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Anatomic distribution of cervicofacial lymphatic malformations based on lymph node groups

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ABSTRACT

Objectives: To evaluate radiographic characteristics and to identify locations of cervicofacial lymphatic malformations in children based on known lymph node groupings.
Methods: Retrospective chart review of pediatric patients with cervicofacial lymphatic malformations who underwent imaging with magnetic resonance imaging (MRI), computed tomography (CT) or ultrasonography (US). Ninety charts were reviewed from November 2005 to June 2015. Demographic information and imaging characteristics were evaluated.
Results: Ninety children were included. The average age at presentation was 52 months (range, 1 day to 170 months). Imaging modalities were MRI in 73 (81%), CT in 7 (8%), US in 6 (7%), and multimodality imaging in 4 (4%) cases. Nearly half (49%) of lesions were found in the parotid and submandibular nodal group, 32% in the cervical group, and 19% in the midline face and oral cavity group. The lymphatic malformations were found on the left in 39 (43%) of cases, on the right in 30 (33%) of cases, and were bilateral in 21 (23%) cases. Nineteen (21%) lesions were macrocystic, twenty-two (24%) were microcystic, and forty-nine (49%) had mixed features. Mixed lesions were more likely to be extensive and involve multiple lymph node groups ($P = 0.0005$). Adjacent lymphadenopathy was present in 20 (22%) among all subjects, with an average size of 1.22 (± 1.92) cm in the short-axis.
Conclusion: The results of this study demonstrate three lymph node groupings in which LM are commonly identified. The midline face and oral cavity lesions are predominantly microcystic, the parotid and submandibular lesions are predominately of mixed morphology, and the cervical lesions are predominately macrocystic and mixed. Further studies are needed to determine if such a classification system demonstrates clinically significant difference in disease progression and response to therapy.

1. Introduction

Lymphatic malformations (LM) are rare benign congenital malformations of lymphatic vessels [1]. The incidence of head and neck LM is approximately 1 of every 2000 to 4000 live births [2]. These lesions typically present as soft, colorless masses that generally grow proportionally with the child. Approximately 90% of lesions are diagnosed by five years of age and can be subject to rapid growth with infection or trauma [3]. Symptoms are commonly related to the size of the malformation and encasement of adjacent structures. Nearly 75% of cases of reported lymphatic malformations manifest in the head and neck region due to the relative predominance of the lymphatic system in this area [4].

Previous classification systems have been introduced in an attempt to categorize lymphatic malformations, however, such schemes do not take into account morphologic or regional lymph node groupings [5]. Historic terms such as cystic hygroma and lymphangioma have largely been abandoned and replaced with more descriptive categories of microcystic, macrocystic or mixed cystic types. Just as the terminology to describe lymphatic malformations has evolved, so have different treatment options that include surgical resection, sclerotherapy, and serolimus. Factors that determine treatment modality often depend on the location and extent of disease. However, the lack of a standardized method

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to classify LM creates a challenge in comparing data in the literature. Therefore, further understanding of the anatomic distributions and morphology of these lesions would be beneficial. The aim of the study was to anatomically map out the radiographic center of each lesion and correlate the resulting histogram with known lymph node groupings in our cohort of children with LM over a ten-year period. We hypothesized that LM would be found in high concentrations near known lymph node groupings given the increased density of lymphatic channels in these areas.

2. Methods

Institutional Review Board approval was obtained from the University of Utah. We retrospectively identified 90 patients with cervicofacial lymphatic malformations under the age of 18 years with imaging at a tertiary-referral institution from November 2005 to June 2015. Patients over the age of eighteen years or those without imaging were excluded. Demographic information of patients including age, gender, and imaging characteristics of the lesion was reviewed. High-resolution magnetic resonance imaging (MRI), computed tomography (CT) or ultrasonography (US) were analyzed. The radiographic center of each lesion was then manually plotted onto a standardized left and right pediatric cervicofacial diagram using axial, sagittal and coronal images. The resulting histogram was then evaluated to identify patterns of distribution, particularly whether the radiographic center corresponded to known lymph node groupings. These lymph node regions included facial, lingual, sublingual, mental, submandibular, parotid, lateral cervical, anterior cervical, posterior cervical and occipital regions [6].

Lesions containing identifiable cysts measuring greater than 2 cm in diameter were termed “macrocystic” while those with smaller cysts or without clear boundaries were labeled “microcystic”. The lesion demonstrating both features were designated as mixed according to the ISSVA classification [7]. Enlarged lymph nodes were defined as measuring greater than 1 cm in short axis and only nodes adjacent to the lesion were included. Statistical analysis was performed using the chi-square test of independence for categorical data and the Pearson's correlation for continuous data.

3. Results

The cohort included a total of ninety children. The average age at presentation was 4.3 years (range, 1 day to 14.2 years). The cohort included 35 (39%) females and 55 (61%) males. Imaging modalities for the 90 patients were MRI in 73 (81%), CT in 7 (8%), US in 6 (7%), CT plus US in 3 (3%), and CT plus MRI in one 1 (1%) case. Table 1 summarizes demographic information of the patients based on LM morphology. More than half of the lesions 49 (54%) had mixed features, while 19 (21%) lesions were macrocystic, and 22 (24%) were microcystic. Lesions with mixed morphology were more likely to be extensive and involve multiple lymph node groups ($P = 0.0005$). Table 2 summarizes the location of the lesions based on morphology with a breakdown of gender, age, laterality and imaging modality. LM have distinct features in three different regions of the head and neck according to radiographic appearance and location, namely 1) the midline face and oral cavity 2) the parotid and submandibular region, and 3) the cervical regions. There were no macrocystic lesions found in the midline face and oral cavity groups. The midline face and oral cavity group included facial (10%), sublingual (4%), lingual (2%), mental (1%) lymph nodes. The parotid and submandibular group included submandibular (40%) and parotid (11%) lymph nodes. The cervical group incorporated lateral cervical (14%), occipital (8%), anterior cervical (7%), and posterior triangle (2%) lymph node subsites.

Table 1
Demographic information based on morphologic lymphatic malformation type.

LM morphologic type			
	Microcystic	Macrocystic	Mixed
Age (months)	67.3 ± 57	44.3 ± 55	48.1 ± 43
Gender			
Male	13 (14%)	14 (15%)	28 (31%)
Female	9 (10%)	5 (6%)	21 (23%)
Total patients	22 (24%)	19 (21%)	49 (54%)
Laterality			
Left	9 (41%)	11 (57%)	19 (39%)
Right	7 (32%)	5 (26%)	18 (37%)
Bilateral	6 (27%)	3 (16%)	12 (24%)
Multiple subsites	6 (27%)	4 (21%)	35 (72%)*
Imaging			
MRI	20 (90%)	13 (68%)	41 (84%)
CT	2 (9%)	4 (21%)	5 (10%)
US	–	4 (21%)	5 (10%)

LM = lymphatic malformation.
*p value 0.0005.

There were 42 children (47%) with extensive lesions that involved more than one anatomic subsite, of which 31 (34%) extended into a different region. The LM were found on the left in 39 (43%) of cases, on the right in 30 (33%) of cases, and were bilateral in 21 (23%) cases. Adjacent lymphadenopathy was present in 20 (22%) children with an average 1.22 (±1.92) cm in the short-axis. The histogram (Fig. 1) summarizes the data regarding laterality and morphology.

4. Discussion

This ten-year experience represents one of the largest series to focus on pediatric radiologic findings of cervicofacial lymphatic malformations. LM can be detected by a variety of imaging modalities. On ultrasound, the lesions are typically hypoechoic and Doppler flow is uncommon. CT may reveal uniloculated or multiloculated cystic masses with low density and may be poorly circumscribed (Fig. 2). Fluid levels can be identified, particularly in cases of prior internal hemorrhage or debris from infection. On MRI (Fig. 3), lesions are hypointense on T1WI and hyperintense on T2WI [4]. Often, MRI demonstrates a lesion with no contrast enhancement within the cystic spaces and septations often show mild enhancement. The majority (81%) of children in our study underwent imaging with MRI. A small number of patients had multiple types of imaging which may have been required to confirm the diagnosis or evaluate missing information from initial imaging studies. MRI is integral in preoperative planning as it can determine the extent of the lesion and status of adjacent structures. MRI is superior in delineating anatomic borders and obviates exposure to radiation but may require sedation based on the child's age.

There is no consensus regarding common sites or characteristics of head and neck lymphatic malformations. The most commonly used system applied is that introduced by De Serres et al. [5]. This anatomic staging system is as follows: stage I describes unilateral infrahyoid disease, stage II unilateral suprahyoid disease, stage III unilateral suprahyoid and infrahyoid disease, stage IV bilateral suprahyoid disease, stage V bilateral suprahyoid and infrahyoid disease, and stage VI bilateral infrahyoid disease. A greater stage suggests a higher risk of complications [8]. Although this classification system may be useful in describing gross cervical disease in relation to disease burden and relation to the hyoid bone, important information such as inclusion of the face and morphology is