■ *Case Report*

Lymphangioma circumscriptum - rare site

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Abstract

22 year old male came to Skin department with grouped vesicles over sternal region since birth along with serous discharge. The biopsy taken from site showed histopathologically Lymphangioma circumscriptum. It was treated with CO2 Laser, and response was good.

Keywords: Lymphangioma circumscriptum, CO2 laser,

Introduction

Lymphangioma circumscriptum (LC) is a term used for lymphatic malformation which is seen in skin, subcutaneous tissue and sometimes muscle.[1] Clinically the condition manifests with fluid filled vesicles seen on the skin. The vesicles may be well defined and discrete, or may be grouped into structures resembling frogspawn. The lymphangiomas may be transluscent when overlying epidermis is very thin, or they may vary in colour from red to blue black when they contain blood. Lymphangioma circumscriptum may present at any age but usually seen at birth or appears during childhood. The commonest sites are the axillary folds, shoulders, flanks, proximal parts of limb and perineum. Lymph weeping (lymphorrhoea) from one or more surface vesicles is common and is likely to increase the risk of infection. Vaporization with carbon dioxide laser can be of palliative benefit[2] and superficial x rays have been successfully used.

Case report

A 22 year old male came to Skin opd with grouped vesicles over chest (Fig 1) since birth along with serous discharge since 2 months. There was no history of pain or pruritus. There was also a tender, oval, swelling of size 3cm x 4cm present over the sternal region since 2 months. The FNAC of swelling reveled hematoma. The biopsy taken from skin lesions showed histopathologically Lymphangioma circumscriptum (Fig 2). His hemoglobin, blood counts, ESR, USG chest within normal limits . It was treated with CO2 Laser, under antibiotic coverage in view of hematoma already present. The patient was reviewed after 3 weeks when all lesions had healed up.

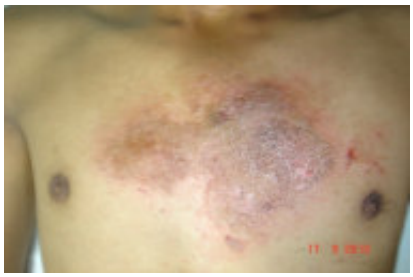

Fig: 1 – Grouped vesicles over the chest

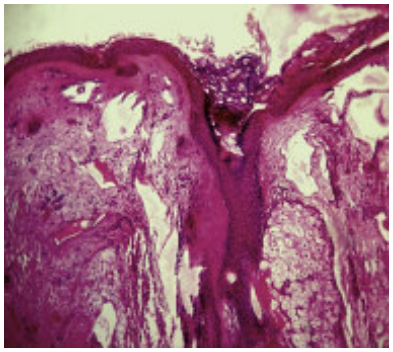

Fig: 2 -Dilated lymph vessels in dermis with acanthosis and hyperkeratosis of epidermis

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Discussion

A lymphangioma or lymphatic malformation represents a congenital proliferation of lymphatic vessels. These conditions account for approximately 4 percent of vascular tumors and 26 percent of benign vascular tumors in children. The predominant types of lymphangiomas are lymphangioma circumscriptum, cavernous lymphangioma, cystic hygroma, and benign lymphangioendothelioma.[3]

The most common form of cutaneous lymphangioma is lymphangioma circumscriptum, which arises in infancy but may occur at any age. They are caused by abnormal dilations of lymph vessels. Most commonly, lymphangioma circumscriptum is found on the proximal extremities, trunk, axilla, and oral cavity but also may occur on the penis, vulva, and scrotum.[4, 5] .Lymphangioma circumscriptum is asymptomatic; however, it can be complicated by excessive drainage and recurrent cellulitis.

The pathogenesis of lymphangioma circumscriptum was first described by Whimster in 1976. He postulated that lymphatic cisterns in the deep subcutaneous tissue arise from early lymph tissue sacs during embryonic development. Furthermore, he noted that these sacs were distinct entities that did not connect to the remainder of the lymphatic system. Finally, he proposed that these sacs are lined by muscle fibers that contract and induce pressure that causes outpouchings in the walls of the sacs. These outpouchings eventually protrude from the skin surface to become what are clinically described as vesicles.[6]

Histolopathologic features of lymphangioma circumscriptum include dilated lymph vessels in the upper dermis that may extend into the subcutis. These dilated vessels cause expansion of the papillary dermis. There may be acanthosis and hyperkeratosis of the overlying epidermis.[7] The lymph fluid within the vesicles may contain erythrocytes, lymphocytes, macrophages, or neutrophils.

Indications for treatment of lymphangioma circumscriptum include both its cosmetic appearance and prevention of complications such as cellulitis. The definitive treatment for lymphangiomas is surgical excision of both the superficial and deep components.[8] However, the recurrence rates can be high, especially in the case of lesions with deeper components. Other palliative treatments that have been reported include superficial x-ray therapy, radiotherapy, argon laser, CO2 laser, 900-nm diode laser, pulsed dye-laser, and sclerotherapy.[9 10]

This case is being presented due to the occurrence in rare site as well as good response to one sitting of CO2 laser treatment.

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