25 Years’ Experience With Lymphangiomas in Children
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Background/Purpose: The management of lymphangioma in children is challenging because complete resection is difficult to achieve in some cases, and recurrences are common. The authors reviewed their experience to assess the risk factors for recurrence and the role of nonoperative treatment.

Methods: A retrospective study over a period of 25 years was carried out. One hundred eighty-six patients with 191 lesions (five patients with de novo lesions in different sites) were treated. There were 90 boys and 90 girls. The average age at diagnosis was 3.3 years (range, fetal life to 17 years) and the average size 8 cm in diameter. Histocytological confirmation was obtained in all patients. The involved sites were head and neck, 89 patients (48%); trunk and extremities, 78 patients (42%); internal or visceral locations (eg, abdominal and thoracic), 19 patients (10%). The treatment consisted of macroscopically complete excision in 145 patients (150 lesions, of which five were recurrences in different sites), partial excision in 10 patients, aspiration in five patients, laser excision in 10 patients, biopsy only in four patients, drainage and biopsy in two patients, and injection of sclerosing agents in 10 patients.

Results: There were 54 recurrences: 44 underwent excision (five of them more than once), and five regressed spontaneously on follow-up. Five other recurrences were stable and not progressing. Recurrences, (defined as clinically obvious disease), were found to be 100% after aspiration, 100% after injection, 40% after incomplete excision, 40% after laser excision, and 17% after macroscopically complete excision. The recurrence rate in the last group was the highest in the head (33%), the least in the internal locations (0%), and intermediate for the cervicothoracic location (13%). There were no significant differences, in terms of outcome, between those who had their surgery immediately at the time of diagnosis (n = 101) and those who had delayed surgery (n = 85).

Conclusions: There were fewer recurrences after macroscopically complete excision. Aspiration and injection had the highest recurrence rate. Risk factors for recurrence included location, size, and complexity of lesions. A period of observation may be useful for infants to facilitate complete excision.

INDEX WORDS: Lymphangioma, mesenteric cyst, cystic hygroma.

LYMPHANGIOMAS are a heterogeneous group of benign vascular malformations of the lymphatic system composed of cystically dilated lymphatics. They commonly present as a mass particularly in the head and neck areas. Because of their size and location, they are sometimes a real therapeutic challenge.1-4

We retrospectively analyzed our experience at The Montreal Children’s Hospital during the past 25 years to assess the risk factors for recurrence and the benefit of nonoperative treatment of childhood lymphangiomas.

MATERIALS AND METHODS

A search was made through our hospital records using lymphangioma and cystic hygroma as a diagnostic code. Our previous and current systems allow coding of diagnosis only if the patient was admitted or underwent an outpatient procedure under general anesthesia.

Our search identified 186 patients with lymphangiomas treated at The Montreal Children’s Hospital over the last 25 years. Their clinical records, imaging, surgical notes, and pathological reports were reviewed. We considered the surgical excision incomplete if it was mentioned as such in the surgical notes or in the pathological reports. Follow-up information about recurrences and complications were taken from the clinical records and from the treating surgeons.

RESULTS

We reviewed 186 patients with 191 lesions. The age at presentation ranged from 26 weeks’ gestation in a fetus to 17 years old, with an average of 3.3 years. Ninety-eight were boys and 88 were girls. Fifty-five lymphangiomas were located in the neck, 34 in the head, 78 in the trunk and extremities, and 19 in the internal organs (Fig 1 and Table 1). The average size was 8 cm. One hundred forty-four patients were referred to us because of the presence of a mass, 13 patients because of abdominal pain, and four patients because of an inguinal swelling. Lymphangioma was found incidentally on x-ray (lytic bone lesions) in two patients and in one patient as a splenic cyst on ultrasound scan. Histocytological confirmation was.
obtained in all patients. Only three had combined hemolymphangiomas.

The diagnosis was based on clinical examination alone in 41% of the cases. The most frequently used diagnostic tools were ultrasonography (US) in 42 patients, computed tomography (CT scan) in 25 patients, and both US and CT scan in 12 patients.

Eighty-five patients (45%) were observed for a period of time (average, 5 years) before undergoing an initial procedure. One hundred forty-four patients underwent complete excision as the initial procedure, 10 patients underwent partial excision, 11 patients had aspiration or biopsy with drainage, and 10 patients underwent laser excision. Injection with steroids, tetracycline, or 50% dextrose was unsuccessful in 10 patients, requiring subsequent surgical excision (Table 2). Only one patient regressed spontaneously without intervention. Emergency interventions were necessary in four newborns because of respiratory compromise resulting from rapid growth of the lesion, of which, two had complete excision, one underwent partial excision, and one had aspiration only. One patient had spontaneous bleeding into a large axillary lesion at 5 weeks of age during observation. She required transfusion and underwent resection a few weeks later. Episodes of infection or cellulitis treated with antibiotics also were noted in a few patients. These infections did not appear to induce regression, nor were they associated with significant complications in our review.

The average follow-up was 3 years (range, 5 months to 16 years); 95% underwent follow-up for at least 12 months. There were 54 of 186 (29%) recurrences, defined as persistence or reappearance of clinically obvious disease. Sixty percent of recurrences occurred within 1 year and 80% within 3 years. Five of the 54 recurrent lesions regressed spontaneously on follow-up (3 of 25 after recurrence of a lesion that was completely excised initially); 45 of 54 (75%) underwent excision, five of which required reexcision; three were managed by laser excision; and one by aspiration on three occasions. The recurrence rate was 100% in the aspiration and injection groups, 40% in the laser and incomplete excision groups, and 17% after macroscopically complete excision (Table 3). The recurrence rate in the complete excision group was 33% in those with head lesions and 0% in those with internally located lesions (Table 4). Metachronous lesions (in different sites) were found in five patients; all underwent excision.

The postoperative complication rate was 18%, distributed equally among expectant and nonexpectant group.

Table 2. Initial Intervention in 186 Patients

<table>
<thead>
<tr>
<th>Procedure</th>
<th>No. of Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Complete excision</td>
<td>144 (77%)</td>
</tr>
<tr>
<td>Partial excision</td>
<td>10 (5.3)</td>
</tr>
<tr>
<td>Laser excision</td>
<td>10 (5.3)</td>
</tr>
<tr>
<td>Aspiration with or without biopsy</td>
<td>11 (6)</td>
</tr>
<tr>
<td>Injection with Steroids/tetracycline or 50% dextrose</td>
<td>10 (5.3)</td>
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</tbody>
</table>
Table 3. Recurrence Versus Procedure

<table>
<thead>
<tr>
<th>Procedure</th>
<th>No of Patients</th>
<th>No. of Recurrences</th>
<th>% Recurrence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Complete excision</td>
<td>144</td>
<td>25</td>
<td>17</td>
</tr>
<tr>
<td>Partial excision</td>
<td>10</td>
<td>4</td>
<td>40</td>
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<tr>
<td>Laser excision</td>
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<tr>
<td>Injections</td>
<td>10</td>
<td>10</td>
<td>100</td>
</tr>
<tr>
<td>Aspiration with or without biopsy</td>
<td>11</td>
<td>11</td>
<td>100</td>
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This included seroma in eight patients, wound infection in six patients, hematoma in five patients, pain at the site of the excision in two patients, recurrent laryngeal nerve injury in one patient, and facial nerve injury in one patient (Table 5). There were no significant complications in reoperations for recurrence. Two patients in our study died, both as a result of their disease. One was a teenage patient with abdominal lymphangiomatosis involving bowel, mesentery, and spleen, who had recurrent infections of the multiple cysts. The other was born prematurely from nonimmune hydrops fetalis with bilateral chylothorax, pleural lymphangiectasia, lymphangiomaticous changes in the left lower lobe, and lymphangiomas involving the neck and scalp; she suffered from pulmonary hypoplasia, and supportive treatment was withdrawn at 5 weeks of age.

DISCUSSION

Overall, lymphangiomas account for about 6% of the benign tumors in the pediatric population. Both sexes are affected equally. Sixty percent are present at birth, and 90% are detected by the end of the second year. They neither become malignant nor have a familial tendency. Nearly 50% of lymphangiomas in our study occurred in the head and neck, 42% in the trunk and extremities, and 10% in the internal organs. The most common single site was the neck with 30% of the lesions. Most lymphangiomas present with a mass or diffuse swelling, which initially grows slowly with the child and after some time might slowly regress. Some investigators make a distinction between cavernous lymphangiomas, which can occur anywhere in the body, and cystic hygromas, which occur in the areas of the embryonic lymph sacs ie, neck, axillae and groins. However, considerable overlap exists and we, like others, have not tried to separate the two entities in our review. The distinction may be interesting embryologically but bears little relevance in the treatment of patients.

Recurrent upper respiratory tract infections or accidental trauma commonly enlarge the mass or worsen the swelling. Such complications were severe enough to require intervention in only five patients (2.6%). Macroglossia secondary to lymphangiomas of the tongue can cause airway obstruction, swallowing difficulty, malocclusion, and speech problems.

History and physical examination usually suggest the diagnosis of lymphangiomas (41% in our study). At times it is necessary to confirm the diagnosis or differentiate the lesion from lipoma, dermoid cyst, branchial cyst, thyroglossal duct cyst, teratoma, salivary gland or thyroid tumors, neurofibroma, and meningoencephalocele. In these instances, radiological evaluation may be required. Soft tissue radiographs are useful to evaluate the airway. Both CT scan and US are helpful in demonstrating the solid or cystic nature of the lesion. The differentiation of lymphangioma from other fluid-filled masses may require needle aspiration or biopsy. A recent report suggests that magnetic resonance imaging is the most useful modality to assess extension.

Surgical excision of localized lymphangiomas is the treatment of choice, but surgeons often worry about the infiltrative nature of some lesions and the difficulty in achieving complete resection. It was possible to perform complete excision in 77% of our patients with a low recurrence rate (17%) and minimal morbidity. Observation for a period of time is warranted for infiltrating lymphangiomas and lymphangiomas causing macroglossia with absence of obstructive symptoms. Lymphangiomas that are localized or that present a diagnostic difficulty can be excised earlier, as is often the case for submandibular lesions. In our study, 45% of patients did not have immediate treatment and remained without significant complications during the expectant period, except for one patient who required transfusion. Some investigators recommend immediate excision because of the danger of severe complications. Others prefer to wait until the child is 2 to 6 months of age or older for the large cervical hygroma. We feel that expectant management is feasible, safe, and advisable in some cases to achieve a
safer complete resection. The disadvantages of this approach include complications (eg, hemorrhage and respiratory distress), frequent follow-up, and the need for a biopsy, in some cases, to rule out malignant or premalignant conditions. We treated one patient (not included in this series) who had a large neck cyst in whom the cytology on the aspirated fluid was suspicious, and the diagnosis of cystic teratoma was confirmed at the time of excision. Expectant management is also advisable for recurrent lesions, at least initially. We noted a spontaneous regression in 12% of patients who had recurrence after initial complete excision (3 of 25).

Some surgeons have recommended conservative management with observation only in asymptomatic patients. The rationale for this management is based on a few documented cases of spontaneous regression. From our review of hospital charts, spontaneous regression was rarely seen in our patients: 1 of 186 patients before any intervention, 3 of 25 who had recurrence after an initial complete excision, and 2 of 29 who had recurrence after another type of procedure. However, because our medical records department can search for a diagnostic code only if a patient has been admitted or operated on, there may have been patients treated conservatively as outpatients whom we could not identify in our study. These cases might have regressed spontaneously.

Aspiration is not a definitive treatment but may be useful for emergency decompression. Over recent years, intraläsional injection of bleomycin, ethibloc, and, more commonly, OK-432 (lyophilized incubation mixture of group A streptococcus pyogenes of human origin) have been used as an alternative treatment with promising results. It has been suggested that OK-432 can be used alone as a primary therapy or after partial surgical excision, or in recurrent lymphangiomas. We do not have experience with these agents. Tetracycline, steroids, and 50% Dextrose had been tried in our study with poor results.

Argon beam ablation and laser technology have been reported to be effective in the treatment of lymphangiommas, especially complex types. Our patient who died in 1986 of diffuse abdominal lymphangiomas might have benefited from the argon beam coagulator if it had been available. Ten patients underwent laser excisions in our study with 40% recurrence, but these often were tongue lesions that were partially resected for debulking.

The type of procedures and the site of the lesions were found to be important factors in the recurrence rate. Neck lesions had a fairly low recurrence rate after complete excision (12%), and internally located lesions had the least recurrence.

In the 186 patients included in this study, there were fewer recurrences in completely excised lymphangiomas with very few significant complications. Aspiration, injections, and laser excision treatments have high recurrence rates. We found no significant differences in outcome between those who had their surgery immediately at the time of diagnosis (101 patients, 55%) and those who had delayed surgery (85 patients, 45%). We conclude that delaying surgery may be useful in young children with large lymphangiomas when nerves and other important structures are at risk. Patience and meticulous dissection are required in those cases to achieve a complete resection. OK-432 and ethibloc are promising alternatives, but their long-term effects remain unknown. Until careful prospective studies are available we feel that these agents should be reserved for patients in whom complete excision does not appear feasible or who suffer from a symptomatic recurrence that would not be easily excised.

ADDENDUM

The hospital office charts of Dr H.B. Williams (plastic surgeon and Surgeon-in-Chief) yielded four more patients with cervical lymphangiomas that regressed spontaneously. The lesions were noted at birth or in the first month of life and all showed regression in the first year. The lesions have regressed completely in all four patients, with follow-up of 2 to 8 years. Although spontaneous regression remains infrequent, these cases support the usefulness of a period of observation.

REFERENCES

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