

CORRELATION OF DERMOSCOPY WITH HISTOPATHOLOGY IN A CASE OF LYMPHANGIOMA CIRCUMSCRIPTUM

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ABSTRACT

A lymphangioma (or lymphatic malformation) is congenital proliferation of lymphatic vessels accounting for approximately 4 percent of vascular tumors and 25 percent of benign vascular tumors in children.¹ One of the predominant types is lymphangioma circumscriptum consisting of multiple, grouped vesicles that may be pink, red, or black, owing to serosanguinous fluid or hemorrhage. These contain lymph fluid and are often said to resemble frogspawn. We present a case of lymphangioma circumscriptum on the right inguinal fold in a 15-year-old girl who was diagnosed clinically based on dermoscopy and histopathology and subsequently treated with sclerotherapy followed by radio frequency ablation.

Keywords: dermoscopy, lymphangioma circumscriptum, hypopyon, sclerotherapy

CASE

A 15-year-old female presented with complain of multiple fluid filled lesions in her right inguinal region, since the age of 8 years. On examination, multiple grouped vesicles with serous and serosanguinous discharge were present in right inguinal region of dimension 8X5 cm with multiple satellite lesions in the periphery. There was also a history of right congenital inguinal hernia surgery at the age of five years. Occasionally the swelling would rupture, discharging serous fluid. Some vesicles exuded viscous serous fluid after being punctured, and the vesicle spaces were filled with pale yellow crusts. The area exhibited a white or yellowish mulberry-like appearance and diffusely thickened underlying skin. There was no associated lymphadenopathy or lymphedema. Diagnosis of lymphangioma circumscriptum was made on the basis of clinical features and dermoscopy finding, which was confirmed by histopathology.



Figure 1 Cutaneous Lymphangioma Circumscriptum

DERMOSCOPY

Multiple yellow lacunae as well as pink to dark red lacunae with variable amount of blood were present in a well circumscribed lesion. Some of the yellow lacunae showed fluid levels with clear fluid in the upper half and yellowish fluid at the bottom.

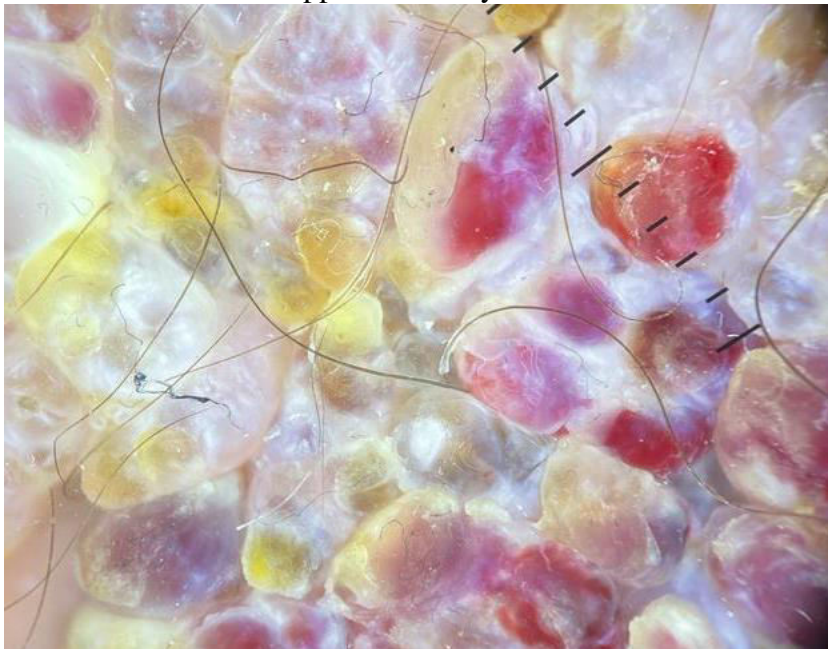


Figure 2 Yellow to pink-red lacunae with pale septa on dermoscopy (40x)

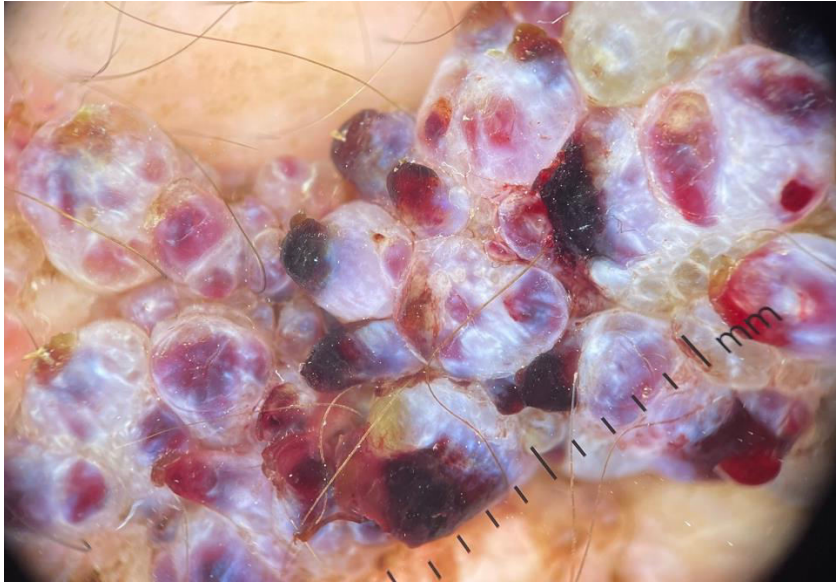


Figure 3 'Hypopyon sign' on dermoscopy

HISTOPATHOLOGICAL EXAMINATION

The sections show stratified squamous epithelium with irregular hyperplasia. The dermis has dilated spaces of different shapes and sizes, lined by a single layer of endothelial cells and containing clear fluid at one end. These channels appear partially enclosed by acanthotic epidermis. There was moderately dense infiltrate in the dermis composed of lymphocytes, a few histiocytes with an attempt towards formation of multinucleated giant cells.

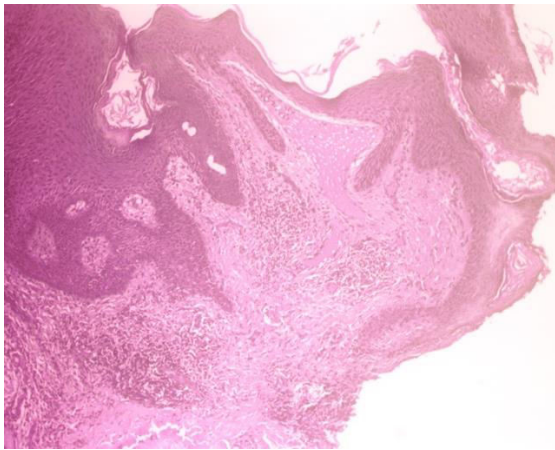


Figure 4 Histopathology of Lymphangioma circumscriptum(40x)

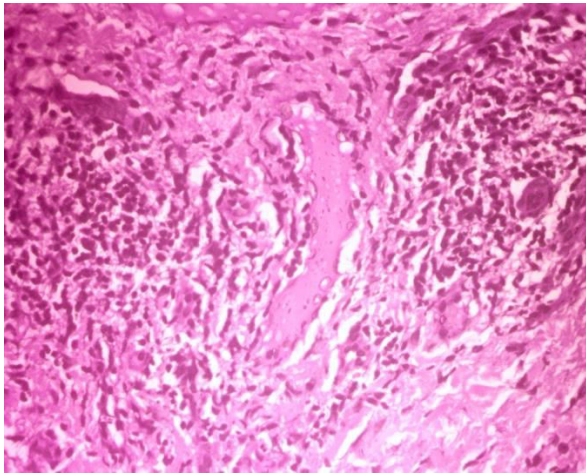


Figure 5 Dilated lymphatic spaces lined by endothelium(100x)

Dermoscopy	Histopathology
Yellow lacunae with pale septa	Dilated spaces with clear fluid lined by single layer of endothelial cells
Pink to dark red lacunae	Dilated spaces lined by endothelium with variable concentration of RBCs

Table 1: Dermoscopy and histopathological correlation

TREATMENT

5 sittings of intralesional bleomycin (1mg/ml) were given at 2 weeks interval, following which the lesion was ablated with radio-frequency in 2 sittings and it healed over 7-10 days leaving behind a hypertrophic scar. The patient was followed up for next 6 months with no signs of recurrence or any new lesion.

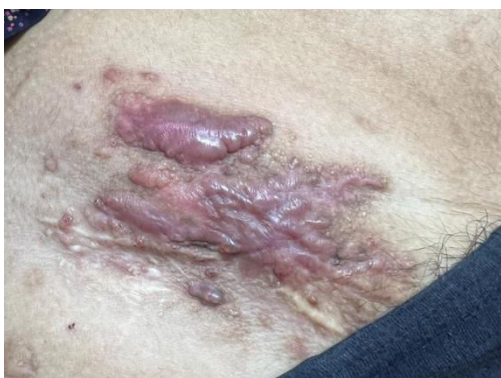


Figure 6 lymphangioma circumscriptum (Post treatment)

DISCUSSION

Lymphangioma circumscriptum, which can develop at any age but most frequently occurs in infants is one of the most prevalent types of cutaneous lymphangioma. The clinical features consist of numerous clear vesicles that may be pink, crimson, or black due to serosanguinous fluid or haemorrhage. They occur as a consequence of aberrant lymph vessel proliferation. Lymphangioma circumscriptum more usually affects the proximal extremities, trunk, axilla, and oral cavity, but it can also affect the penis, vulva, and scrotum.^{2,3}

The accuracy of diagnosing lymphangioma circumscriptum has been greatly improved by the non-invasive technique of dermoscopy. Dermoscopically, it has been proposed that CLC displays two distinct patterns: yellow lacunae surrounded by light septa without blood and yellow to pink lacunae alternating with dark-red or bluish lacunae due to blood.⁴ These findings and histologic traits were strongly related.⁵ The former can be distinguished by dilated lymphatic vessels in the dermis, the subcutaneous layer, or both. These are filled with lymphatic fluid. The latter has variable red blood cell concentrations. Some of them resembled the "hypopyon" of the eye or the "half-and-half" blister observed in Sneddon-Wilkinson disease due to the typical accumulation of fluid in the lowest portion of the lacuna.⁶

Treatment for lymphangioma circumscriptum is done to improve appearance of the lesion as well as to prevent complications like cellulitis. The effective therapy for lymphangiomata is the surgical excision of both the superficial and deep components.⁷ However, recurrence rates might be high, especially when lesions have deeper components. Additional palliative treatments include radiation therapy, superficial x-ray therapy, argon laser, CO2 laser, 900-nm diode laser, pulsed dye laser, and sclerotherapy.^{8,9,10}

Although hemangiomas react to sclerosing agents more well than lymphangiomata do, intralesional sclerotherapy has improved and reduced the requirement for other forms of therapy. For lymphatic malformation, a number of sclerosing drugs have been utilised, including picinabil (OK-432), bleomycin, doxycycline, acetic acid, alcohol, and hypertonic saline. Even while sclerotherapy is becoming more and more popular, there is no agreement on the best kind to employ.

Bleomycin in addition to being antiangiogenic, is also antiviral, antineoplastic, and antibacterial. The US Food and Drug Administration has approved systemic bleomycin as a chemotherapeutic drug. Skin cancer, persistent warts, keloids, hypertrophic scars, hemangiomas, VM, telangiectasias, condyloma acuminata, and cutaneous leishmaniasis are among the conditions for which bleomycin is used off-label in dermatology.¹¹

REFERENCES

- 1) Patel GA, Schwartz RA. Cutaneous lymphangioma circumscriptum: frog spawn on the skin. *Int J Dermatol.* 2009;48:1290–1295.
- 2) Vlastos A-T, et al. Lymphangioma circumscriptum of the vulva: a review of the literature. *Obstet Gynecol* 2003; 101:946
- 3) Swanson DL. Genital lymphangioma with recurrent cellulitis in men. *Int J Dermatol* 2006; 45:800
- 4) Arpaia N, Cassano N, Vena GA. Dermoscopic features of cutaneous lymphangioma circumscriptum. *Dermatol Surg* 2006; 32:852854.
- 5) Amini S, Kim NH, Zell DS, Oliviero MC, Rabinovitz HS. Dermoscopic-histopathologic correlation of cutaneous lymphangioma circumscriptum. *Arch Dermatol* 2008; 144:16711672.
- 6) Gencoglan G, Inanir I, Ermertcan AT. Hypopyon-like features: new dermoscopic criteria in the differential diagnosis of cutaneous lymphangioma circumscriptum and haemangiomas. *J Eur Acad Venereol.* 2012;26:1023–1025.

- 7) Browse NL. Surgical management of 'lymphangioma circumscriptum'. Br J Surg 1986; 73:585
- 8) Eliezri YD, Sklar JA. Lymphangioma circumscriptum: review and evaluation of carbon dioxide laser vaporization. J Derm Surg Oncol 1988; 14:357
- 9) Lapidoth M. Treatment of lymphangioma circumscriptum with combined radiofrequency current and 900nm diode laser. Derm Surg 2006; 32:790
- 10) Bikowski JB, Dumont AMG. Lymphangioma circumscriptum: treatment with hypertonic saline sclerotherapy. J Am Acad Dermatol 2005; 53:442
- 11) Saitta P, Krishnamurthy K, Brown LH. Bleomycin in dermatology: A review of intralesional applications. Dermatol Surg 2008;34:1299-313. †