75% occurring in the neck.[11] These lesions are described as soft, compressible, infiltrative and commonly cross tissue planes.[12] Complications include hemorrhage or infection, which can present as sudden enlargement of preexisting mass. Our patient was being followed on a regular basis with a plan for sclerosing the lesion if it increased in size. An acute infection can at times result in involution due to the inflammatory process. Due to incomplete involution, he was treated with elective intervention. Evaluation of the pediatric patient with a lateral neck mass starts with a detailed history, and physical that focuses on the age, time of onset,

exposure history and duration of symptoms as well as associated systemic symptoms such as fever, chills, or preceding viral syndrome.[1] Chronicity often helps to distinguish between infectious vs. congenital processes however some congenital lesions will not manifest until acutely infected. The physical exam focuses on palpation of the mass to assess the size, location, painfulness, features of the overlying skin and mobility.[1] For masses adjacent to the floor of mouth as described in our patient with a dermoid cyst, it is important to evaluate the oral airway and tongue position. If there is any suspicion of impingement on the airway or a potential for it with the expansion of the lesion, a flexible laryngoscopy should be performed. An early otolaryngology consultation in this situation would be appropriate. When the distinction between the acute manifestation of a congenital mass and infectious lymphadenitis is not obvious, further workup should be pursued. Standard laboratory evaluation such as white blood cell count may not be the most helpful in this setting. Imaging will be the key in making the distinction.

Imaging evaluation consists of ultrasound and cross-sectional modalities. Ultrasound is a cost-effective, noninvasive, and readily available imaging modality ideal for the initial assessment as it can easily distinguish between solid mass and cystic mass.[2] It has the advantage of avoiding radiation exposure, which is extremely important in the pediatric age group. Brachial cleft anomalies on ultrasound appear as a unilocular, anechoic or hypoechoic cystic mass.[13,14] Majorities of them are derived from the second brachial cleft.[3] The typical appearance on ultrasound is a well-defined cyst without any internal debris that is located along the medial border of the sternocleidomastoid muscle and posterior boarder of the submandibular gland.[15] It is also found superficial to the great vessels of the neck.[15] An acutely infected cyst may mimic an abscess and have an ill-defined thick wall with internal debris. However, the location of these lesions will often be different from those of abscesses.[13,15] Suppurative lymphadenitis or abscess usually has a thick and ill-defined boarder with an internal collection of debris.[15] It can be multilocular, and several nodes together may form a conglomerate mass mimicking a mass lesion.[14] The capsule of the abscess can be hypervascularized and visible on Doppler ultrasound.[15] Regional reactive lymphadenitis may be present as well as surrounding edematous soft tissue and obliterated fascial planes.[15] Ultrasound is also the initial imaging choice for both dermoid cysts and lymphatic malformations. A dermoid cyst on the US would show a well-defined hypoechoic mass tissue displacement. Heterogeneous appearance may be seen due to the presence of fat, osseous, or dental structures.[16] In contrast, lymphatic malformations would reveal a compressible, multilocular cystic mass with septations of variable thickness. Fluid levels may be seen if hemorrhage occurs.[16]

CT and MRI of brachial cleft cysts often demonstrate a low attenuation cystic mass that is homogeneous in appearance and located in its typical anatomical position: superficial to the carotid artery and jugular vein, posterior to the submandibular gland and medial to the anterior sternocleidomastoid muscle.[14,15] Infected cysts can be more heterogeneous, hyperattenuated and have a more irregular rim.[14] Their appearance can also mimic cancerous metastatic lymph nodes. If clinical suspicion of malignancy is high based on history and the rest of the physical examination, fine needle aspiration (FNA) is recommended to exclude malignancy.[15] Brachial cleft cysts on MRI shows high T2-weighted signal given the intracystic fluid with occasional hyperintensity depending on the degree of internal proteinaceous content such as mucous and epithelial cells.[13,15] On CT, an abscess has rim enhancement, low density, surrounding fat stranding with loss of fascial planes.[15] MRI will show a lesion with low intensity on T1-weighted images and high intensity on T2. They are often located in the submandibular triangle and can be superficial.[14,15] Usually there is surrounding edematous soft tissue and obliterated fascial planes.[15]

On non-contrast CT, a dermoid cyst usually appears as a low-density, unilocular, and well-circumscribed mass. Fat, mixed-density fluid and calcification (<50%) may also be seen. Specifically, there is a "sac-of-marbles" appearance, in which fat coalesces into small nodules within the lesion. Importantly, the presence of calcifications and cystic spaces distinguishes dermoid cysts from lipomas.[16] The demonstration of a fat-fluid level on MRI or CT is diagnostic for a cervical dermoid cyst.[6] Lymphatic malformation on CT demonstrates poorly circumscribed, multiloculated, hypodense masses with fluid attenuation. MRI typically demonstrates low or intermediate signal intensity on T1-weighted images and hyperintensity on T2-weighted images, indicating proteinaceous material.[16]

Differentiation of these four pathologies is important given that management differs significantly. Brachial cleft anomalies should be surgically excised after the resolution of an acutely infected state.[3] Surgical dissection is made difficult by active infection and can lead to incomplete resection and recurrence. For cervical lymphadenitis, depending on the severity of the symptoms, management may range from serial examination, oral empiric antimicrobial therapy, to needle aspiration and incision/drainage. Due to their tendency to rupture, dermoid cysts require surgical excision as first-line

treatment.[17] Finally, management of infected lymphatic malformation is controversial and includes observation (spontaneous resolution), serial aspiration, sclerosant agents, or surgical resection.[11,18]

CONCLUSION

Painful enlarging unilateral neck masses are a common presentation to the pediatric ED. The differential diagnosis includes infected brachial cleft cysts, suppurative lymphadenitis, dermoid cysts, and infected lymphatic malformations. Some require urgent surgery, others admission for antibiotic therapy and many can be managed electively on an outpatient basis. Differentiation of these four pathologies as detailed above will guide treatment pathways. A thorough history and examination should focus on chronicity, associated local and systemic signs of infection. Imaging using ultrasound, CT or MRI can help distinguish between disease processes, help determine the acuity of the condition and appropriate direct consultations.

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