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Giant Lymphangioma Circumscriptum: A Differential of Verrucous Lesions

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Abstract: Lymphangioma circumscriptum (aka superficial lymphatic malformations) occurs due to the abnormal migration of lymphatic tissue that leads to failures in the communication and drainage of the lymphatic system. They usually present as groups of translucent papules and vesicles on the skin or mucous membranes. Its presentation as a giant verrucous crusted plaque on proximal arm in a young female of 16 years duration is unusual and emphasizes the relevance of this case report. Diagnosis of this unusual presentation was confirmed with histopathology. This case is presented here for its rarity, to increase its diagnostic awareness and to exemplify the fact that utmost vigilance and awareness of the entity is required to avoid missing diagnosis and to ensure proper treatment.
Keywords: Lymphangioma circumscriptum, Giant verrucous, Lymphatic malformation.

INTRODUCTION

Lymphangiomas are hamartomatous malformations of deep dermal and subcutaneous lymphatic channels. Lymphangioma circumscriptum (LC) is the commonest form of cutaneous lymphangioma in which superficial lymphatic cisterns communicate with skin blisters via thin walled vessels. Typical clinical findings are small fluid-filled vesicles forming plaques on the skin or mucosa. Alternatively, the surface of lymphangioma may appear extremely warty and may be mistaken for fungal infections, tuberculosis or viral warts [1]. Wart like LC is rare and reported most commonly in veneral organs as small lesion. We herein report a case of giant LC located in upper extremity and mimicking a wart.

CASE REPORT

A 22 year old female presented with a long standing history of an asymptomatic lesion on the left proximal arm. Lesion started initially as a reddish eruption following trauma which gradually increased in size to the present size. Patient complained of a foul smelling discharge from the lesion since last 15 days. Clinical examination revealed a well defined giant verrucous plaque with dirty yellow crusting in the left arm measuring around 20x10cm in size. Giant plaque showed distal papulo-nodular growth with sero-sanguineous discharge. Few scattered brownish papules were seen in the perilesional skin (Fig. 1). Because of the giant verrucous nature of the lesion, diagnostic hypothesis included: skin tuberculosis, chromomycosis, sporotrichosis and common wart. Laboratory analysis identified no significant abnormality. Purified protein derivative and viral markers were nonreactive. Tissue culture for tuberculosis and fungal infections proved negative. Culture sensitivity of serosanguineous discharge showed mixed bacterial infection. Since diagnosis remained undefined, biopsy was performed.

Microscopic examination showed proliferation of dilated lymphatic spaces lined by flattened endothelium in the superficial and papillary dermis. Some of the vascular spaces contained clear pink fluid with occasional presence of erythrocytes. The surrounding stroma showed lymphocytic infiltration (Fig. 2, a & b). The overlying epidermis showed hyperkeratosis with focal neutrophilic crust and acanthosis. In accordance with the clinical and pathological findings, a diagnosis of lymphangioma circumscriptum was made and resection of the lesion was advised. Patient refused any surgical intervention and was therefore advised to avoid mechanical trauma and cleanse the local area daily with antiseptic agents. We recommended topical antibiotic ointment application for any ruptured lesion. Patient did not develop any new lesions or any other complication on one year follow up.

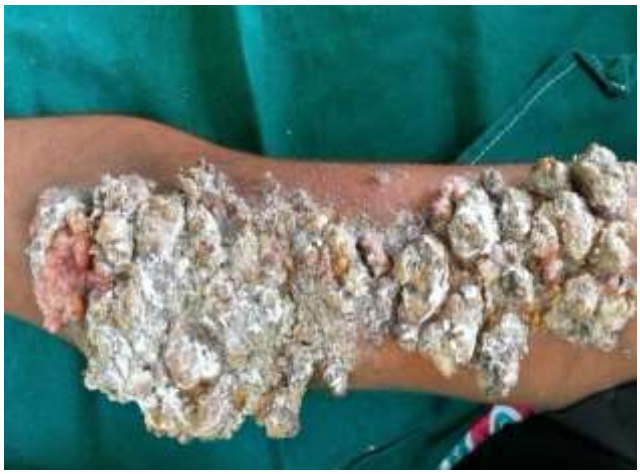

Fig. 1: Giant verrucous plaque with dirty yellow crusting on left arm of the patient

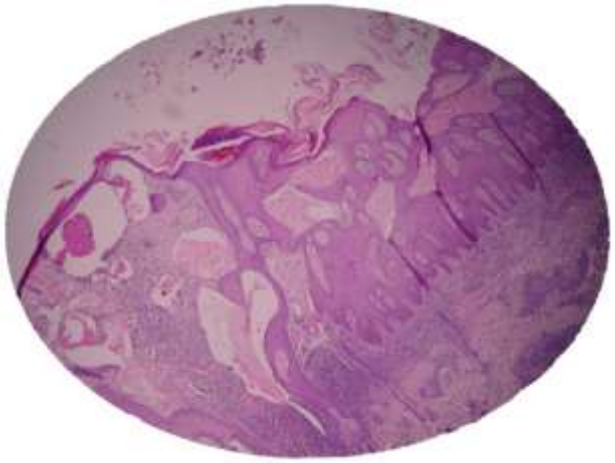

Fig. 2a: Dilated lymphatic spaces in papillary dermis with lymphocytic infiltration in adjacent stroma (Hematoxylin and Eosin, x10)

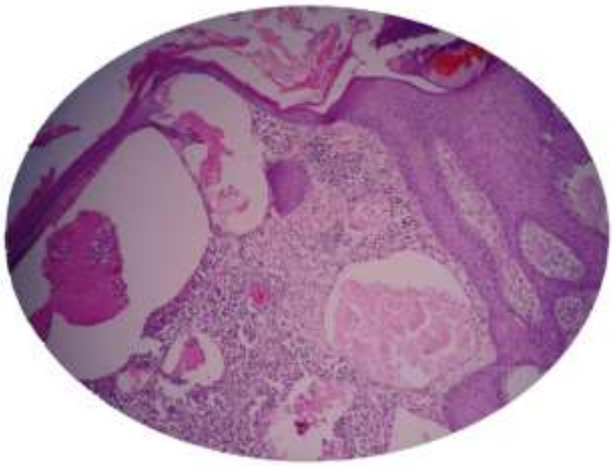

Fig. 2b: Dilated lymphatic spaces in papillary dermis with lymphocytic infiltration in adjacent stroma (Hematoxylin and Eosin, x40)

DISCUSSION

Lymphangiomas are malformations of the lymphatic system [2]. There is benign proliferation of lymph vessels divided into superficial/microcystic (LC) and deep/macrocystic type (lymphangioma cavernosum/cystic hygroma) [3]. LC commonly appears at birth, but may also occur at any age [4]. Clinically, the lesion appears as a cluster of small, cutaneous, translucent vesicles of varying sizes resembling 'frog spawn' or as diffuse swelling in a particular area. These vesicles may develop a pink or red hue upon trauma and subsequent haemorrhage. Although uncommon, these swelling have accompanying verrucous alterations giving them a warty appearance. Microscopically, these vesicles are dilated lymphatic channels that cause papillary dermis to expand. They may be associated with acanthosis and hyperkeratosis. These channels lined by flat endothelial cells are numerous in the upper dermis and often extend to subcutaneous tissue. The deeper vessels tend to have a larger calibre with a thicker wall comprising of smooth muscle. These vessels are filled with lymphatic fluid with occasional presence of erythrocytes [5].

Although exact etiology is unknown, but lymphatic obstruction has been suggested as cause of disease in some reports [6]. Its dermatopathology has been well described by Whimster *et al.* [7] as a cluster of small, thin-walled vesicles communicating with collection of lymphatic cisterns in deep subcutaneous tissue. These deeper cisterns are lined by smooth muscle cells which contract and serve to pump lymph into the superficial vesicles over extended periods of time. Evidence to support this hypothesis has been provided by various lymphangiographic studies that show presence of large multi-lobular cisterns in the dermis and that do not communicate with adjacent normal lymphatics. Diagnosis is based on clinical and histopathological findings.

Though considered a benign lesion, pain, infection and haemorrhage following trauma may occur. Lesion may be single or multifocal, circumscriptive or infiltrative. Lymphangiomas represent a vascular malformation during embryonic development. Vascular endothelial growth receptor (VEGF)-C and VEGF receptor-3 are active in lymphangiomas formation [1]. Differential diagnosis (d/d) includes lymphedema, lymphangiectasis, verrucae, molluscum, hemangioma, tuberculosis or fungal infection [2]. In some cases such as the present one, clinical presentation is so atypical (Giant lesion (20x10cm) on the proximal arm, progression in form of verrucous plaque, late onset), that confirmation by biopsy is necessary.

Main goal of treatment is typically cosmetic and prevention of any complications. In LC, although

dilated vascular channels are present beneath papillary dermis, there are cases with a deep seated component. A deep seated component may cause a high recurrence rate even after excision. Thus, the only radical cure available is to surgically remove the superficial component as well as the deeper lymphatic cisterns. Other treatment modalities used with variable success include electrocautery, cryotherapy, sclerotherapy, laser therapy and observation [8, 9].

In our report, a 20x10 cm giant lesion with verrucous appearance was reported on the proximal arm. Rarely, the surface of the lesion may be warty, but the uncommon feature most commonly is seen in genital organs of adults as small papules and may be misdiagnosed as genital warts or molluscum. The proximal arm may be a common site for LC, but it is an uncommon one for a wart like lesion. LC are generally lesions 1–2 mm in diameter, and some giant ones have been reported, but a 20x10 cm giant lesion is uncommonly large for a LC mimicking a wart [10].

Initial presentation as a slow growing giant verrucous/warty lesion reinforces the relevance of this report in view of rarity of the case and the necessity of correct diagnosis for preventing inappropriate treatment. It also stresses on the fact that lymphangioma should be included in the d/d of verrucous lesion of the extremities.

CONCLUSION

Lymphangioma circumscriptum usually presents as small groups of papules and vesicles on the skin. Rarely does it present as a giant verrucous lesion on the extremities. Knowledge of lymphangioma circumscriptum as a differential of verrucous lesion may help ensure proper treatment for the patients.

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