VARIED PRESENTATION AND MANAGEMENT OF LYMPHANGIOMAS IN INFANCY AND CHILDHOOD-
OUR EXPERIENCE IN DISTRICT LEVEL TEACHING HOSPITAL

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ABSTRACT

Lymphangiomas are developmental defects of the lymphatic channels that belong to a large spectrum of vascular malformations usually located in the head and neck region. They present variably and occur anywhere there are lymphatic vessels. These are seen in infants and children primarily. Retroperitoneal lymphangiomas pose diagnostic challenge as well as its surgical management.

OBJECTIVE

Our study focused on the location, clinical presentation, and management of lymphangiomas in infancy and childhood.

METHODS

The medical records of children with lymphangiomas admitted to MGM Hospital, Warangal, Telangana State over a period of 7 years from April 2009 to March 2016 were reviewed for age with gender, location, mode of presentation, management, and its outcome.

RESULTS

We managed 50 children with lymphangioma in last 7 years at MGM Hospitals, Warangal. The age range from 9 days to 10 years. About 65% of children were in the age group of less than 5 years with female-to-male ratio of 2:1. The youngest was 9 days and the eldest was 10 years. In 20 patients (40%), lymphangioma was present in the neck, 18% in axilla (9 patients), 10% in over the chest wall/abdominal wall, 10% over the back, 10% over the limbs (5 patients each) respectively, 8% over the perineal region (4 patients), 2% in retroperitoneum (1 patient), and 2% in the floor of the mouth (1 patient). Surgical excision was the treatment of choice and performed in all cases.

CONCLUSIONS

Lymphangiomas are rare congenital malformations seen in infancy and childhood. They are usually asymptomatic at the time of clinical presentation. They are diagnosed by ultrasonography. CT and MRI scan are performed in doubtful cases. Total or subtotal excision was the treatment of choice by preserving the surrounding important structures.

KEYWORDS

Congenital Malformation, Cystic Hygroma, Lymphangioma, Total or Subtotal Excision.


INTRODUCTION

Lymphatic malformations or lymphangiomas are rare benign hamartomas that result from maldevelopment of primitive lymph sacs.¹ Approximately 50% are present at the time of birth and 90% are diagnosed before the age of 4 years. They are most frequently found in the neck (75%) and axilla (15%) while 10% found in mediastium and abdominal cavity including mesentery, retroperitoneum.²³ The incidence range from 1.5 to 2.8 per 1,000 livebirths.⁴

There are three theories of origin for this malformation. 1) A blockage or arrest in the normal growth of primitive lymph channels during embryogenesis, 2) The primitive lymph sacs do not reach the venous system, 3) During embryogenesis, lymphatic tissue is laid in the wrong area.⁵ Surgical management remains the best possible treatment still today although alternative treatment such as sclerotherapy have been proposed to reduce the impact and complications of surgery.⁶⁷ The present study focuses on the varied clinical presentation and location of lymphangiomas, their management, and further followup in infancy and childhood.

MATERIALS AND METHODS

A retrospective analysis of 50 cases admitted in MGM Hospitals, Warangal, during the period of 7 years from April 2009 to March 2016 were undertaken to review the clinical presentation, location, and management of lymphangiomas. All patients were within the age group of 9 days to 10 years with maximum (30 patients) less than 5 years old. Both the sexes were included in our study. Female-to-male ratio was 2:1 (Table 1). Site of lymphangiomas were shown in Table No-2 and Fig No: 1a, b, c, d, e. 20 patients presented with swelling over the neck, 9 patients in the axilla, 5 patients each over the chest/abdominal wall, back, and extremities, respectively. 4 patients in the perineal region, one patient within the retroperitoneum, and one patient within the floor of the mouth. All patients were subjected to radiological
investigations like ultrasonography, colour Doppler study, CT scan was done in 10 patients especially in neck cases and a case of cystic lump in abdomen and MRI scan was performed in 5 doubtful cases especially over chest, abdominal wall, and back swellings to confirm lymphangiomas.

Ultrasonography with high resolution probe showed multilocular cystic lesions (macrocystic variety) in 35 patients of our series (Fig. 2a). They showed multiloculated hypoechoic lesions of varied sizes filled with clear fluid. Another 4 patients were also subjected to CT scan for better delineation of extent and type of lymphangioma (Fig. 2b). Colour Doppler study was performed in 10 patients, which helped us to determine the relation towards major vessels especially in neck and axilla.

In one patient, we noticed a huge retroperitoneal cyst on abdominal ultrasonography, which was subjected to CT scan and confirmed as huge cystic cavernous lymphangioma (Fig. 4a). MRI scan was performed in 5 patients who presented with mass on back and perineal region. All patients were subjected to surgical excision of the cysts. However, total excision was possible in macrocystic lymphangiomatosis, subtotal excision was performed in 60%-70% of patients to preserve the neurovascular bundles in the neck and axilla.

RESULTS
All the 50 patients were subjected to surgical excision with incorporation of drain in the form of corrugated rubber drain or vacuum suction drainage. In total, 35 patients, total excision of the lymphangiomatosis was performed especially in macrocystic type located on the neck and axilla (Fig. 2c, d and 3a, b respectively). In one of the case, submandibular gland was adherent to the mass and it was removed along with the mass. In few cases in the neck, the cysts were seen extending behind the carotid sheath and as well in some extending into the supraclavicular fossa. Here, we had to take care during dissection and some of the cysts were marsupialised to prevent damage to the carotids as well as subclavian vessels.

In 10 patients, subtotal excision was performed who were diagnosed to have cavernous lymphangiomas to preserve the neurovascular bundles surrounding them. In one patient with huge retroperitoneal cyst, it was arising from the pelvis and occupying the whole sigmoid mesocolon and extending into the abdomen getting blood supply from inferior mesenteric artery, hence sigmoid colon resection was inevitable. There were two to three large cysts with few small cysts near the retroperitoneum (Fig. 4b, c). In 5 patients, we observed the lymphangiomas over the extremities, especially in one patient on the left forearm and subjected to total excision, which turned out as microcystic variety (Fig. 5a, b). This patient developed wound dehiscence due to poor healing or premature suture removal as we had removed them on 7th postop day.

Postoperatively, there was localised collection in 15 patients, which was relieved by intermittent needle aspiration. Localised wound infection was observed in 20 patients. Wound dehiscence in 5 patients, which needed secondary suturing in 2 patients. Rest of the patients wound healed spontaneously by daily dressings (Fig. 6). Nerve palsy was developed in 5 patients especially in the neck mainly cervical branch of facial nerve presented as mild deviation of the angle of mouth. It was relieved postoperatively by physiotherapy.

There was recurrence in 5 patients, which was due to infection in some and due to residual microscopic cysts. They were treated conservatively by administering antibiotics and anti-inflammatory agents. Aspiration was performed in 2 patients.

Regular followup weekly for the first 3 months and later fort nightly was advised to all patients at the time of discharge. All patients were observed regularly for 6 months after the surgical removal of lymphangiomas and later we lost the followup. 5 patients turned up to our OPD with recurrence and were treated successfully by antibiotics in inflammatory cases and sclerotherapy. None of them required surgery.

<table>
<thead>
<tr>
<th>Age</th>
<th>No. of Patients</th>
<th>Sex</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;1 Year</td>
<td>6</td>
<td>4</td>
</tr>
<tr>
<td>1-5 Years</td>
<td>30</td>
<td>20</td>
</tr>
<tr>
<td>5-10 Years</td>
<td>14</td>
<td>10</td>
</tr>
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**Table 1: Showing the Demographic Details**

<table>
<thead>
<tr>
<th>Site</th>
<th>No. of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Over the Neck</td>
<td>20</td>
</tr>
<tr>
<td>In the Axilla</td>
<td>9</td>
</tr>
<tr>
<td>Over the Chest/Abdominal Wall</td>
<td>5</td>
</tr>
<tr>
<td>Over the Back</td>
<td>5</td>
</tr>
<tr>
<td>Over the Extremities</td>
<td>5</td>
</tr>
<tr>
<td>Over the Perineal Region</td>
<td>4</td>
</tr>
<tr>
<td>Retroperitoneum</td>
<td>1</td>
</tr>
<tr>
<td>Floor of the Mouth</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>50</td>
</tr>
</tbody>
</table>

**Table 2: Showing the Site of Lymphangiomas**

<table>
<thead>
<tr>
<th>Complication</th>
<th>No. of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wound Infection</td>
<td>20</td>
</tr>
<tr>
<td>Seroma</td>
<td>15</td>
</tr>
<tr>
<td>Nerve Palsies</td>
<td>5</td>
</tr>
<tr>
<td>Wound Dehiscence</td>
<td>5</td>
</tr>
<tr>
<td>Recurrence</td>
<td>5</td>
</tr>
</tbody>
</table>

**Table 3: Showing Postoperative Complications**

**Fig. 1: Showing Varied Presentation of Lymphangiomas:**

**Fig. 1a: Over the Right Breast**
Fig. 1b: In the Right Axilla

Fig. 1c: In the Neck-Left Supraclavicular Region

Fig. 1d: Over the Abdominal Wall in Left Hypochondrium

Fig. 1e: Over the Back-in Infrascapular Region

Fig. 2a: Lymphangioma in the Right Submandibular Region in the Neck

Fig. 2b: USG of Neck showing Hypoechoic Lesions of Various Sizes

Fig. 2c: CT Scan showing a Fusiform Isodense Lesion 4.5x3x2 cm Noted in the Right Side with Density Values in the Range of 40-60 HU Suggestive of Cystic Hygroma

Fig. 2d: Postoperative Image showing Scar and Drain

Fig. 2: Shows the Lymphangioma in the Neck:
Fig. 3: Showing Lymphangioma in the Left Axilla:

Fig. 3a: Clinical Presentation

Fig. 3b: Intraop Image showing Macrocytic and Microcytic Masses

Fig. 4: Shows the Retroperitoneal Lymphangioma:

Fig. 4a: Clinical Presentation showing Lump in Abdomen

Fig. 4b: CT scan image showing a very large ovoid-shaped hypodense thinly septated cystic lesion of 13x11x7 cms with average density values in the range of 20-30HU extending cranially from level of L2 and caudally up to level of pelvic inlet displacing the small bowel peripherally.

Fig. 4c: Intraop Image showing the Huge Cyst Adherent to the Sigmoid Colon, which was Removed by Resecting the Sigmoid Colon

Fig. 5: Showing Lymphangioma Over the Left Forearm

Fig. 5a: Clinical Presentation showing Swelling in the Left Forearm

Fig. 5b: Intraop Image (Microcytic Type) showing Complete Excision

Fig. 6: Showing the Postoperative Wound Healing

Fig. 6a: Healthy Scar
Lymphangiomas are rare congenital malformations of lymphatic system seen all over the body, more frequently in the cervical region. The incidence is 1.5 to 2.8 per 1,000 newborns. Previous authors like Ashcraft in the old literature has named them as cystic hygroma major and minor in the neck which constituted about 67%. Nowadays, with the advent of prenatal ultrasonography, most of the lymphangiomas were been detected in utero.

Traditionally, they are classified into 3 types. They are capillary, cavernous, and cystic lymphangiomas, which has no clinical usefulness. In 1996, Mulliken classified into macrocystic (formerly cystic hygroma), microcystic (formerly cavernous lymphangioma), and mixed type, which was adopted by International Society for the Study of Vascular Anomalies. Macroscopic lymphangioma is commonly seen on the neck and axilla. Microcystic and mixed type of lymphangiomas are seen in tongue and floor of the mouth. The lesions maybe present at the time of birth in 50%-70% of children or prenatally and 80%-90% are present within 5 years of life. The lymphangiomas tend to grow slowly with the child and are asymptomatic at diagnosis, but they become symptomatic due to sudden enlargement followed by infection or spontaneous bleeding into some of the cysts. The most common presenting symptom is presence of soft cystic mass. The mass maybe small and unnoticed in early infancy period and gradually increased in size due to incidental trauma at the site or haemorrhage in the cyst. Neck swellings were diagnosed by the presence of soft, lobulated, compressible swelling located in the posterior cervical triangle and brilliantly transilluminant. Children with uncomplicated lymphangioma look healthy. Clinical presentation depends on the site, which include disfigurement, mass, pressure effects (respiratory distress or dysphagia), acute abdomen, distension of the abdomen, lump abdomen, or intestinal obstruction. 85% of lymphangiomas, which are present in the neck are unilateral and quite large in size. Pathologically, lymphangiomas are cysts or pockets of lymphatic fluid collection, which consist of multiple cysts connected to each other by small lymphatic channels. They contain clear, straw-coloured fluid unless infection or bleeding occurs into it.

Diagnosis was based on physical examination, ultrasonography, CT scan, and MRI in some cases. Physical examination showed as a soft, multilobulated mass with brilliant transillumination. However, in case of infection or intracystic haemorrhage, transillumination was negative. Ultrasonography helped in classifying the lesion (macrocystic, microcystic, or mixed), location, relation to surrounding structures, and extent of the lesion. Prenatal ultrasonography helps in the diagnosis of lymphangiomas in the intrauterine life particularly when they are located in the neck region. Doppler study shows relationship to the vessels surrounding the lesion, CT and MRI are contributory to ultrasonography for better delineation of the mass in relation to the neighbouring important or vital structures and this helped in planning the surgical management. CT imaging of lymphangioma appears as multilocular transpatial masses of fluid attenuation. MRI scan was the more accurate technique for evaluating the extent of the tumour, relationship of the tumour with the neurovascular structures, and associated venous anomalies.

Surgical removal of lymphangioma is the treatment of choice. Total or subtotal excision of the lymphangioma is preferred. Complete excision is recommended to prevent the incidence of recurrence. Microcystic lymphangiomas have the tendency to infiltrate and extend into and around the neighbouring structures making complete excision difficult. However, subtotal excision is recommended to avoid the injury to surrounding vital structures especially in the neck region. Prognosis varies with the type of malformation (microcystic, macrocystic, or mixed variety), location of the lesion, mucosal involvement. Apart from surgical removal, other treatment modalities are available. Many authors have described about various other techniques like aspiration, sclerotherapy with alcohol and drugs like bleomycin and OK-432 as different ways of treatment.

In our series, all patients underwent surgical removal in the form of total or subtotal excision. Postoperative complications were observed more in mixed variety with subtotal excision (Table 3). Wound infection was seen in 40% of our series, which was treated with antibiotics. In 10% of our patients developed fascial nerve palsy, wound dehiscence respectively. Seroma was developed in 30% of our patients, which was treated by intermittent needle aspiration.

CONCLUSIONS
Our study discussed 50 patients with varied presentation of lymphangiomas seen all over the body. Majority of them found on the neck and axilla (67%), which were removed completely by surgical excision. However, we noticed few postoperative complications, which were treated successfully by intermittent aspiration and antibiotics. We believe ultrasonography helps in detecting the lesion and should be the basic investigation of choice in all the cases. Colour Doppler study will help in detecting the relation of lymphangiomas with the major vessels while CT and MRI scan are complementary investigations in diagnosing intra-abdominal cysts in the region of neck and back. It shows the relation with the surrounding important structures and planning the surgical management.
REFERENCES