

## Review Article

# Lymphangioma circumscriptum of the skin

## 皮膚局限性淋巴管瘤

JTHT Yu 余浩德 and KC Yau 游高照

---

Lymphangioma circumscriptum consists of persistent localised or diffused translucent and red vesicles. They are primary or acquired hamartomatous lymphatic malformations and can occur anywhere on the body. Destructive treatment is frequently unsuccessful and is characterised by rapid recurrence. Surgical excision is curative but often involves extensive area of skin. Observation of this benign entity is probably the best strategy.

局限性淋巴管瘤由持續的局限或彌散的紅色透明小泡組成。為原發或繼發的淋巴錯構瘤，可出現於全身任何地方。破壞性療法常常無效並迅速復發，外科手術可以治癒，但常影響廣泛皮區。對此良性疾患，觀察可能是最佳策略。

**Keywords:** Lymphangioma circumscriptum

**關鍵詞：**局限性淋巴管瘤

---

## Introduction

Lymphangiomas are uncommon, hamartomatous malformations of the lymphatic system that involve the skin and subcutaneous tissues and was first described by Fox and Fox in 1879.<sup>1</sup> The present classification used divides these lesions into two

major groups based on the depth and the size of these abnormal lymph vessels.<sup>2</sup> The group with superficial vesicles is called lymphangioma circumscriptum (LC) and the deep-seated group includes cavernous lymphangioma and cystic hygroma. LC is a rare benign disorder of unknown aetiology involving the lymphatic channels in the deep dermal and subcutaneous layers. LC can occur as a primary abnormality or secondary to damaged lymphatic channels.

---

**Social Hygiene Service, Department of Health, Hong Kong**

JTHT Yu, BM BCh(Oxon), MRCP(UK)

**Public Health Laboratory Centre, Department of Health, Hong Kong**

KC Yau, MBChB(CUHK), FRCPA

Correspondence to: Dr. JTHT Yu

Yau Ma Tei Dermatology Clinic, 12/F, Yau Ma Tei Specialist Clinic, 143 Battery Street, Kowloon, Hong Kong

## Objective

To promote proper identification and management of this rare entity by presenting a review of cases in Hong Kong with illustrations of its clinical presentation and pathology.

## Method

Skin biopsy reports from territory wide dermatology and social hygiene clinics in the Department of Health, Hong Kong were screened manually for the time period from 1st January 1995 to 1st January 2005. Eleven cases of LC were identified on histology.

## Result

All eleven cases of LC identified were of the primary type. There were seven female (64%) and four male (36%). The mean age was 19.5 years (range 4-46 years). The most common site of presentation was the buttock and thigh (55%), followed by the trunk (36%). Five patients reported bleeding and discharge from the lesions. Four patients had itchy vesicles and three patients complained of pain. None reported secondary infection of the vesicles. Nine patients were offered symptomatic treatment with topical antibiotic while two patients received cryosurgery. In both patients treated with cryosurgery, the vesicles recurred after a few months.

## Discussions

LC can occur anywhere on the body including the skin and mucous membranes. Several classifications of lymphangioma have been proposed, but they are often confusing. The most accepted one distinguishes the two main forms of LC: primary and acquired.<sup>2</sup> Both forms have a similar clinical appearance characterised by

clusters of diffuse thin-walled, translucent vesicles up to 5 mm in diameter and filled with clear lymphatic fluid (Figure 1). The presence of varying amounts of blood may give the vesicles a pink colour. Differential diagnoses include haemangioma, herpes infections and molluscum contagiosum. These lesions can also have a warty appearance on their surface and can be confused with viral warts.<sup>3,4</sup> Though similar in terms of clinical features and symptoms, the primary and acquired forms of LC differ in terms of patient's age at first appearance, size of the lesion and distribution over the body (Table 1).

The largest series was reported by Peachey et al.<sup>2</sup> This series comprised of 65 cases at a female to male ratio of 2:1, comparable to our series. However, about half of the patients in Peachey' series presented before the age of five; which was much earlier than our findings. The most common sites of involvement in the primary form were the proximal extremities, the



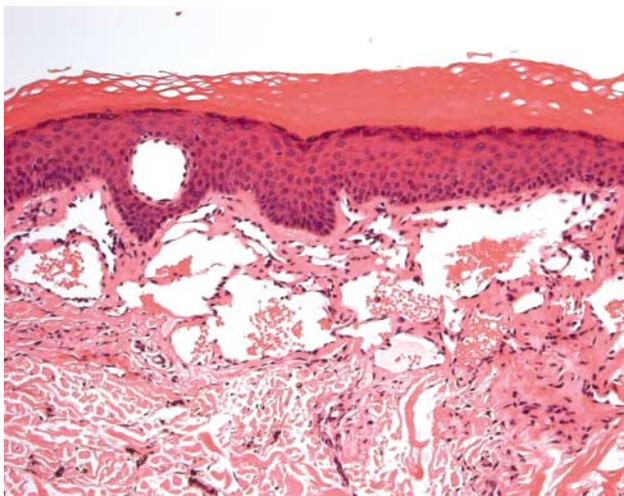
**Figure 1.** Lymphangioma circumscriptum on proximal thigh.

**Table 1.** Classification of lymphangioma circumscriptum

	Classic	Acquired
Localisation	Proximal limbs	Various sites
Size	≥1 cm <sup>2</sup>	≤1 cm <sup>2</sup>
Age	<30 y	Any age
Symptoms	Oozing of fluid from vesicles, crusting, infection, swelling and pain	

buttocks followed by the trunk. This was also consistent with the clinical features in our local series. However, they can sometimes be found in the intestines, pancreas and mesentery.<sup>5,6</sup> Deeper cystic lesions usually occur in areas of loose and areolar tissue such as neck, axilla and groin. Their skin involvement ranges from small, well-demarcated areas to large, diffuse regions with unclear borders.

The aetiological factors for primary LC are not clear. Primary lesions are considered as a circumscribed developmental defect of the lymphatics. The typical history of primary LC involves the development of a small number of vesicles during early childhood. Subsequently, they increase in number, and the area of skin involved continues to expand. Vesicles and other skin abnormalities may not be noticed until years later. LC is not known to have any association with any congenital disorder although there was one Korean case report suggesting that LC can be found in Cobb syndrome. Cobb syndrome describes the association of a spinal angioma and a cutaneous angioma such as nevus flammeus or angiokeratoma of the corresponding dermatome.<sup>7</sup>

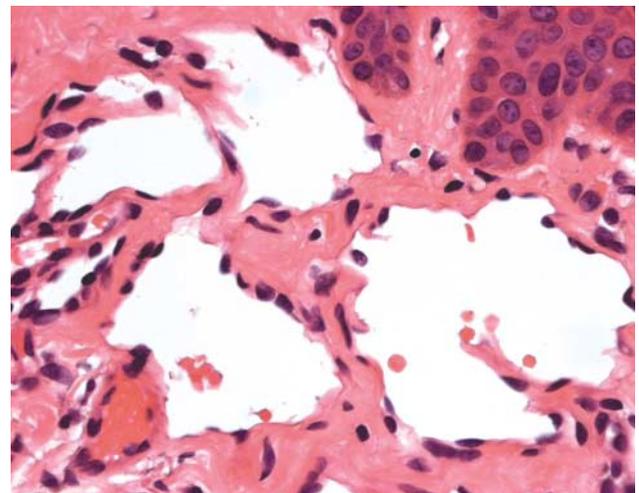


**Figure 2.** Numerous dilated lymphatics expanding the papillary dermis. The overlying epidermis shows mild acanthosis and some of the dilated lymphatics contain red blood cells (H&E, Original magnification x 10).

In acquired LC, architectural disruption of previously normal lymphatic channels leads to sequestration and further dilatation of the existing lymphatics. Predisposing factors include surgery and radiotherapy at axilla for breast cancer,<sup>8</sup> at vulvar sites in cervical cancer<sup>9</sup> or at different sites in tuberculosis.<sup>10</sup> Acquired LC usually presents at an older age and can be found at any sites following damage of the existing lymphatic system.

## Histopathology

Microscopically, the vesicles in LC are greatly dilated lymphatic channels that cause the papillary dermis to expand. They may be associated with acanthosis and hyperkeratosis (Figure 2). These channels are numerous in the upper dermis and often extend to the subcutis. The deeper vessels seem to have a larger caliber, and they often have a thick wall that contains smooth muscle. The lumen is filled with lymphatic fluid, but it often contains red blood cells, lymphocytes, macrophages, and neutrophils. These channels are lined by flat endothelial cells (Figure 3). The interstitium often has numerous lymphoid cells and shows evidence of fibroplasia.



**Figure 3.** The lymphatic channels are lined by single layer of bland endothelium (H&E, Original magnification x 40).

Whimster studied the pathogenesis of LC.<sup>11</sup> He proposed that the basic pathologic process is the collection of lymphatic cisterns in the deep subcutaneous tissue. These cisterns might arise from a primitive lymph sac that fails to connect with the rest of the lymphatic system during its embryonic development. The lymphatic cisterns have a thick coat of muscle fibers. Rhythmic contractions of the smooth muscle increase the intramural pressure and subsequently dilated channels protrude from the cistern walls toward the skin forming the LC. Whimster's observations were supported by lymphangiographic and radiographic studies.

## Management

Lymphangiomas are benign hamartomatous malformations. This uncommon pathology poses a diagnostic challenge because its clinical appearance at presentation can mimic infectious processes such as herpes zoster, molluscum contagiosum and genital warts, leading to inappropriate treatment. Magnetic resonance imaging is required to define the entire anatomy of the lesion.<sup>12</sup>

Treatment modalities are the same for both primary and acquired LC. These modalities include surgical excision,<sup>11</sup> laser therapy,<sup>13,14</sup> sclerotherapy,<sup>15</sup> electrocoagulation<sup>16</sup> and cryosurgery.<sup>17</sup> According to the "Whimster's hypothesis", only complete surgical excision of the deep lymphatic cistern can cure the patient. However, this may not be feasible in some situations when the involvement is extensive. Recurrence is the rule with other forms of destructive therapy. Thus a conservative approach for symptomatic control such as bleeding, infection and pain is a more acceptable strategy. Radiotherapy should be avoided in treating LC as two cases of lymphangiosarcoma were reported arising from LC.<sup>18</sup>

## Conclusion

LC is benign lymphatic malformation with few complications. Treatment is difficult for high recurrence rate is associated with simple destructive therapy. Curity is possible but often needs extensive surgical excision of skin and underlying tissue.

## References

1. Fox T, Fox TC. On a case of lymphangiectodes with an account of the histology of the growth. *Trans path Soc Lond* 1879;30:470.
2. Peachey RD, Lim CC, Whimster IW. Lymphangioma of skin. A review of 65 cases. *Br J Dermatol* 1970;83: 519-27.
3. Darmstadt GL. Perianal lymphangioma circumscriptum mistaken for genital warts. *Pediatrics* 1996;98(3 Pt 1): 461-3.
4. Gupta S, Radotra BD, Javaheri SM, Kumar B. Lymphangioma circumscriptum of the penis mimicking venereal lesions. *J Eur Acad Dermatol Venereol* 2003; 17:598-600.
5. Amadori G, Micciolo R, Poletti A. A case of intra-abdominal multiple lymphangiomas in an adult in whom the immunological evaluation supported the diagnosis. *Eur J Gastroenterol Hepatol* 1999;11:347-51.
6. Gleeson MJ, McMullin JP. Cystic lymphangiomas of the adrenal gland. *Br J Urol* 1988;62:93-4.
7. Shim JH, Lee DW, Cho BK. A case of Cobb syndrome associated with lymphangioma circumscriptum. *Dermatology* 1996;193:45-7.
8. Leshin B, Whitaker DC, Foucar E. Lymphangioma circumscriptum following mastectomy and radiation therapy. *J Am Acad Dermatol* 1986;15(5 Pt 2): 1117-9.
9. Ambrojo P, Cogolludo EF, Aguilar A, Sanchez Yus E, Sanchez de Paz F. Cutaneous lymphangiectases after therapy for carcinoma of the cervix--a case with unusual clinical and histological features. *Clin Exp Dermatol* 1990;15:57-9.
10. Di Leonardo M, Jacoby RA. Acquired cutaneous lymphangiectasias secondary to scarring from scrofuloderma. *J Am Acad Dermatol* 1986;14: 688-90.
11. Whimster IW. The pathology of lymphangioma circumscriptum. *Br J Dermatol* 1976;94:473-86.
12. 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.
13. Huilgol SC, Neill S, Barlow RJ. CO(2) laser therapy of vulval lymphangiectasia and lymphangioma circumscriptum. *Dermatol Surg* 2002;28:575-7.
  14. Lai CH, Hanson SG, Mallory SB. Lymphangioma circumscriptum treated with pulsed dye laser. *Pediatr Dermatol* 2001;18:509-10.
  15. Nishi T. Lymphangioma of the labia minora with deep lymphatic involvement. *Br J Obstet Gynaecol* 1998; 105:926-7.
  16. Heuvel NV, Stolz E, Notowicz A. Lymphangiectases of the vulva in a patient with lymph node tuberculosis. *Int J Dermatol* 1979;18:65-6.
  17. Handfield-Jones SE, Prendiville WJ, Norman S. Vulval lymphangiectasia. *Genitourin Med* 1989;65: 335-7.
  18. King DT, Duffy DM, Hirose FM, Gurevitch AW. Lymphangiosarcoma arising from lymphangioma circumscriptum. *Arch Dermatol* 1979;115: 969-72.