



## LYMPHANGIOMA CIRCUMSCRIPTUM -A CASE REPORT

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<p><b>Article Info</b> <i>Received 15/12/2015</i> <i>Revised 27/12/2015</i> <i>Accepted 12/02/2016</i></p> <p><b>Key words:</b> Lymphangioma circumscriptum, Lymphorrhoea,</p>	<p><b>ABSTRACT</b> Lymphangioma circumscriptum is a deep dermal and subcutaneous lymphatic channel malformation, which involves the skin, subcutaneous tissue and muscles occasionally. It is often seen at birth or can appear during childhood. The sites which are commonly involved are proximal parts of limbs, shoulders, axillary folds and perineum. It clinically presents as multiple fluid -filled vesicles which may be either discrete or grouped similar to a frog spawn. We report a case of Lymphangioma circumscriptum in a 10 year old girl who presented with complaints of erythematous raised skin lesion over left axilla for the past 7 years.</p>
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### INTRODUCTION

Tilbury Fox and Colcott Fox first described lymphangioma circumscriptum under the name “lymphangiectodes” in the year 1879 and the present term was coined by Malcolm Morris in 1889. It usually presents as multiple fluid filled vesicles (lymph blisters) which bulges over the skin surface. It may be either translucent or red to blue or black in colour. This condition usually presents with recurrent oozing (lymphorrhoea) containing clear fluid in most of the cases.

### CASE REPORT

A 10 year old girl came to our skin OPD with the complaints of reddish raised lesions over the left axilla for the past 7 years. It is gradually progressing in size. She also gives history of similar complaints in the past, for which she underwent a surgery 7 years back. The lesion recurred following the surgery. No history of pain, discharge or itching over the lesion. Dermatological examination revealed multiple grouped erythematous vesicles present in left axillary region. On palpation, it is cystic in consistency. There is no warmth or tenderness and does not bleed on touch. A linear scar of 1cm is seen below the lesion (figure.1). Systemic examination was normal.

Biopsy done and histopathology showed papillary dermis containing solitary and grouped cystic spaces which are lined by endothelial cells in which red blood cells and lymphatic fluid are present. Deeper dermis and subcutaneous fat showed thickened muscular wall with dilated lymphatics – features of Lymphangioma circumscriptum. All baseline routine investigations were done and found to be within normal limits.

**Figure 1. Clinical picture showing multiple erythematous grouped vesicles in the axillary region.**



## DISCUSSION

Lymphangioma circumscriptum (LC) is a hamartomatous malformation of deep dermal and subcutaneous lymphatic channels [1-4]. Synonyms: microcystic lymphatic malformation, capillary lymphangioma, lymphangiectasia, dermal Lymphangioma [5]. It accounts for about 4% of all vascular tumours and 26% of benign vascular tumours in children. It can occur at any age but usually presents at birth or can appear in childhood (5 years). Preachy et al sub divided LC into two groups depending up on the age of onset and extent of involvement – classic and localized variant.

The pathogenesis of LC was stated by Whimster in 1976<sup>(1)</sup>. During embryonal development, a collection of subcutaneous lymphatic cisterns arise which are not continuous with the normal lymph conducting pathways. Dilatation of lymphatic channels occurs as a result of tissue drainage into these abnormal lymphatic channels. When the dermal contractile muscles lining these cisterns contracts, it results in retrograde flow of lymphatics into skin, producing protrusions. Acquired LC occurs in late age are due to infections (filariasis, tuberculosis or lymphogranuloma venereum) and damage due to radiotherapy that results in injury to the deep collecting lymphatic channels. The most commonly involved sites are proximal parts of limbs, shoulders, axillary folds, neck, breasts and perineum. Rarely involves scrotum and oral cavity. Clinically presents as multiple discrete or grouped vesicles which may be either translucent or red to blue or black in colour due to secondary haemorrhage and the surface may be verrucous.

## REFERENCES

1. Saroona Haroon, Sheema H Hasan. (2012). Lymphangioma circumscriptum in the scrotum: a case report. *Journal of medical case reports*, 6, 233.
2. Bikowski JB, Dumont AMG. (2005). Lymphangioma circumscriptum: treatment with hypertonic saline sclerotherapy. *J Am Acad Dermatol*, 53(3), 442-4.
3. Sachdeva S. (2011). Lymphangioma circumscriptum treated with radiofrequency ablation. *Indian J Dermatol*, 56:77-78.
4. Marissa Heller MD, Stephanie Mengden MD. (2008). Lymphangioma circumscriptum. *Dermatology Online Journal*, 14(5), 27
5. Mohan H. Kudur and Manjunath Hulmani. (2013). Extensive and invasive lymphangioma circumscriptum in a young female: A rare case report and review of the literature. *Indian Dermatol Online J*, 4(3), 199–201.
6. Mortimer PS. (2010). Disorders of lymphatic vessels. *Rooks text book dermatology*. Vol 3, Chap 48, 26.

It is often associated with oozing of the lymphatic fluid from the vesicle (lymphorrhoea), pain and pruritis. Complications that can occur are haemorrhage, ulceration, secondary infection and seldom malignant transformation (squamous cell carcinoma). LC can be associated with cystic hygroma and lymphedema of lower limb commonly.

Treatment for a clinically evident LC is indicated when there is cosmetic disfigurement and to prevent from complications such as cellulitis. Treatment of choice is radical excision since inadequate surgical excision results in formation of widespread subcutaneous malformation. Palliative therapy includes Intralesional administration of sclerosants (picibanil, doxycycline or hypertonic saline), vaporization with CO2 laser, argon laser, pulse dye-laser, 900 nm diode laser, electrocautery, radiotherapy, X ray therapy and observation [6,7].

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## CONFLICT OF INTEREST:

The authors declare that they have no conflict of interest.

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