

Lymphangioma Circumscriptum Of The Breast

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Citation

Puneet, S Tiwary, R Khanna, S Singh, A Khanna. *Lymphangioma Circumscriptum Of The Breast*. The Internet Journal of Dermatology. 2005 Volume 3 Number 2.

Abstract

Lymphangioma circumscriptum (LC) is a rare benign condition characterized by the appearance of persistent clusters of thin walled vesicles within a thickened area of skin, which may contain palpable fluctuant cyst (1). The first reported description of LC was done by Fox and Fox in 1878 and they named it as lymphangiectodes (2). The term lymphangioma circumscriptum is first used by Morris in 1889(3). We report here two cases of lymphangioma circumscriptum on the breast.

INTRODUCTION

Lymphangioma Circumscriptum (LC) is usually seen in breast following mastectomy and axillary clearance or radiotherapy (3). Primary LC of breast is very rare. We are reporting two cases of LC of breast without prior history of surgery or radiation therapy, due to its rare presentation.

CASE REPORT 1

40 years female presented with multiple vesicles on the upper part of right breast with recurrent attack of discharge from the vesicles without any fever or pain. A possibility of hidradenitis was kept and the patient was given antibiotics without any relief. The excision was done and the wound was primarily closed. The biopsy came out to be a case of lymphangioma circumscriptum. At follow up of six months there was no recurrence.

CASE REPORT 2

45 years female presented with a progressive increasing vesicles on the right breast and their vesicles increased towards the right axilla (Fig.1). There was discharge from the vesicles with some pain. Examination revealed multiple vesicles with warty appearance. The excision of the lesion was done which revealed it as lymphangioma circumscriptum. On follow up at one year there was no recurrence of the disease.

Figure 1

Figure 1: Right Breast showing multiple vesicles of lymphangioma circumscriptum



DISCUSSION

Lymphangioma circumscriptum (LC) is an uncommon condition of the skin and subcutaneous tissue characterized

by the presence of clusters of thin walled vesicles filled with the clear fluid but occasionally contains discolored fluid by the presence of altered blood, over a slightly thickened area of subcutaneous tissue which may contains palpable fluctuant cyst (1).

The classic lesion of LC appears at birth or in early childhood. However they may arise spontaneously in later life although rarely after fourth decade (4). It is comparatively more common in females (5). The etiology is not fully understood, but the majority of cases are thought to represent congenital abnormality, possibly resulting from sequestered island of lymphatic tissue that develops without any connection to normal lymphatic channel (1). Trauma including surgery and radiation therapy has been implicated in some cases as the causative agent (4). It can occur spontaneously in a congenitally lymphedematous extremity without prior surgery or radiation therapy (4). It can occur at any location including breast, thigh, buttocks neck and axilla. These are multiple lesions but are generally localized to one specific area (4). When vesicles are confined to an area less than 7 cm in diameter it is referred to as lymphangioma circumscriptum. When size is greater than 7 cm it is termed as lymphangioma diffusum (1).

The abnormal vessels of lymphangioma circumscriptum represents sequestered lymphatics, which consist of collection of muscle coated cisterns lying deep in the subcutaneous plane and communicating via dilated dermal lymphatics with superficial vesicles (1). The diagnosis is clinical with histological confirmation. Imaging is valuable in delineating the depth of penetration and the border of the lesions and its relationship to vital structures. The natural course of these lesions is slow progression of the lesion both in size and area with leakage of fluid, infection, local discomfort from hypertrophy and irritation from clothing and movement.

Surgery is the most appropriate treatment of LC. The excision includes deep lymphatic cisterns with wide subcutaneous tissue followed by primary skin closure or skin grafting. It is postulated that by separation of the cisterns from the vesicles causes involution and regression. Data indicates that lesions smaller than 7cm respond well to surgery while larger than 7 cm do not (5). The other modality of treatment available includes radiotherapy, electrocautery, carbon dioxide laser therapy and topical dressings (5,6). In our both cases excision with primary closure could be achieved without recurrence.

In conclusion breast LC without prior surgery or radiotherapy is very rare. They represent the congenital abnormality of lymphatic vessels that may present later. Surgery is the treatment of choice.

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