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Lymphangioma Circumscriptum: Successfully Treated by Surgical Excision

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Authors' contributions

This work was carried out in collaboration between both authors. Author FTT designed the case report and wrote the draft of manuscript. Author BMV managed the literature search. Both authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Aim: Lymphangiomas are malformations of the lymphatic system that are located in the skin and subcutaneous tissues. We report a case of invasive lymphangioma circumscriptum successfully treated by surgery.
Presentation of a Case: A 10-year female presented with asymptomatic lesions over the left side of the abdomen since birth. Dermoscopy showed characteristic multiple yellow lacunae surrounded by septa, as well as with few lacunae showing hypopion sign. Histopathology of the lesion was suggestive of lymphangioma circumscritum.
Discussion: Depending on the size and depth of the lymph vessels, lymphangiomas are separated into two categories; the superficial group and deeper group. The superficial group contains only lymphangioma circumscriptum, whereas deeper group is comprised of caverneous lymphangioma and cystic hygroma. Of the three types lymphangioma circumscriptum is the most

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common. Diagnosis of lymphangioma circumscriptum was made and surgical excision of the lesions was done.
Conclusion: we report a case of invasive lymphangioma circumscriptum which is managed successfully by surgical excision.

Keywords: Lymphangioma circumscriptum; surgical treatment; hypopyn sign; dermascopy.

1. INTRODUCTION

Lymphangiomas are congenital lymphatic malformations accounting for 4% of all vascular tumours and 25% of benign vascular growths in children. Lymphangioma circumscriptum (LC) is a rare benign skin disorder involving hamartomatous lymphatic malformation of deep dermal and subcutaneous lymphatic channels. The term “Lymphangioma circumscriptum” was coined by Morris et al. in 1889. Usually LC is a well circumscribed lesion presenting as multiple clusters of clear or red vesicles on the surface of skin like frog spawn. Rarely, extensive and invasive forms can be seen. The term ‘circumscriptum’ appears to be a misnomer for an extensive lesion. We report a case of rare extensive type of lymphangioma circumscriptum in a young teenage girl, managed by surgical excision with acceptable cosmesis.

2. CASE REPORT

A 10-year female presented with asymptomatic lesions over the left side of the abdomen since birth. It gradually, increased in number with lesions becoming bluish and reddish in color with occasional oozing. For these complaints she had consulted a local doctor and had received tab. propranolol 10 mg once daily for 1 month with topical steroids and found no improvement. On local examination, there were multiple violaceous to hemorrhagic grouped vesicles of varying size over the left side of abdomen with surrounding hyperpigmentation (Fig. 1). Dermoscopy showed characteristic multiple yellow lacunae surrounded by septa, as well as with few lacunae showing hypopion sign. (Fig. 2) Histopathology of the lesion was suggestive of lymphangioma circumscriptum. With clinical, dermoscopy and histopathology, a diagnosis of lymphangioma circumscriptum was made and surgical excision of the lesions was done with acceptable cosmesis. However it was found during surgery that the lesions had a cystic extension, horizontally from left iliac crest to midline, and vertically extending up to external oblique muscle on the left side. (Fig. 3A, 3B).

3. DISCUSSION

Lymphangioma circumscriptum or microcystic lymphatic malformation is a rare benign skin disorder involving hamartomatous lymphatic malformation of deep dermal and subcutaneous lymphatic channels characterized by clusters of thin walled vesicles on the surface of skin like frog spawn. These vesicles are filled with either clear fluid or blood tinged fluid [1].

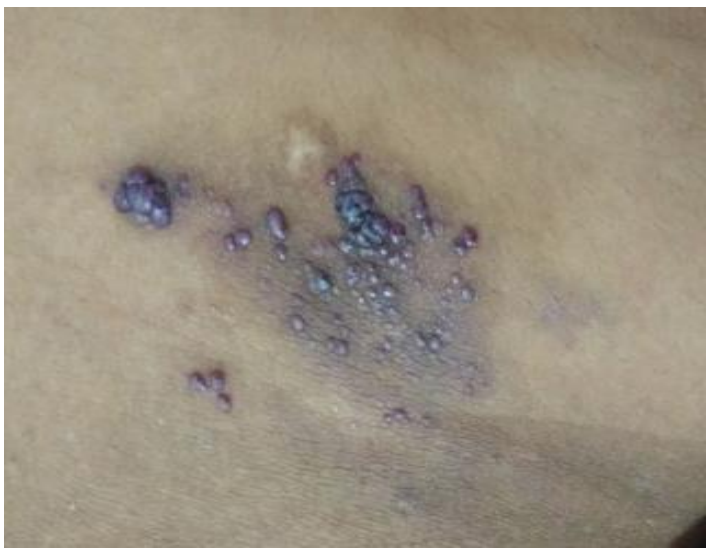

Fig. 1. Lymphangioma circumscriptum showing frog spawn appearance

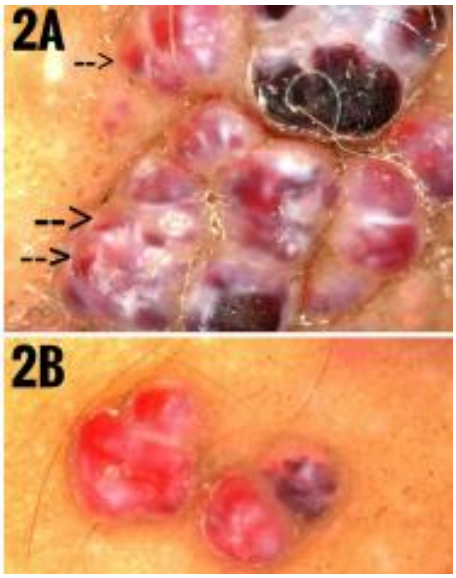

Fig. 2. Dermoscopy showing hypopyn sign

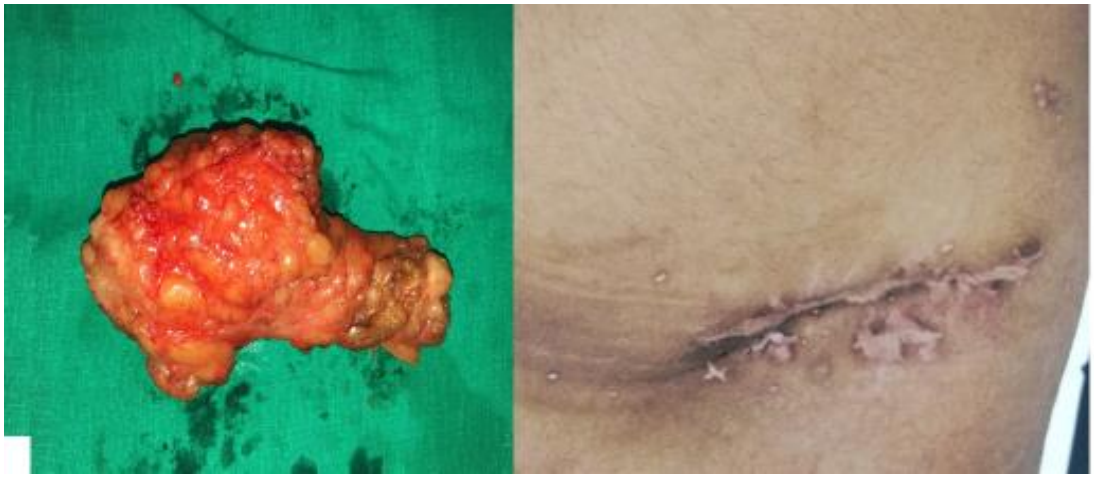

Fig. 3A . Intraoperative, showing extensive Lymphangioma circumscriptum

Fig. 3B. Post operative showing better cosmetic result

The pathogenesis of lymphangioma circumscriptum was first described by Whimster in 1976. It was postulated that large muscle coated cisterns were present in subcutaneous tissue, disconnected from the normal network of lymphatic vessels. However, they are connected to dermal lymph channels. The vesicles result from the dilatation of lymphatics in papillary dermis caused by back pressure. Based on this pathogenesis, it was suggested that excision of subcutaneous cisterns while leaving overlying skin might offer a better cosmetic and acceptable option to the patient [2].

In 1970, Peachey et al. divided LC into classic and localized forms. The classic variety of LC is seen at or soon after birth, and is often larger than 1 cm^2, seen over the proximal limbs. The localized form can be seen at any age and is smaller than1 cm^2. LC can occur as a congenital or acquired form. The congenital form will be larger in presentation and the acquired form will be small and circumscribed [1]. The LC is asymptomatic in its localized form, but the most common symptom is oozing of clear fluid mixed with blood, which occurs spontaneously or after minor trauma and rarely, extensive and invasive forms are seen, which can involve deep fascia, muscles of abdomen, buttocks and large vessels, including iliac and femoral arteries [3]. The common associations of LC are lymphedema of lower limb and cystic hygroma, and there few

reports of Proteus, Cobb and Maffucci syndromes [3].

The differential diagnosis of LC include lymphangiectasis, hemangioma, angiokeratoma, angiosarcoma, metastatic carcinoma to the skin, verrucae, molluscum contagiosum, warts, and epidermal nevi.

The dermatoscopic examination of LC reveals a lacunar and saccular pattern. There are brownish lacunas limited by pale septa or reddish areas depending on the blood content. A new dermascopic pattern reveals Hypopyon like feature [4]. It is because of sedimentation of blood, cellular components lying at the bottom and serum at the upper part, leading to transition from dark to light in some lacunae [5,6]. Apart from histology and lymphangiography, MRI is very useful in the diagnosis of LC and the modality of choice in the management of LC [3].

The surgical excision is the main modality of treatment, although the recurrence is common, with a cure rate of 75%.Other treatment options are carbon dioxide laser, pulsed dye lasers, intense pulse light, sclerotherapy, cryotherapy, superficial radiotherapy, radiofrequency ablation [7] and electrocautery. Mosche Lapidoth et al. have successfully used the combination of radiofrequency current and 900 nm diode laser in treating six patients of LC. The primary goal of treatment is to remove or destroy the diseased lymphatics and subcutaneous components that serve as a nidus for recurrence [1][2].

4. CONCLUSION

lymphangioma circumscriptum, although localised, rarely, extensive and invasive forms are seen. Surgical excision is the main modality of treatment, but the recurrence is common. We report this case, because of its rarity of internal extension and surgical removal as treatment of choice because, if not removed, can serve as a nidus for recurrence. We also highlight the importance of dermoscopy as a non-invasive tool to differentiate it from hemangioma.

CONSENT AND ETHICAL APPROVAL

As per international standard parental written and informed consent of the patient and the ethical approval has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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