Total, Subtotal, and Partial Surgical Removal of Cervicofacial Lymphangiomas

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**Objectives:** To compare different surgical interventions for the treatment of extensive cervicofacial lymphangiomas and to define the minimal extent of surgery necessary to control disease.

**Design:** Retrospective study. Mean ± SD follow-up was 31 ± 4 months after surgery. Surgical procedures were grouped as follows: (1) total removal, (2) subtotal removal (all cystic structures removed, small plaques of cyst walls left attached to vital structures), (3) partial removal (major cysts removed, some partially resected cystic structures left in place), and (4) incision and aspiration with subsequent compression bandage. Control of disease was defined as no recurrent or residual tumor or as recurrent or residual tumor less than 10% of initial tumor size without evidence of growth on several postoperative examinations and without clinical symptoms or aesthetic disfigurement.

**Patients:** Twenty-one patients with cervicofacial lymphangiomas (>3 cm in maximum diameter) without thoracic involvement were evaluated. Fifteen patients were 6 years or younger and 6 were older than 6 years. No surgery was yet performed in 3 patients, for a total of 24 surgical interventions in 18 patients.

**Setting:** Hospitalized care in 2 referral centers.

**Results:** After total removal, disease was controlled in 5 of 5 cases; after subtotal removal, in 8 of 9 cases; after partial removal, in 1 of 7 cases; and after incision and aspiration with subsequent compression bandage, in 0 of 3 cases. Two complications were encountered—1 fully reversible paresis of the marginal branch of the facial nerve and 1 secondary healing.

**Conclusions:** Surgical removal of cervicofacial lymphangiomas is a safe treatment modality. Disease control can be achieved if all cystic structures are removed. Small plaques of cyst walls attached to vital structures may be left in place. If small cystic extensions of lymphangiomas are only opened and left in place or if lymphangiomas are only drained following compression bandage, symptomatic residual tumor or recurrence is frequent.


**Lymphangiomas** are relatively rare tumors occurring most frequently in the first decade of life. They originate from lymphatic vessels that are unable to drain their contents into the venous system. Lymphangiomas frequently present as painless soft tissue swelling in the head and neck. Different histologic varieties of this tumor (lymphangioma simplex, cavernous lymphangioma, and cystic lymphangioma) frequently coexist within one mass. The tumor is usually slow growing, may rarely regress spontaneously, and sometimes expands rapidly. Bleeding and infection can rapidly increase the size, threatening the airway. In some reported cases, lymphangioma appears following minimal cervical trauma.

Small lymphangiomas may be asymptomatic, whereas large, often polycystic lymphangiomas with multiple extensions may compromise the airways or interfere with feeding and swallowing. They may lead to deviatory development or pressure-induced lesions of the cervicofacial skeleton, restrict ocular movement, or obstruct the ear canal. In addition, large lymphangiomas are often disfiguring. These extensive lesions demand therapeutic intervention.
PATIENTS AND METHODS

All patients with lymphangiomas of the head and neck treated between 1993 and 1997 in the Departments of Otorhinolaryngology, Universities of Ulm, Germany, and Tîrgu-Mureș, Romania, were evaluated. To assess preoperative symptoms, tumor size, and details of the surgical procedure, the medical records were retrospectively reviewed and findings from magnetic resonance imaging (MRI), computed tomography (CT), and ultrasound examinations were analyzed. Patients with tumors less than 3 cm in largest diameter were excluded. Indications for surgery; size, growth pattern, and localization of tumor; mode of surgery; and clinical outcome were categorized.

Symptoms indicating surgical intervention were grouped as skeletal deviation or bony erosion, dyspnea, dysphagia, aural or ocular involvement, and aesthetic disfigurement without functional impairment. Tumor size and growth pattern were classified according to the largest diameter of the lesion in horizontal and vertical planes as assessed by preoperative MRI scans, CT scans, and/or ultrasound examinations, and histological specimens obtained during surgery: size I, 3 to 5 cm largest diameter; size II, greater than 5 cm in largest diameter without invasive growth pattern; and size III, greater than 5 cm with invasive growth pattern. Affected regions were categorized as neck, tongue and oral cavity, parotid and buccal area, other regions, and several regions involved concomitantly. The classifications of indications for therapeutic intervention, tumor size, growth pattern, and affected regions are summarized in Table 1.

Completeness of surgical removal was categorized as total removal, subtotal removal, partial removal, and incision and aspiration. The classification of completeness of surgical removal is outlined in Figure 1 and Table 2.

All patients underwent a follow-up examination including ultrasound examinations of the neck, tongue, and parotid region. If parts of the lesion had been hidden behind structures impermeable for ultrasound preoperatively, postoperative MRI or CT was performed (8 cases). Computed tomography was only performed in adult patients to avoid radiation exposure of young patients. Mean ± SD follow-up was 31 ± 4 months. The presence of residual or recurrent tumor was categorized as follows: no tumor, no tumor detectable at follow-up examination; controlled, recurrent or residual tumor less than 10% of initial size and without any clinical symptoms or aesthetic disfigurement and without evidence of growth on several postoperative examinations; and recurrence, recurrent or residual tumor greater than 10% of initial size, with clinical symptoms or aesthetic disfigurement, or evidence of growth.

Various therapeutic modalities have been proposed for treatment of lymphangiomas of the head and neck, including repeated aspiration and percutaneous embolization with various sclerosing agents such as piciba-nil (OK-432), fibrin glue, or corticosteroids. Also, intravenous cyclophosphamide has been used for treatment of supralaryngial lymphangioma. These modalities have been found to be ineffective and associated with severe complications by some authors, who consider total surgical removal of the lesion to be the treatment of choice. However, a more conservative surgical approach is advised by others. Most authors agree that surgery should never be excessive and vital structures must never be sacrificed.

Because the latter statements concerning the extent of surgery seemed imprecise, we attempted to grade the completeness of surgical removal of exten-
sive cervicofacial lymphangiomas based on the experience of 21 cases. The clinical outcome in relation to the completeness of surgical removal of these lesions was evaluated.

### RESULTS

Twenty-one patients with extensive cervicofacial lymphangiomas were evaluated. Six patients were younger than 1 year, 9 patients were between 1 and 6 years old, and 6 patients were older than 6 years. Twelve patients were male and 9 patients were female. Eleven patients were treated at the Department of Otorhinolaryngology, University of Tirgu-Mures, and 10 patients were treated at the Department of Otorhinolaryngology, University of Ulm.

Indications for surgery were dysphagia in 2 patients, dyspnea in 1, involvement of the orbit with impairment of ocular movement or obstruction of external ear canal in 3, false posture of the head in 3, and bony erosion in 2. In these patients, a varying degree of disfigurement was also present. Disfigurement alone without functional impairment was the indication for surgery in 10 patients.

Only tumors greater than 3 cm in largest diameter were included. In 9 patients, the tumor was 3 to 5 cm in diameter (size I), in 10 patients it was greater than 5 cm without infiltrating growth pattern (size II), and in 2 patients it was greater than 5 cm with infiltrating growth pattern (size III).

Various regions of the head and neck were affected by the malformations. The buccal and parotid region was the origin of the malformation in 5 patients, the neck in 7, the mouth and tongue in 1, other regions (forehead, occiput, and temporal region including the eyelid) in 3, and several regions were simultaneously involved in 5.

Of the 21 patients observed, no surgery was yet performed in 3. In 1 patient with lymphangioma of the tongue and mouth, surgery was postponed because of cardiac disease; the child is tracheotomized and nourished by percutaneous gastrostomy. In the 2 remaining patients without surgery, functional impairment is minimal although severe disfigurement exists. The parents did not yet consent to surgery in these cases. In 6 patients, 2 procedures have been performed, totaling 24 surgical procedures in 18 patients.

Size and location of tumor and mode of surgical procedure as outlined above were considered relevant confounders. A confirmatory analysis could not be applied because of the small number of cases. Thus, the results of the various types of surgical intervention in relation to the factors considered relevant for prognosis are descriptively presented in **Table 3**.

Total removal of the tumor was the most reliable procedure. No persistent or recurrent disease was observed in this group. Total removal was feasible in 4 of 10 interventions in size I tumors, in 1 of 11 interventions in size II tumors, and in 0 of 3 interventions in size III tumors. Subtotal removal as described above also was a reliable surgical modality. In 5 of 9 cases no tumor was found at the last postoperative examination and disease was controlled in 3 of 9 cases. A recurrence was found after 1 of 9 interventions.

Partial removal yielded unreliable results in the observed patients. Disease recurred in 6 of 7 cases; in 1 of 7 cases, no tumor was found. Incision and aspiration with subsequent compression resulted in recurrence in 3 of 3 cases. The 3 patients had to undergo revision surgery.

Two complications were observed. In 1 patient with size III tumor, a reversible paresis of the marginal branch of the facial nerve was observed and in another patient a wound healing impairment had to be treated surgically.

### COMMENT

Lymphangiomas of the head and neck are rare malformations of the cervicofacial lymphatic system. The clinical significance of this entity mainly depends on the size of the lesion. The completeness of surgical removal depends on the size of the lesion; surgery should be performed as soon as possible.

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**Table 3. Results of Various Surgical Interventions**

<table>
<thead>
<tr>
<th>Tumor Size</th>
<th>Localization</th>
<th>Total Resection</th>
<th>Subtotal Resection</th>
<th>Partial Resection</th>
<th>Incision/Aspiration</th>
<th>No Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>I (10 interventions in 9 patients)</td>
<td>Neck</td>
<td>2, no tumor</td>
<td>1, no tumor; 1, controlled</td>
<td>1, recurrence</td>
<td>2, recurrence</td>
<td>1, no result</td>
</tr>
<tr>
<td></td>
<td>Buccal/parotid area</td>
<td>1, no tumor</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td></td>
<td>Other regions</td>
<td>1, no tumor</td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td></td>
<td>Neck</td>
<td></td>
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<tr>
<td></td>
<td>Buccal/parotid area</td>
<td></td>
<td>1, controlled</td>
<td></td>
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<tr>
<td></td>
<td>Oral cavity/tongue</td>
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<td></td>
<td>Other regions</td>
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<tr>
<td>II (11 interventions in 10 patients)</td>
<td>Neck</td>
<td></td>
<td></td>
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<tr>
<td></td>
<td>Buccal/parotid area</td>
<td></td>
<td>1, controlled</td>
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<td></td>
<td>Oral cavity/tongue</td>
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<td></td>
<td>Other regions</td>
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<td></td>
</tr>
<tr>
<td></td>
<td>Several regions</td>
<td>1, no tumor</td>
<td></td>
<td>1, recurrence</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Several regions</td>
<td>1, controlled;</td>
<td>1, recurrence</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Total interventions</td>
<td>5/7, no tumor</td>
<td>5/9, no tumor; 3/9, controlled; 1/9, recurrence</td>
<td></td>
<td>1/7, no tumor;</td>
<td>6/7, recurrence</td>
</tr>
<tr>
<td>III (3 interventions in 2 patients)</td>
<td>Total interventions</td>
<td>5/5, no tumor</td>
<td></td>
<td></td>
<td>3/3, recurrence</td>
<td>3/3, no result</td>
</tr>
</tbody>
</table>

* Ellipses indicate data not applicable.
and localization of the tumor. In small lymphangiomas without functional impairment or aesthetic disfigurement, therapeutic intervention is not necessary. The tumor may rarely regress spontaneously, sometimes it can expand and become symptomatic. Large tumors may interfere with glutation, respiration, vision, hearing, regular posture of the head and neck, bone development, or aesthetic appearance. In these cases, therapeutic intervention is mandatory.

VARIOUS MODALITIES have been described to treat lymphangiomas of the head and neck and several authors consider surgical removal the method of choice, because there are few complications and revision surgery is not rendered unduly difficult when compared with revision following obliterative techniques. However, statements regarding the completeness of surgical removal necessary to achieve the best benefit for the patient are vague. Most authors agree that vital structures should not be sacrificed to achieve total tumor removal. It remains unclear how much tumor should be removed and how much can be left in place.

To estimate the extent of surgery necessary to obtain a satisfactory result, we attempted to categorize the completeness of tumor removal in a group of 21 patients with extensive cervicofacial lymphangiomas. The surgical interventions were grouped into 4 categories: total removal, subtotal removal, partial removal, and incision and aspiration. For total removal, no remnants of tumor were left in place. For subtotal removal, all cystic structures were removed, and only flat, plaquelike small remnants were left in place when firmly attached to nerves or vessels. In the partial removal intervention, not all cystic structures were removed, but some cysts were only in part resected and saclike remnants were left in place. In the incision and aspiration with subsequent compression treatment, the obvious cystic structures either were punctured using a syringe for fluid aspiration or were incised, spreading scissor blades several times within the lesion, and finally applying a compression bandage with or without a wound drain. These basic surgical concepts are illustrated in Figure 1.

The criteria for success used in this evaluation are rather stringent when compared with criteria used by others. The clinical outcome was categorized in 3 groups, ie, no residual or recurrent tumor (no tumor), residual or recurrent tumor less than 10% of the initial size without any functional or aesthetic deficits (controlled), and tumors greater than 10% of the initial size and/or tumors causing functional or aesthetic deficits (recurrence). In contrast to malignant tumors, radical eradication of the disease is not the aim of surgery, but that functional and aesthetic integrity is achieved. Thus the first 2 categories are considered good outcomes, whereas the recurrence category is considered a therapeutic failure.

To compare the outcome of different surgical techniques, the severity and extent of disease in the various treatments must be approximately equally distributed. Three indicators for severity and extent of disease were considered relevant: symptoms indicating surgical intervention, size and growth pattern of the lesion, and regions affected. These factors have also been demonstrated to influence clinical outcome by Ricciardelli and Richardson. The existing classifications of Landing and Farber and Bill and Summer did not describe the relevant confounders sufficiently and the McGill and Mulliken classification was not detailed enough to describe the factors considered relevant in the observed patients. The classification used is not complete; for instance, bleeding, a common problem in lymphangiomas of the tongue and mouth, did not occur in these patients and was not included. The classification merely intends to achieve rather equal distribution of possible confounders over the various treatment arms in this evaluation.

Cervicofacial lymphangiomas are often diagnosed in second-trimester fetuses using ultrasonography. They are classified into septated and nonseptated lesions. In fetuses with septated lymphangiomas, abnormal karyotypes prevail in more than 80% and these fetuses rarely survive. Most patients in this report had polycystic lymphangiomas with a septated appearance in ultrasound examinations. No chromosomal abnormalities were known in this group.

Total tumor removal proved successful in 5 of 5 cases, subtotal tumor removal in 8 of 9 cases, partial removal in 1 of 7 cases, and incision and aspiration in 0 of 3 cases. Total removal is the method of choice, if it can be achieved. It was most frequently performed in smaller tumors not involving the oral cavity and could not be achieved in tumors larger than 5 cm with invasive growth pattern. When the oral cavity and tongue are involved, total tumor removal may result in severe functional impairment.

Subtotal removal is defensible in cases where total removal results in functional deficits due to tumor size, growth pattern, or localization. If the lymphangioma has an infiltrative growth pattern, resection of organs with total parotidectomy, submandibulectomy, and selective neck dissection may be necessary (Figure 2). Aggressive surgical procedures such as wide resection of cervicofacial skin and temporary midline mandibulotomy to obtain total tumor removal were not performed and subtotal removal was considered sufficient in cases with extensive tumor.

Partial removal of lymphangiomas proved unreliable and is considered adequate only for lymphangiomas involving the tongue, pharynx, or larynx. In these cases, reduction of tumor with the carbon dioxide or Nd-YAG laser might alleviate the symptoms and sclerotherapy using picibanil (OK-432) may be an effective treatment. Precise preoperative evaluation of tumor size and extent that includes MRT is necessary to avoid unintentional partial removal of lymphangiomas. Because lymphangiomas can exhibit a growth pattern resembling infiltrative tumors, extensive surgery with exposure of various vital structures of the head and neck and removal of organs such as salivary glands or thyroid must be expected and lie within the capabilities of the surgeon. Meeting these requirements, partial removal of lymphangiomas should be avoided in favor of subtotal removal in most cases. Revision surgery of
lymphangiomas may be rather exacting and should be prevented if possible. Incision and aspiration is also not a valid therapeutic concept. It can be performed to relieve acute symptoms or to facilitate securing the airways in emergencies.

Although extensive lymphangiomas were treated in the patients observed, the complication rate was low. In 1 patient, a completely reversible paresis of the marginal branch of the facial nerve was observed and another patient required wound revision for an infection.

Cervicofacial lymphangiomas are rare. It is thus problematic to gather sufficient numbers of cases in a prospective study to obtain good-quality data for confirmatory statistical evaluation of different therapeutic concepts. In addition, surgical procedures cannot be blinded and potential bias owing to the judgment of the examiner exists. These are also severe drawbacks of the present evaluation. In this study, preoperative data were based on retrospective chart reviews, and the operative notes were used to determine the amount of remaining lesions. Moreover, the assessment of tumor size based on preoperative MRI, CT, and ultrasound examinations may not always reflect the actual size of the lesion. However, all patients were carefully examined at their initial admission, including CT or MRI, except in 4 patients with size I lesions. In these patients, the lymphangioma could be visualized entirely using ultrasonography and no CT or MRI scan had been obtained preoperatively. The documentation of patient data meets contemporary quality standards. All patients were reexamined at the hospital using a standardized examination plan to obtain follow-up data of good quality. At follow-up, an ultrasound examination was performed in all patients. If parts of the lesion had not been visible using ultrasonography preoperatively, postoperative CT in adults and postoperative MRI in children had been performed. The mean follow-up of 31 months is considered sufficient to assess the short-term outcome. Taking the drawbacks of the study design and the limited number of cases into consideration, the data obtained are suggestive that total or subtotal removal of extensive cervicofacial lymphangiomas is a reliable and safe surgical concept. Partial removal leaving sac-like remnants in place may frequently result in symptomatic recurrences. Reformation of lymphangiectatic cysts with impaired venous drainage is the probable reason for failure of this treatment modality. Incision and aspiration with subsequent compression of cervicofacial lymphangiomas may help establish the airways in emergencies, but is not suitable for definite treatment.

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Figure 2. Three-year-old girl with cervicofacial lymphangioma. A, T-weighted coronal magnetic resonance image with gadolinium contrast. B, Intraoperative site (circle indicates mastoid; hollow arrow, greater auricular nerve; triangle, trunk of facial nerve; and solid arrow, lymphangiectatic cysts dispersed in the parotid tissue). C, Histological overview (white arrow indicates ductal epithelium; black arrow, lymphangiomatosus cavities between the acini) (immunohistochemical stain against cytokeratin, magnification ×80); D, Same girl 2 years after surgery.
REFERENCES