Giant Lymphangioma Circumscriptum – A Therapeutic Challenge

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Abstract: Lymphangioma Circumscriptum (LC) is a rare condition which poses a therapeutic challenge when it is extensive. Here we report the largest lymphangioma circumscriptum in English literature till date along with technical considerations in its surgical management.

Keywords: Lymphangioma circumscriptum, lymphangioma diffusum, lymphangioma, giant lymphangioma, microcystic lymphatic malformation, surgical excision.

1. INTRODUCTION

Lymphangioma is a rare hamartomatous lesion of the lymphatic system. The present classification divides this entity into two groups based on the depth and size of abnormal lymphatic vessels. The group with superficial vesicles is called lymphangioma circumscriptum (LC) and the deep seated group constitutes cavernous lymphangioma and cystic hygroma. This paper reports a case of giant lymphangioma circumscriptum over the left thigh along with successful surgical management.

2. CASE REPORT

A 35 year old male presented with multiple vesicular lesions involving nearly the whole of left thigh, noticed since childhood which was gradually progressive. There were multiple pink coloured vesicles over the anterior thigh (Figure **1a**) and red coloured vesicles over medial and posterior aspect of thigh along with a hard nodule of size 5 x 4 cm over the posterior aspect of thigh (Figure **1b**). After duplex scanning to exclude the hemangiomatous component, the edge biopsy of the lesion revealed lymphangioma circumscriptum.

The lesion was excised supra-fascially with gross margin of 1-1.5cm. Lymphatic sinusoids entering into the inter-muscular septum were identified and ligated. Excision over the anterior thigh was bloodless, but excessive bleeding over posterior thigh. Meshed split skin graft was done on tenth day after lymphorrhea subsided. Graft take up was 85-90% with graft loss over the base of femoral triangle (Figure **1e**).

Histology: Focally thinned out stratified squamous epithelium arranged in papillary fold. Papillary and reticular dermis showed dilated lymphatic channels lined by flattened epithelium. Stroma showed dense lymphocytic infiltration and aggregates of lymphocytes forming follicles. Focal peripheral area showed lobules of congested capillaries (Figure **1c**). There were areas of haemorrhage seen on the lesion from posterior thigh but no hemangioma (Figure **1d**) or malignancy.

3. DISCUSSION

LC or microcystic lymphatic malformation is a hamartomatous lymphatic malformation of benign aetiology involving skin and mucous membrane. It was first described by Fox and Fox in 1879 [1]. Whimster described its pathology as subcutaneous muscle coated lymphatic cisterns which receive the lymph from surrounding tissue, instead of draining into normal lymphatic system, drains into dermal lymphatics through communicating channels [2].

LC can be divided based on the size into two types, the classical variety (<7cm) and extensive or giant (>7cm) [3]. The largest described LC till date has been described by Kudur *et al,* [4] measuring 20 x 15 cm over the anterior abdominal wall. In our patient the lesion was measuring 30 x 50 cm, making it the largest LC reported.

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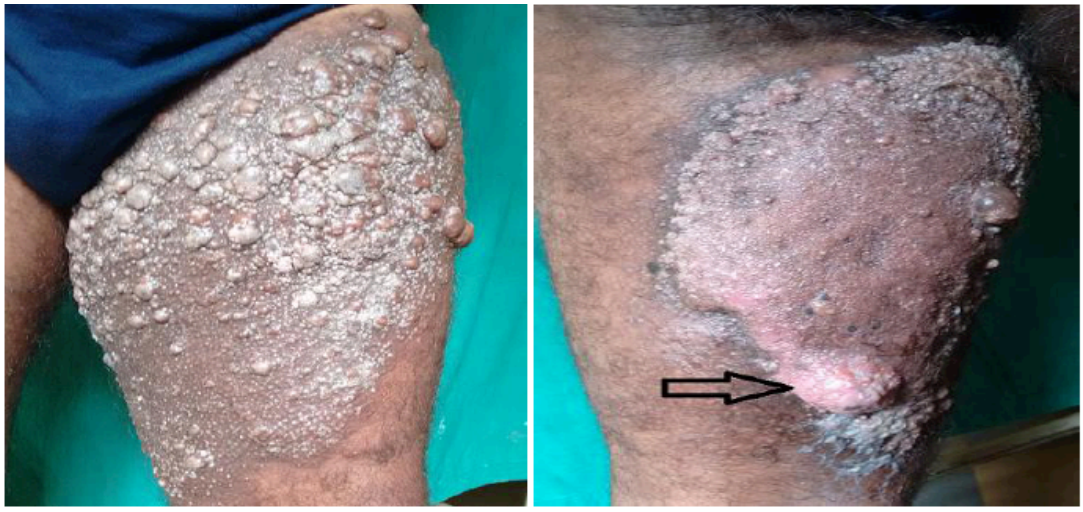

Figure 1a-b: Clinical photograph of anterior and posterior aspect of left thigh. The arrow mark shows the hard nodule.

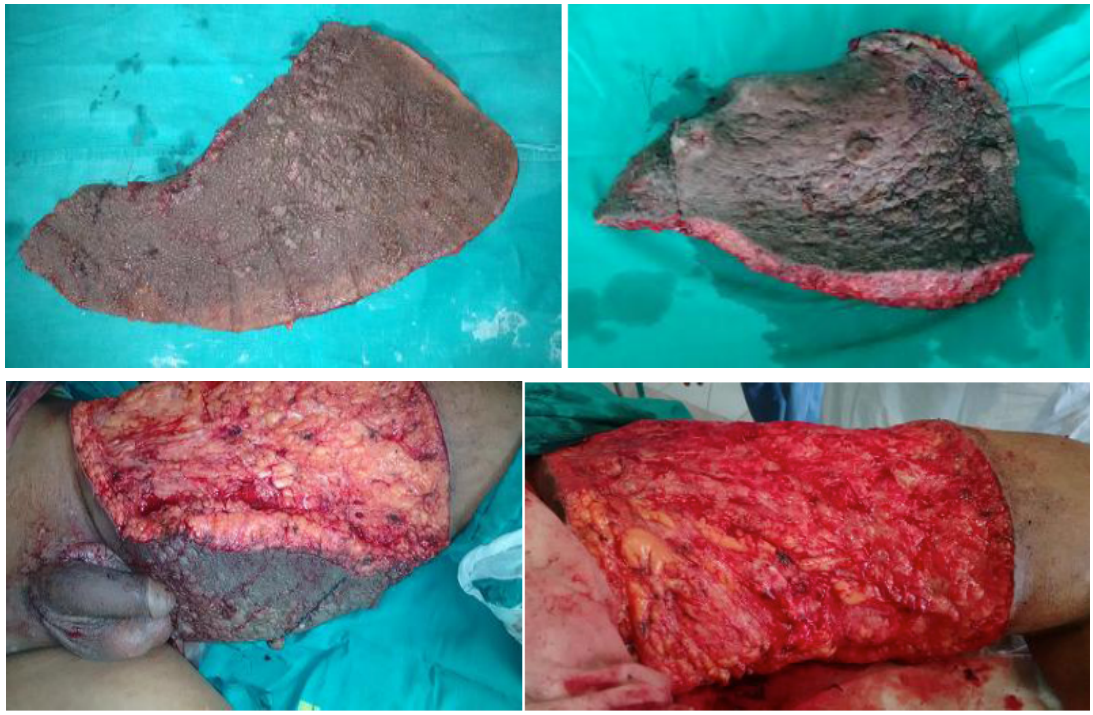

Figure 1c-d: Histology showing dilated lymphatic channels lined by flattened epithelium in papillary and reticular dermis in both while area of congested capillaries and haemorrhagic spots in the latter.

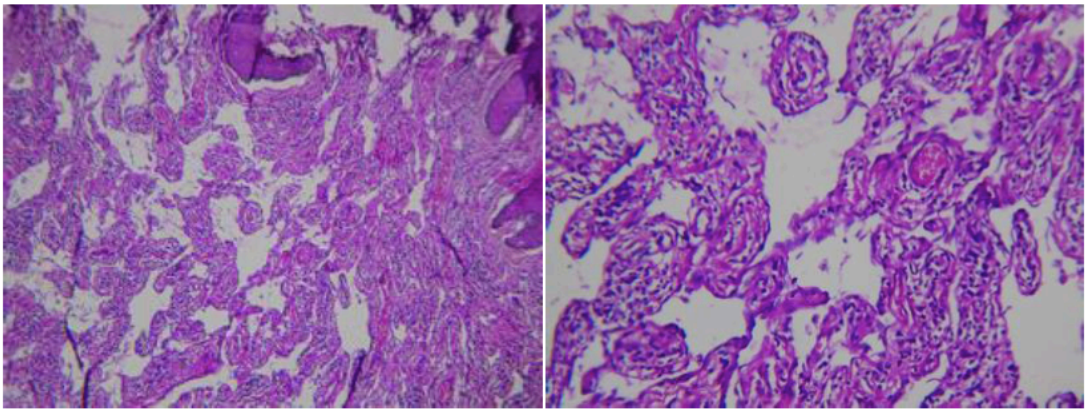

Figure 1e: Post-operative clinical picture showing good graft take.

Table 1:

Authors' Surgical Tips for Excision of Extensive Type of LC
Wider and deeper excision till fascia to radically remove all lymphatics.
Identification of large sinusoids and ligation and/or cauterization to prevent lymphorrhea and subsequent wound infection and graft loss.
Special precautions near the base of the femoral triangle at its entry into the lymph nodes and adjacent vital structures to prevent injury.
Delay the skin grafting till the lymphorrhea subsides.
Meshed collagen dressing may be preferred which would prevent wound infection and help in draining of lymphorrhea.
Meshed skin grafting wound increase the graft uptake.
Frequent change of dressing as compared to regular skin graft to prevent the maceration of the graft and subsequent graft loss.
Calcium alginate may be preferred over the graft to increase the absorption of lymphorrhea

Although benign in nature, it poses a diagnostic challenge as it mimics other infectious etiology such as herpes zoster, molluscum contagiosum and genital warts resulting in treatment failure [5]. We made a definitive diagnosis with a tissue biopsy in our case and biopsy remains the gold standard and one case of squamous cell carcinoma has been described arising from LC in the literature [7]. Although MRI can be used to pre operatively to define the lesion, its role is not clear [6].

The treatment options are excision, laser, sclerotherapy, electro coagulation and cryosurgery [4]. Only complete surgical excision of the deep lymphatic cistern can cure the patient [2].

Excessive bleeding could be encountered, either due to previous hemorrhage or possible hemangioma component and it was due to earlier hemorrhage in our case. Lymphorrhea due to the opening of the large lymphatic sinusoids, subsequent infection and graft failure and this necessitates accurate identification and ligation of these thin walled sinusoids (Table **1**).

CONCLUSION

Giant Lymphangioma circumscriptum poses a therapeutic challenge and technical modifications may have to be adopted to prevent recurrence and improve the graft uptake.

CONFLICT OF INTEREST STATEMENT

All the authors declare that there is no conflict of interest.

REFERENCES

[1] Fox T and Fox TC. On a case of lymphangiectodes with an account of the histology of the growth. Trans Path Soc Lond 1879; 30: 470.

[2] Whimster I. The pathology of lymphangioma circumscriptum. Br J Dermatol 1976; 94: 473-86. https://doi.org/10.1111/j.1365-2133.1976.tb05134.x

[3] Aykut K, Ozdemir C and Acikel U. Infected giant lymphangioma circumscriptum treated with a combination therapy. Ann Vasc Surg 2010; 24(7): 953.e11-2. https://doi.org/10.1016/j.avsg.2010.02.048

[4] Kudur MH, Hulmani M. Extensive and invasive lymphangioma circumscriptum in a young female: A rare case report and review of the literature. Indian Dermatol Online J 2013; 4(3): 199-201. https://doi.org/10.4103/2229-5178.115516

[5] Yu JTHT and Yau KC. Lymphangioma circumscriptum of the skin. Hong Kong J Dermatol Venereol 2016; 14: 129-133.

[6] McAlvany JP, Jorizzo JL, Zanolli D, Auringer S, Prichard E, Krowchuk DP, et al. Magnetic resonance imaging in the evaluation of lymphangioma circumscriptum. Arch Dermatol 1993; 129: 194-7. https://doi.org/10.1001/archderm.1993.01680230078009

[7] Wilson GR, Cox NH, McLean NR, et al. Squamous cell carcinoma arising within congenital lymphangioma circumscriptum. Br J Dermatol 1993; 129: 337-339. https://doi.org/10.1111/j.1365-2133.1993.tb11859.x