LYMPHATIC MALFORMATIONS-
COMPRESSIBLE MICRO MACROCYSTIC LESIONS

Dr A. Rekha M.S.¹, Keerthi Kumaran², R. Anukiran³

¹ Professor of surgery, Sri Ramachandra Medical College Chennai 116.
² Intern in general surgery Sri Ramachandra Medical College, Chennai 116.
³ Medical student, Sri Ramachandra Medical College, Chennai 116.

Abstract - Vascular malformations include arterial venous and lymphatic anomalies. Micro and macrocystic lesions constitute the lymphatic malformations. While surgical excision remains the gold standard, other modalities are considered for recurrent lymphangiomas and for those involving vital structures. Newer modalities include sclerotherapy, bleomycin, intralesional interferon and interstitial thermography.

Keywords - lymphatic malformations, lymphangioma, excision, sclerotherapy

AIM OF THE STUDY
To analyse the incidence, demographics and anatomical location of lymphatic malformations (LM) presenting to our hospital and to review literature on lymphatic malformations and their management.

METHODOLOGY
This was a retrospective study conducted at Sri Ramachandra Medical College over a one year period. Patients with a diagnosis of lymphatic malformation were included in the study. Patient medical records, discharge summaries and histopathology reports of all the patients were identified and reviewed.

DEMOGRAPHICS
The total number of patients in the study was 13, with 7 males and 6 females distributed between 0 – 50 years. The average age of the study group was 25years with 61.5% of the cases presenting in the first decade of life. About 23% of the cases were in the age group 21-30 years. The age groups 11- 20 years and 41-50 years accounted for 7.69% of cases each (Graph 1) In the 0 -10 years age class interval, there were more males (38.46 %) than females (23.07 %). However the overall incidence was about equal in both sexes. The highest incidence (53.85 %) was observed in the craniofacial region which included head, neck and oral cavity. 23.08% cases involved the lower limb (Graph 2).

The clinical presentation in all patients was a painless swelling (100%) which showed a slow increase in size. The other symptoms included a sense of heaviness (in limb lesions), slight weakness in power (lesion over the eyelid).Fig 1d and localized gigantism in one patient(fig 1a). The patients with macroglossia(fig 1b) also had difficulty in articulation due to mechanical obstruction. Examination showed that all lesions were soft and demonstrated compressibility(fig 2). The neck lesions demonstrated transillumination(fig 1c). A clinical diagnosis of lymphangioma was made in all patients. Investigations done included CT (Fig3) and MRI to document the extent of the lesion and to provide a road map for excision. Treatment included surgical excision in all patients when feasible.
DISCUSSION

INTRODUCTION

Vascular malformations are congenital lesions that arise due to disturbances in the embryogenesis of the vascular system. They can be further divided according to the flow of blood into slow-flow (capillary, venous, lymphatic), high-flow (arterial) and combined slow-flow-high flow. Lymphatic malformations (LM) can be classified based on their size, into microcystic (diameter < 1 cm), macrocystic (diameter > 1 cm) and combined microcystic-macrocystic (1, 2).

The lymphatic system develops around the fifth to sixth weeks of gestation. Lymphatic malformations consist of dilated lymphatic channels which can occur due to blockage or arrest of normal growth of the primitive lymph channels during embryogenesis. There is abnormal budding of lymphatic channels and sequestration of lymphatic tissue that retains its embryonic potential. Another theory proposes that the primitive lymphatic sac does not communicate with the
venous system. LMs can be associated with karyotypic abnormalities like trisomy 13, 18, 21, Turner’s syndrome, Noonan’s syndrome. Rarer syndromes include Fryn’s syndrome, multiple pterygium syndrome and achondroplasia. Cowchock Wapner Kurtz syndrome is a lethal disease comprising cystic hygroma, cleft palate and lymphedema. Gorham Stout syndrome is characterized by dissolution of bone caused by lymphangiomas or hemangiomas(3,4).

**Graph 2 showing the anatomical distribution of lymphatic malformations in males and females.**

Lymphatic malformations most commonly occur in the cervicofacial region accounting for almost seventy-five percent of the cases. Cystic hygroma is the most common lymphatic malformation. Other common sites, outside the head and neck, include the axilla, shoulder, chest wall, mediastinum, abdominal wall, and thigh.

The common clinical presentation includes the presence of a mass which is soft, smooth, compressible, transilluminant and often shows an episodic growth especially in the setting of an infection. They are usually seen in the neck, axilla or thigh and are often present at birth. Macrocystic lesions are large, soft masses under normal colored or bluish skin. Microcystic lesions present as small, raised lesions with clear fluid. Lymphangioma circumscriptum refers to lymphatic malformations that occur in the superficial layer of the skin and present as clusters of papules or vesicles. Lymphatic malformations may cause significant asymmetry or deformity of adjacent structures- for example craniofacial malformations may be associated with involvement of eyelids, cheeks, orbit.

The most common complication is intralesional bleeding. It can be due spontaneous or traumatic rupture of the cyst. Infection of the cyst can be due to leakage of the lymphatic fluid which causes subsequent microbial infection. Other complications can arise due to the specific anatomical location of the lymphatic malformation. For example – a lesion in the gastrointestinal tract can lead to protein loss, a lesion in the thoraco-abdominal region can lead to chylothorax or chylous ascites. A malformation compressing the trachea can cause respiratory difficulty. Feeding difficulties can be present in children if the lesions involve the oropharynx or hypopharynx.

**DIAGNOSIS**

Imaging modalities like ultrasound, MRI and CT are mainly used to assess the size and extent of the lesion. They can also be used to determine the relationship with adjacent structures to plan for appropriate surgical techniques. On MRI, lymphatic malformations show a typical solid appearance with low intensity on T1-weighted spin-echo image, which is equal to that of venous VMs and less intense than that of hemangiomas. A typical rim enhancement can be seen on administration of contrast. Microcystic malformations are hyperintense on T2-weighted sequence. Ultrasound plays a very important role in prenatal diagnosis of large lesions. CT can help in differentiating lymphatic malformations from other vascular malformations(4,5,6).

**TREATMENT AND MANAGEMENT**

Surgical excision of the lymphatic malformation can be done in superficial lesions (lymphangioma circumscriptum) or deep lesions (cystic hygroma). The extent of the lesion must be carefully determined prior to the surgery and extreme caution must be exercised to prevent injury to the adjacent structures(6,7). Incomplete excision can cause recurrence or repeated infection. Infected lymphangiomas should be first treated with intravenous antibiotics, followed by definitive surgery after resolution of infection.

Serial aspiration of the cysts has shown promising results. Percutaneous drainage and sclerotherapy(8,9,10) can be employed as first line treatment or for recurrence after surgical excision. A fibrin sealant may be used after aspiration of the cyst. Common side effects of repeated aspiration include bleeding, infection and recurrence.

Endovascular treatment with sclerotherapy can also be done especially for lesions that are surgically inaccessible or have a wide extension beyond deep fascia to involve muscles, tendons or bone. It is particularly effective for macrocystic lesions. Some commonly used sclerosants are doxycycline, ethanol, bleomycin(10,11), sodium tetradecyl sulfate. Some of the potential side effects are tissue necrosis, pulmonary embolism, nerve injury, venous thrombosis. Chronic complications include perilesional fibrosis. A newer sclerosant OK-432 (Picibanil- derived from a less virulent strain of type 3, group A streptococcus pyogenes) has the main advantage of absence of perilesional fibrosis and has been proposed as first line treatment over the past decade(12).

Newer treatment options include interferon-alpha treatment. IFN-alpha acts by inhibiting the proliferation of endothelial cells lining the lymphangioma by having an indirect effect on the growth factors involved in angiogenesis(13). IFN-alpha therapy has been successfully employed for lesions which are surgically irresectable. Some side effects include fever, anorexia, fatigue and rarely serious complications such as spastic diplegia.

Laser treatment can be employed as a part of the comprehensive treatment plan(14, 15). Carbon dioxide laser is a novel treatment option especially in lymphangiomas of the oral cavity. Studies have shown decreased perioperative blood loss as well as decreased surgical time and early patient
recovery. Pulsed dye laser has been used for the excision of lymphangiomas of the oral cavity.

Radiofrequency ablation(16) is a new technique which employs high frequency laser to selectively debulk superficial lesions and microcysts especially those found intraorally(17,18,19). It destroys tissues at low temperatures(40-70°C) without damaging the adjacent healthy tissue. The reduced thermal energy prevents the regrowth of tissue, thereby reducing the chances of postoperative complications like recurrence, bleeding or infection(20).

Magnetic resonance controlled laser induced interstitial thermotherapy (MR-LITT) is being used primarily for minimally invasive tumors like hepatic metastatic deposits. However it has potential use in ablating lymphangiomas, especially in the head and neck region. This MR-LITT technique has the important advantage of accurate topographic and spatial identification of the lesion, thereby reducing the incidence of complications like bleeding, necrosis or incomplete ablation(21). In comparison to conventional surgical techniques, this technique results in decreased hospital stay for patients and reduced chance of infection and recurrence.

Alternative treatment options under research include use of oral propranolol, intraläsional bleomycin and intraläsional bevacizumab(23) for intractable lymphatic malformations.

REFERENCES