Lymphangioma Circumscriptum Of The Breast

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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 (2). 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.

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 (\(4\)). The other modality
of treatment available includes radiotherapy, electrocautery,
carbon dioxide laser therapy and topical dressings (\(5\)). 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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