Surgery of lymphatic malformations in oral and cervicofacial regions in children

Zhi-Min Lei, DDS, MSc, a Xing-Xing Huang, DDS, MSc, b Zhi-Jun Sun, DDS, PhD, c Wen-Feng Zhang, DDS, MSc, d and Yi-Fang Zhao, DDS, MSc, e Wuhan, People’s Republic of China

WUHAN UNIVERSITY

Objective. The objective of this study was to determine prognostic factors and complication rates related to the surgical management of lymphatic malformations of oral and cervicofacial regions in children.

Study design. The charts of 117 children operated on for oral and cervicofacial lymphatic malformations were retrospectively reviewed. Treatment outcomes were analyzed for correlation with several factors, including age at presentation, sex, associated symptoms, anatomical site of involvement, extent of disease, operative complications, histological pattern, and recurrence. The χ² test was used to compare treatment failure rates and complication rates between patients with and without these factors.

Results. The tongue was the most frequent site of involvement (40.17%). The lesions with oral and facial involvement had a higher failure or recurrence rate (29.23%) than those with cervical involvement (8.33%; P < .05), and lesions with involvement of multiple sites had a higher recurrence rate (48.28%) than those with involvement of a single site or 2 sites (11.67%; P < .01). Although patients who underwent surgical procedure at less than 1 year of age had a higher recurrence rate (28.95%) than those more than 1 year (19.61%) of age, and microcystic lesions had a higher recurrence rate (28.33%) than macrocystic lesions (13.79%), no significant difference was found between them (P > .05). The lesions with involvement of 3 or more sites had significantly higher operative complication rates (37.8%) than those with involvement of 1 or 2 sites (15.58%; P < .01).

Conclusion. Factors correlated with a worse prognosis in lymphatic malformations of oral and cervicofacial regions include the involvement of the oral cavity and/or face and involvement of multiple anatomical sites, which also may be associated with higher operative complications. (Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2007;104:338-44)

Lymphatic malformations are developmental defects of the lymphatic system. They are frequently present at birth and are most commonly found in the head and neck regions, but also involve other sites such as the axilla, shoulder, chest wall, mediastinum, abdominal wall, and thigh. Although accepted treatment regimens include sclerotherapy, aspiration, laser, and observation, surgical excision of the lesion is the main treatment modality for lymphatic malformation in the head and neck. Lymphatic malformations involving the oral and cervicofacial regions are especially problematic from a functional and cosmetic point of view. This paper reported our results in 117 children under the age of 17 years with lymphatic malformations of the oral and cervicofacial regions and evaluated prognostic factors and surgical complication rates in their treatment.

PATIENTS AND METHODS

The charts of 117 children operated on between 1985 and 2005 for oral and cervicofacial lymphatic malformations at the Hospital of Stomatology, Wuhan University, China, were retrospectively reviewed. Factors related to treatment outcomes were evaluated, including age at presentation, sex, associated symptoms, anatomical site of involvement, extent of disease, surgical complications, histological pattern, and recurrence.

Soft tissue lesions were excised as completely as possible, and special attention was paid to identify and preserve important nerves and vessels. A tongue reduction procedure was carried out for extensive lesions involving the entire tongue. Surgical complications were noted, including injury to the facial nerve, bleeding or hematoma, infection, and contour defect.
primary anatomic location of disease was defined by the collective information obtained from the clinical examination, imaging studies, and surgical findings. The ultrasound and surgical and histological findings were reviewed, and the lesions were subclassified into macrocystic (greater than 2 cm) and microcystic (less than 2 cm) types according to the diameter of the largest cystic cavity within the lesion.9

Patients without any lesions or residual effects of treatment were considered an excellent outcome. Patients classified as good had a small symptom-free lesion that was not a cosmetic concern. Fair results indicated persistent or recurrent disease that showed improvement but still remained symptomatic. A poor outcome referred to those patients having significant symptoms together with moderate or progressive disease.10

Information on follow-up was obtained by reviewing the medical charts and contacting the parents of the children by telephone or regular mail. Treatment failure was defined as the existence of a recurrent or persistent mass lesion after the first surgical procedure.5 Treatment failure rates and complication rates were compared using the χ² test between patients who presented with these factors and those who did not (SPSS, Version 11.0 for Windows, SPSS Inc., Chicago, IL). P ≤ .05 was statistically considered significant.

RESULTS

Of 117 children, there were 78 males and 49 females (ratio, 1:0.63). The average age at presentation was 42 months (range, 6 months to 17 years). Eighty-three patients (70.94%) presented before 2 years of age. The single most common presenting symptom was a mass, occurring in 95 patients. Associated symptoms included infection (33 patients), eating problems or dysphagia (17 patients), airway obstruction (5 patients), bleeding (3 patients), and malocclusion (3 patients). Two patients required a tracheotomy at the time of diagnosis. The tongue was the most frequent site of involvement; 47 (40.17%) patients had involvement of the tongue region, followed by 19 patients with involvement of the cheek (16.24%), 17 patients with involvement of the anterior neck (14.53%), and 16 patients with involvement of the lips (13.67%). Areas of less involvement were 8 patients with involvement in the submandibular region (6.84%), 5 patients with involvement of the parotid gland (5.13%), and 2 patients with involvement of the oral floor and posterior neck (1.71%), respectively. Involvement of the oral cavity and face was significantly greater than the neck.

All 117 patients included in our series were treated by surgical excision. The average age at surgical procedure was 40.3 months. The lesions with involvement of a single site and macrocystic lymphatic malformations were easily excised and had excellent outcome (Fig. 1). Lymphatic malformations of the neck were either a single cystic mass or multiple noncommunicating cavities with intervening septa of variable thicknesses and were easily dissected from adjacent tissues (Fig. 2). However, care should be exercised during surgical procedure to identify and preserve important cervical structures such as the internal jugular vein, internal or common carotid artery, hypoglossal nerve, and vague nerve. Extensive or diffuse lymphatic malformations, especially lesions with involvement of oral and oropharyngeal mucosa or skeletal anomalies, were frequently poorly defined, difficult to eradicate, and had functional or cosmetic defects after surgical procedure (Fig. 3).

Eighty-nine patients had a follow-up period of 6 months to 11 years, with an average of 3.7 years. Recurrence or persistent disease was found in 21 children (24%) after the initial surgical treatment. Seven had no further treatment, but 14 underwent additional surgery at an average age at surgical removal of 43.6 months and an average follow-up of 5.10 years. Five patients who underwent additional surgical procedure were clinically free of disease. Seventy-three patients (82.02%) were graded as excellent outcome or good, 16 patients (17.98%) as fair, and no patient as poor. It was found that several factors were associated with treatment outcome (Table I). Factors that correlated with a worse prognosis were the presence of a mass at 3 or more anatomical sites and facial or oral involvement. Although microcystic lesions and children operated on below 1 year of age had a higher recurrence rate, no significant difference in recurrence rate was found compared with macrocystic lesions and children operated on who were more than 1 year of age.

Twenty-six complications associated with surgical procedure were demonstrated in 117 patients, and some patients had more than 1 complication. They included damage of the facial nerve (6 patients) and vagus (1 patient), infection (7 patients), hematoma or seroma (4 patients), external deformity (7 patients), and salivary fistula (2 patients). Four patients with paralysis of marginal branches of the facial nerve resolved spontaneously 3 to 6 months postsurgical procedure; another 2 patients had permanent damage. Five infections resolved after a course of oral antibiotics and 2 required intravenous antibiotic therapies. Operative complications were closely associated with the extent of disease, and lesions with involvement of 3 or more sites had a higher complication rate (37.84%) than those with 1 or 2 sites (15.58%; P < .01). However, no significant differences in the complication rate were found between
lesions with involvement of oral and facial regions (25.58%) and those of the neck (12.9%), and between microcystic lesions (26.51%) and macrocystic lesions (11.76%; \( P > 0.05 \)). Patients who underwent surgical removal before 1 year of age and more than 1 year of age had similar operative complication rates, 23.53% and 21.21%, respectively. Recurrence rates or operative complications were not related to sex or associated symptoms.

**DISCUSSION**

Lymphatic malformations are typically present in young children. The present study showed 70.94% occurred before 2 years of age, which is similar to other

---

Fig. 1. Microcystic lymphatic malformations of the tongue. 

**A**, Many lymph-filled vesicles on the dorsum of the tongue. 
**B**, Excision of the lesion. 
**C**, Excellent function of the tongue 5 months after surgical procedure.
Fig. 2. Macrocystic lymphatic malformations of the neck. A, preoperative view. B, Complete resection of the lesion with overlying redundant skin. C, No recurrence 1 year after surgical procedure.
In our patients, the tongue was the most frequent site of involvement, and the anterior and submandibular triangles of the neck were involved more than the posterior triangle. However, Ravch et al. reported the involvement of the neck in the highest percentage of patients, and similar distribution among the posterior, anterior, and submandibular triangles. The difference of sites of involvement is possibly because patients treated by Ravch et al. were from the Department of Otolaryngology at the Hospital for Sick

Fig. 3. Lymphatic malformations of the left facial and submandibular regions. A, Preoperative appearance of the face. B, Panoradiograph showing the involvement of the left mandible. C, Significantly improved facial contour after excision of the left facial and submandibular lesions, but patient has asymmetric deformity in the lower face because of hypertrophy of the mandible.
Table 1. Prognostic factors in 89 patients with lymphatic malformations

<table>
<thead>
<tr>
<th>Factors</th>
<th>Recurrence</th>
<th>Recurrence rate (%)</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age of operation</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>≤1 y</td>
<td>11/38</td>
<td>28.95</td>
<td></td>
</tr>
<tr>
<td>&gt;1 y</td>
<td>10/51</td>
<td>19.61</td>
<td>&gt;.05</td>
</tr>
<tr>
<td>Site of involvement</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Neck</td>
<td>2/24</td>
<td>8.33</td>
<td>&lt;.05</td>
</tr>
<tr>
<td>Oral and face</td>
<td>19/65</td>
<td>29.23</td>
<td>.01</td>
</tr>
<tr>
<td>Extent of disease</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1 or 2 sites</td>
<td>7/60</td>
<td>11.67</td>
<td></td>
</tr>
<tr>
<td>3 or more sites</td>
<td>14/29</td>
<td>48.28</td>
<td>&lt;.01</td>
</tr>
<tr>
<td>Pathological pattern</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Microcystic</td>
<td>17/60</td>
<td>28.33</td>
<td>&gt;.05</td>
</tr>
<tr>
<td>Macrocystic</td>
<td>4/29</td>
<td>13.79</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>21/89</td>
<td>24.00</td>
<td></td>
</tr>
</tbody>
</table>

Recurrence also may be influenced by an imbalance of angiogenic mediators. Sidle et al.\textsuperscript{16} examined specimens from 12 pediatric patients who underwent surgical excision of cervicofacial lymphangioma for expression of angiogenic inducer vascular endothelial growth factor and angiogenic inhibitor pigment epithelium-derived factor by using immunohistochemical analysis. They demonstrated recurrent specimens had increased histological evidence of angiogenesis as well as increased vascular endothelial growth factor and decreased pigment epithelium-derived factor activity compared with nonrecurrent lesions.

The least successfully treated lymphatic malformations are those located in the face or oral cavity. Ravch et al.\textsuperscript{5} reported that 10% of patients with involvement limited to the neck suffered recurrences, compared with 60% when the face or oropharynx was involved. Our results showed the lesions of the neck had a lower recurrence rate than that of lesions of the oral cavity and face. This difference could be explained by the fact that in the face and oral cavity, most lesions are microcystic and infiltrative, anatomical boundaries are less delineated, and structures have cosmetic as well as functional importance. Rowley et al.\textsuperscript{17} found that lymphatic malformations located below the level of the mylohyoid muscle generally may be safely resected within the first 12 months of life and during 1 procedure. Lymphatic malformations above the level of the mylohyoid muscle are frequently poorly defined, the planes between the malformation itself and normal anatomy may be difficult to dissect, and complete surgical resection can be quite a challenge. Operative complications associated with resection of lymphatic malformations in the oral and cervicofacial regions include damage to surrounding structures—particularly nerves and blood vessels—infection, and scarring. Permanent nerve paralysis, which is reported to occur in 6% to 76% of patients,\textsuperscript{1,3,5,12,13} occurred in only 3 cases in our series. The marginal mandibular branch is the nerve most frequently damaged. The cervical sympathetic trunk, the recurrent laryngeal, the hypoglossal, the vagus, and the spinal accessory nerves can occasionally be damaged.

Several authors\textsuperscript{5,12} found no correlation between prognosis and the interval from diagnosis to treatment. Balakrishnan and Bailey\textsuperscript{18} recommend early surgical removal before the lesion has grown and further involved normal tissue and before repeated infections have caused increased fibrosis. Findings of Ravch et al.\textsuperscript{5} indicate that when there is an involvement of sites associated with higher surgical morbidity such as the parotid gland, or sites in which complete removal is less likely such as in the tongue, a watchful observation period should be considered. When surgical removal is
indicated, it should be performed without delay, regardless of age. In light of our results, the time of surgical removal or age of the child was not significantly related to the recurrence and operative complications, and surgical removal can be safely performed after the age of 6 months. Surgical resection of the diffuse and extensive lymphatic malformations involving the oral cavity and face is technically demanding; incomplete removal is common because of a desire to avoid excision of critical structures. Therefore, it is necessary to further explore better therapeutic strategies such as sclerotherapy or laser for managing lymphatic malformations in these regions.

CONCLUSIONS
We reviewed the charts of 117 children with lymphatic malformations of the oral, cervicofacial regions. The overall recurrence rate or failure rate was 24%. Negative prognostic factors were found to include the involvement of the oral cavity and/or face and involvement of multiple anatomical sites. The age at presentation, associated symptoms, and pathological pattern were not significantly associated with the final outcome. The lesions with involvement of 3 or more sites also had higher operative complications. Surgical removal can be safely performed after the age of 6 months. Because incomplete removal of the diffuse and extensive lymphatic malformations involving the oral cavity and face is common, it is mandatory to further explore better therapeutic strategies for managing these lesions.

REFERENCES

Reprint requests:
Yi-Fang Zhao, DDS, MSc
Department of Oral and Maxillofacial Surgery
School and Hospital of Stomatology
Wuhan University
237 Luoyu Road
Wuhan, People’s Republic of China 430079
yifang@public.wh.hb.cn