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# A Rare Case of Lymphangioma Circumscriptum

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**Abstract:** ***Background: Lymphangiomacircumscriptumis a rare cutaneous lymphaticmal formation that occur especially on children. Lymphangioma account for 4% of all vascular malformation. This is the third case had been reported in the last 9 years on Department of Dermatology Venereology Dr. M. Djamil Hospital Padang. Case: A case of lymphangioma circumscriptum in 13 years-old male was reported. There were multiple blister on upper left back and left upper arm that not felt itchy or pain that gradually increased in number and size since 6 months ago. Dermatologic state: multiple pinkish to hemorrhagic grouped of vesicles. Diagnosis is lymphangioma circumscriptum. Dermoscopy revealed of multicoloured lacunae, two tone lacunae, hypopyon and a white lines Histopathology showed achantosis and hyperkeratosis and dillated lymphatic channels contain with amorf mass. MRI showed a lymphangioma circumscriptum. Patient was consulted to surgery department for have a wide excision. Discussion: Clinical, dermoscopic, histhopatological and MRI features were describe in this case report in order to be easily recognize LC, because it is a rare case and also optional management of the cutaneous lymphangioma circumscriptum. Management of LC is challenging due to the lack of satisfactory treatments and reccurency.***

**Keywords:** dermoscopy, hypopion sign, lymphangioma circumscriptum, multicoloured lacunae, rare case

## 1. Introduction

Lymphangioma circumscriptum (LC), also known as “capillary lymphangioma,” “lymphangiectasia, ” and “dermal lymphangioma, ” is a rare benign skin disorder involving hamartomatous lymphatic malformation of deep dermal and subcutaneous lymphatic channels. Peachey *et al*. classified LC into two main forms: classic and localized. The classic form usually appears at or soon after birth and involves proximal limbs. It is thought to be derived from muscular lymphatic cisterns which failed to segment during embryonic development. Clinically, lesions appear as vesicular and do not progress into warty plaque. On the contrary, the localized form is seen at any age and has no site predilection.[1]

Lymphangioma circumscriptum can also be divided into congenital and acquired forms. Congenital LC results from local malformation of lymphatics and manifests at birth or before 5 years of age, whereas acquired form occurs secondarily due to obstruction of lymphatics commonly in the vulvar region and can manifest at any age secondary to pelvic surgery, radiation therapy, and infection such as tuberculosis, Crohn's disease, and so on. The common sites are axillary folds, shoulder, upper arm, scrotum, penis, rectum, and vulva.[1]

Lymphangioma circumscriptum develops as a result of collection of subcutaneous lymph cisterns during embryonic development, which are not connected to the lymphatic system and therefore unable to drain the lymph received from the surrounding tissue. The cisterns are lined with muscle that contracts and, by applying pressure, produces protrusions on the skin. Acquired LC develops due to injury to deep collecting lymphatics, caused by radiotherapy damage or infections such as filariasis, lymphogranuloma venereum, or tuberculosis in advanced age. The exact etiology of LC is unknown, but various growth factors such as vascular endothelial growth factor-C (VEGF-C) and VEGF-D and their receptors on the lymphatic endothelial cells may have a role.[1]

LC is usually asymptomatic. Vesicles that contain lymphatic fluid are compared with frog spawn which are pink to copper-colored while secondary hemorrhage gives it red or black color. As the clinical presentation of LC may vary from pseudovesicles to nodules or wart-like lesions, it requires histopathology.

Histology shows dilated lymphatics which contain red and white blood cells in the epidermis and the papillary dermis, lined by flat endothelial cells. Sometimes, the epidermis shows acanthosis and hyperkeratosis with widening of papillary dermis. The deeper dermis shows wide ectatic channels with a lining of endothelium containing lymph.[1]

Lymphangioma circumscriptum LC is treated for cosmetic reasons and to prevent complications such as cellulitis. The definitive treatment for lymphangiomas is surgical excision. It has highest cure rate with recurrence rate of 17%–23% which is usually due to an improper surgical approach or inadequate excision of the tumor. Surgery has the highest risk of complications such as scarring, keloid formation, hematoma, wound infection, and nerve injury. Other treatment options include scelotherapy, cryotherapy, pulsed dye laser, electrocoagulation, $CO_2$ laser, and topical imiquimod cream. Recurrence is a rule with other forms of destructive therapies.[1]

## 2. Case Report

A 13 year-old boy, presented with asymptomatic, multiple blister which gradually increased in number and size since 6 months ago on upper left back and upper left arm. Initially

±6 years ago patient complaint of one blister on his upper left back. The blister is round, small (as big as green pea), not painful and not itchy. The blister splitted a week later and contain clear fluid. The blister was dried but then slowly grow on the same place with the same size. ± 6 months ago, the blister increased in number and size. Some blister becoming pinkish and blackish in color. Occasionally the blister would rupture and leak clear fluid, and sometimes mixed with blood, or blood, it is provoked by minor trauma like rubbing with clothes or scratching. There was no white mass (like a rice grin) came out from the blister. ± 3 months ago, the blister extended also appeared in the upper left arm. Patient never treated the complaint before.

Dermatological examination showed grouped of vesicles, multiple colour pinkish to hemorrhagic grouped of vesicles.

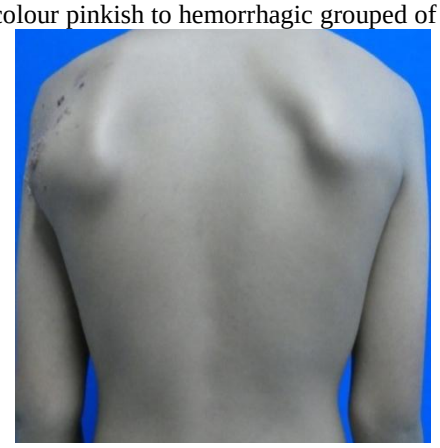

**Figure 1:** Showed the location of lesion in upper left back and left upper arm

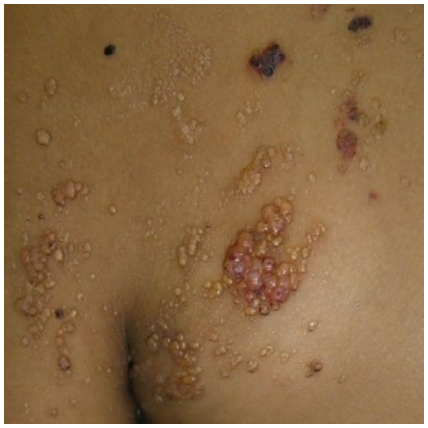

**Figure 2:** showed grouped of vesicles, multiple colour pinkish to hemorrhagic grouped of vesicle.

Dermoscopy examination showed two tone lacunae (colour transition from dark (at the bottom lacunae) to light (at the upper part lacunae), multicoloured lacuna and white lines

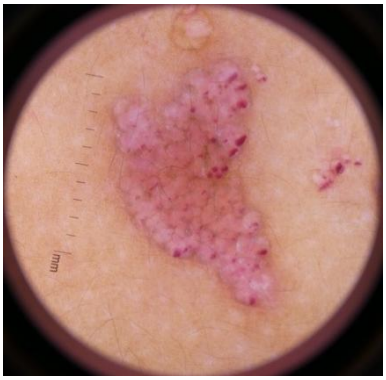

**Figure 3:** Dermoscopy examination showed two tone lacunae (hypopion sign).

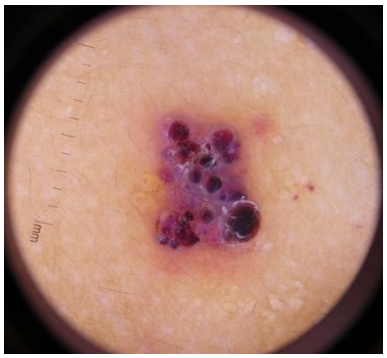

**Figure 4:** Dermoscopy examination showed pink lacunae, dark lacunae and white lines.

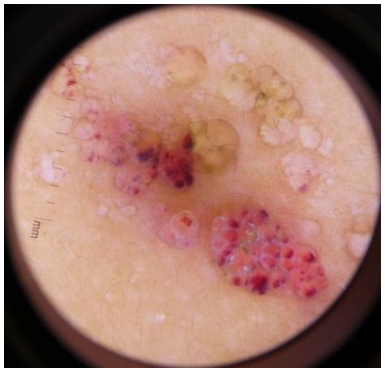

**Figure 5:** Dermoscopy examination showed multicoloured lacunae with hypopion sign.

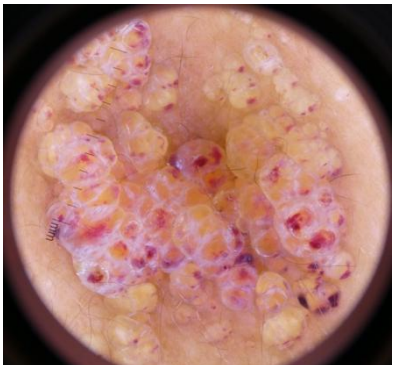

**Figure 6:** Dermoscopy examination showed yellow to pink lacunae with dark red lacunae and white lines.

Histopathological examination showed achantosis and hyperkeratosis in epidermis. In papillary dermis there were dillated lymphatic channels contain with amorf mass, some vessels channels contain valves, and containing lymphocytes

and few erythrocytes. These channels are lined by flat endothelial cells. Lymphocyte cells also appears in dermis, especially around the proliferation of vessels

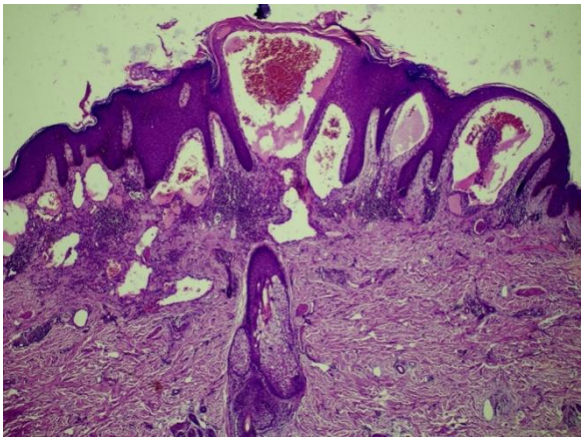

**Figure 7:** Histhopatological examination showed hiperkeratosis, achantosis, amorf mass/lymph and lymphocites

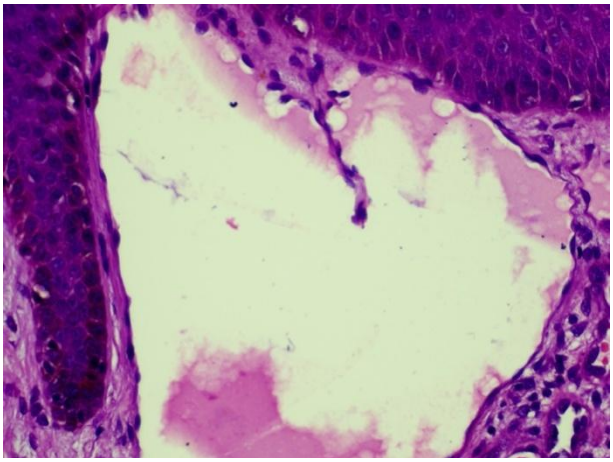

**Figure 7:** Histhopatological examination showed amorf mass/lymph and valves

The magnetic resonance imaging (MRI) showed serpiginous structure in cutaneous and subcutaneous tissue on leftscapula to left axilla, which are hypointense in T1W1 and T2 Flair and hyperintense T2W1 and T2FS.

Diagnosis of lymphangioma circumscriptum based on clinical practice, dermoscopy examination, mapping the extent and location with the MRI and confirmed by histhopatologic examination.

There is no specific treatment in our department. We referred the patient to surgical department for surgical excision

## 3. Discussion

Lymphaticmal formations or lymphangiomas are rare benign hamartomas. Lymphangioma can occur at any age, but majority are seen in children. Approximately 50% present at birth and 90% are diagnosed before the age of 2 years.[1] In the United States, congenital lymphaticmal formations occur in 1.2 to 2.8 per 1000 livebirths. Lymphangioma circum scriptum is the most common congenital lymphaticmal formation, although its true incidence is unknown.[2] Yu JTHT, etal. (Hongkong, 2006) reported 11 cases of lymphangioma circumscriptum recorder from 1st January 1995 to 1st January 2005 that were diagnosed by histopathology examination.[3] There is no official report about incidence of lymphangioma in Asia and Indonesia recently. In Indonesia there were somecasereport had been reported. Medikawati in 2014 etalfrom Bali, Olinaetal from Palembang, Adisty etalfrom Surabaya and three cases of lymphangioma circumscriptum of vulva in Jakarta and Surabaya.[4, 5, 6] . In out patiens of Dermato-Venerology Department in Dr. M. Djamil Hospital, this is the third case in the last 9 years.

Lymphangioma circumscriptum is a congenital cutaneous lymphaticmal formation characterized by translucent vesicles resembling “frogs pawn.” The vesicles harbor clear or serosanguinous fluid composed of varying degrees of lymph and blood. They mayrange in size from minute vesicles to larger bullae. Specifically, 2-4 mm clusters of vesicles with pink, red, or black discoloration as a by product of hemorrhage are common. Vesiclerupture commonly occurs, and repetitiv erupture can induce epidermal hyperplasia and hyperkeratosis, leading to a verrucous appearance. Lesions with a hemorrhagic component can become darkly pigmented. Other symptoms include lymphorrhea (leakage of lymphatic fluid), pruritus and pain.[2, 7, 8] In our patient we found various vesicle lesion from translucent vesicles, pink, and black discoloration of vesicles. Sometimes patient complaint itch and scratch the lesion it leads to leaked the clear fluid or hemmoraghae discharge from lesion. Zaballosetal ( Spain, 2017) reported total of 45 cases of lymphangioma circumscriptum were affected 26 women (58%) and 19 men (42%), ranging in agefrom 5 to 77 years (mean 29.7 years), mostlesion were located on the trunk (49%).[9] Our patient were 13 years old boy and the location of lesion is on the posterior shoulder and upper left arm.

Dermoscopy as a useful non invasive tool for aiding diagnosis. Lacunae are defined as multiple, clustered, well-demarcated, yellowish, reddish or dark-coloured structures with a roundto oval shape. The histopathological correlation of these lacunae is the presence of dilated, thin-walled vessels in thepapillary dermis which are thehallmark of many vascular tumours. The whitish, light tan to yellow coloration is due to lymphatic fluid. The pinkish or red dish colour is due to the presence of red blood cells in the dilated lymphatic channels. When they are partially or completely thrombosed the lacunae can be dark violaceous, blue- black or black in colour. These are called dark lacunae. In somecases, a two-tone lacunaor a colour transition from dark (at the bottom) to light (at the upper part) in the same lacuna was observed. This phenomenon has been named the hypopyon sign in the literature and it is due to these dimentation of blood (red or dark in colour) in the dilated lymphatic channels.[9, 10, 11]

Zaballos et al (Spain, 2017), reported dermoscopic images in 45 cases of lymphangioma sircumsriptum. Lacunae is the most common structure found in lymphangioma sircumsriptum (89% of cases), followed by red or dark-coloured in 18 cases (45%), yellowish or whitish in 14 cases

(35%) and multicoloured in eight cases (20%). The second most common dermoscopic structure was the presence of vascular structures, which were found in 82% of cases, followed by white lines (47%), the hypopyon sign or two-tone lacunae (42%) and scales (7%).[11] In our patient, dermoscopy revealed of multicoloured lacunae, two tone lacunae, hypopyon, and also white lines.

Histology remains the gold standard for definitive diagnosis although dermoscopy plays a major role in the diagnosis. Histopathology is characterized by acanthosis and papillomatosis of epidermis with numerous ectatic lymphatic channels in the upper dermis which may often extend through deep layers.[8] Haley et al also reported in dilated lymphatic vessels with a flattened endothelial lining contais lymph innsuperficial dermis.[2] Fatima et al ( Pakistan, 2015) also reported histhopatological finding from lymphangioma circumscriptum are acanthotic squamous epithelium with papillomatosis which lymphatic channels composed of ectatic dilated vessels with serum and inflammatory cells in their lumina. The lymphatic channels were seen in deeper layers along with lymphocytic aggregates.[12] In our case we found achantosis and hyperkeratosis in epidermis. In dermis we found dillated lymphatic channels contain with amorf mass, some vessels channels contain valves, and also lymphocytes and few erythrocytes. These channels are lined by flat endothelial cells.

Radiological exploration is necessary to look for deep localization. MRI can detect an involvement of the subcutaneous and deeper skin layer also to define the entire anatomy of the lesion.[2] MRI examination revelead serpiginous structure in cutaneous and subcutaneous tissue on left scapula to left axilla, which are hypointense in T1W1 and T2 Flair and hyperintense T2W1 and T2FS.

Lymphangioma circumscriptum is benign, and treatment is no required in asymptomatic patients. Indications for treatment include lymphorrhea, recurrent infections, cosmetic concerns, bleeding, and pain.[2] The primary goal of treatment is to remove or destroy the diseased lymphatics and subcutaneous components that serve as a nidus for recurrence.[7]

Various modalities of treatment include surgical excision, laser therapy, sclerotherapy, electrocoagulation and cryosurgery. Lasers, mainly the pulse dye laser (PDL) and $CO_2$ laser achieve good cosmetic results and resolution of their symptoms with minor and infrequent side effects such as dyspigmentation and mild scarring, but the recurrence rate was significantly high. Cryotherapy uses very low temperatures to cause vasoconstriction immediately followed by reactive vasodilation resulting in cellular necrosis. This, in combination with laser therapy, may have a synergistic effect for LC. Emer et al. failed to observe clinical change in an LC lesion after two treatments with PDL delivered two weeks apart.[12-13] Sclerotherapy is a useful method for the treatment of LC, but the side effects, such as pain, inflammation, hyperpigmentation, ecchymosis, localized urticaria, and skin necrosis, are considerable.[12-13] Radiotherapy is an alternative treatment for large or unresectable lesions with limited treatment options, but it is remains controversial as cases have been reported in which LC developed as a complication of previous radiotherapy sessions.[12] From review article by Perper et al ( Saudi Arabia, 2017) reported cryotherapy may prove beneficial for treating LC when combined with the PDL lasere Electrocoagulation may be valuable treatments for LC. Imiquimod is another potential noninvasive treatment for LC but in some case lesions become more prominent after following treatment.[13, 14]

The surgical excision is the main modality of treatment, although the recurrence is common, with a cure rate of 75%, because it is can eliminate the deep subcutaneous cisterns. However lymphangioma circumscriptum often has larger dermal and subcutaneous components than are clinically apparent, making complete excision difficult. All subcutaneous tissue superficial to deep fascia must be excised to encompass the deep cisterns, often necessitating extensive excision and reconstruction with flaps or skin grafts.[2, 7] Taj et al (India, 2019) reported surgical excision as the treatment for his patient, when during surgery the lesion of his patient had a cystic extension, horizontally from left iliac crest to midline, and vertically extending up to external oblique muscle on the left side.[7] Recurrence rates are around 9% after a single excision and 5% after reexcision. [2] Nair et al reported surgical has a highest cure rate and recurrence rate about 17%–23% which is usually due to an improper surgical approach or inadequate excision of the tumor. Surgery has the highest risk of complications such as scarring, keloid formation, hematoma, wound infection, and nerve injury.[1] Based on explanation above patient were treated with surgical excision because it is definitive treatment, lowest recurrence and it is available in our department.

Prognosis on this patient is bonamforquoadvitam and dubiaadbonam for quoadsanationamanddubiaad malam for quoadcosmeticum, and quoad function. Lymphangioma circumscriptum is not life-threatening. The lesions are caused by lymphaticmal formation and recurrence is usual, even with the main therapy which is surgery. There is possibility of scars after treatment and the skin's function is disturbed.

## 4. Conclusion

Lymphangioma circumscriptum is benign and rarely and extensive lymphatic malformation. The treatment of LC is quite challenging. According to previous reports many modalities has been described, but surgical excision is the main modality treatment.

## References

[1] Nair PA, Kota RK, Singhal RR, Gandhi SS. Verrucous lymphangioma circumscriptum in a child. J NTR Univ Health Sci. 2019;8:72-4.
[2] Haley MD, Tyring SK. A 22 year old man with painful vesicles on his flank. Jama. 2019
[3] Yu J, Yau K. Lymphangioma circumscriptum of the skin. Hong Kong J Dermatol Venereol 2006;14:129-33
[4] Medikawati IR, Wardhana M, Darmaputra IGN. Lymphangioma circumscriptum yang

diterapidenganbedahlistrik. Medicina. 2015; 45; 3; 176-181.
[5] Olina R, Yuliawati, Sari YM, Yahya YF. Malformasilimfatikmikrositik yang tidakumum.Buku program danabstrak 2014. 1-5
[6] Adisty DR, Zulkarnain I. Studiretrospektif : insidensidanpenatalaksanaaangenodermatosis. Berkalailmukesehatankulitdankelamin. 2016; 28;2;35-41
[7] Taj FT, Vyshak BM. Lymphangioma circumscriptum : successfully treated by surgical excision. Ajrdes. 2019;2(1):1-4
[8] Jouari OEL, Senhaji G, Benkirane S, Baybay H, Douhi Z, Elloudi S, et al. Cutaneous Lymphangioma Circumscriptum. SM Vasc Med. 2018; 3(1): 1014.
[9] Zaballos P, Pozo LJ, Argenziano G, Karaarslan IK, Landi C, Vera A. Dermoscopy of lymphangioma circumscriptum : A morphological study of 45 cases. Australian journal of dermatology. 2017;59;3:189-193.
[10] Massa AF, Moreira AI, Menezes N, Osório-Ferreira E, Baptista A. Cutaneous Lymphangioma circumscriptum - dermoscopic features. An Bras Dermatol. 2015;90(2):262-4
[11] Gomides MDA, Costa LD, Berbert ALCV, Janones RS. Cutaneous lymphangioma circumscriptum: The relevance of clinical, dermoscopic, radiological, and histological assessments. *Clin Case Rep*. 2019;7:612–615.
[12] Fatima, S., Uddin, N., Idrees, R., Minhas, K., Ahmad, Z., Ahmad, R., Kayani, N., Arif, M. (2015). Lymphangioma circumscriptum: clinicopathological spectrum of 29 cases. Journal of the College of Physicians and Surgeons Pakistan, 25(9), 658-661.
[13] Perper M, Cervantes J, Eber AE, Hsu VM, Alharbi M, et al. Lymphangioma Circumscriptum: Treatment Modalities for this Unyielding Condition. J ClinInvestigat Dermatol. 2017;5(1): 2.
[14] Puri N. Treatment options of lymphangioma circumscriptum. Indian Dermatol Online J. 2015 Jul-Aug; 6(4): 293–294

## Author Profile

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