

Case Report

Lymphangioma Circumscriptum - A Rare Case

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ABSTRACT

Lymphatic malformations or lymphangiomas are benign proliferations of lymphatic vessels. They account for approximately 4% of all vascular tumours and 26% of benign vascular tumors. Lymphangioma are classified into a superficial lymphangioma circumscriptum and a deeper cavernous lymphangioma. Here we present a case of 35 year old female with firm keratinizing lesions in bunches on anterior aspect left thigh. Provisional diagnosis of lymphangioma circumscripta was made. Lesion excised up to deep fascia and primary closure done. Biopsy confirmed our diagnosis.

Key Words: Excision, Frog spawn, Hamartoma, Superficial lymphatics.

INTRODUCTION

Lymphangioma circumscriptum (LC) is an uncommon hamartomatous malformation of superficial lymphatics present as translucent, discrete or grouped vesicles resembling frog spawn appearance which contains clear fluid. [1] Primary lymphangioma circumscriptum usually present at birth or develops in early childhood. Secondary LC induced by impairment of lymph flow. Most common sites of LC are the proximal extremities, trunk, axilla and oral cavity but also occurs on the penis, scrotum and rarely in the vulva. [2,3] Diagnosis is mainly by clinical features and radiological modalities supplement it. Treatment in most of the cases is surgical excision.

CASE REPORT

A 35 year old female patient came to our outpatient department with multiple lesions over the anterior aspect of left thigh since last 10 years. The lesions gradually increased in size and number. It is

associated with history of intermittent discharge from the lesions which is clear in nature. No history of sudden increase in the size of lesions. Not associated with pain and fever. No history of similar lesions in the past and anywhere else on the body. No history of similar complaints within the family.

On examination, multiple, firm keratinizing lesions in bunches and clusters are found on anterior aspect of left thigh. (Figure 1) Skin around the vesicles showed dark pigmentation. No active discharge is observed at the time of examination. No palpable lymphadenopathy on bilateral limbs. External genitalia and other limb are completely normal. Routine laboratory investigations were done and found within normal limits. Based on clinical features, a provisional diagnosis of Lymphangioma circumscripta is made. Full thickness of the skin along with the lesion is excised, (Figure 2) primary closure done and a sub cutaneous corrugated rubber drain is placed.



Figure 1: Lesion over anterior Aspect of left thigh



Figure 2: Full thickness of the skin with lesion excised



Figure 3: Dilated Lymph channels In Upper dermis



Figure 4: Thinned out Epidermis

Histopathological examination of the lesion revealed dilated Lymph vessels containing lymphatic fluid lined by single layer of epithelial cells in papillary dermis along with thinned out epidermis. Thus our diagnosis of lymphangioma circumscripta is confirmed. (Figure 3 and 4)

DISCUSSION

Lymphatic malformations are broadly classified into superficial lymphangioma circumscripta and deeper cavernous lymphangioma. Another type of congenital malformation, the cystic hygroma is considered as a variant of cavernous lymphangioma. Lymphangioma circumscripta was first discovered by Fox and Fox in 1879 under the name lymphangiectodes and the present term was given by Morris in 1889. Lymphangiomas are nothing but the damaged portions of

lymphatic system which can promote the production of lymph and increase in volume of lymph inside it leads to larger growth in the contiguous tissue. [4,5]

Preach *et al* divided lymphangioma circumscripta into two groups, classical and localized. Classical LC differs from the localized variety by age of onset and extent of involvement. Common sites involved are neck, axilla, breast, chest and buttocks. Lesions have been described in the vulva, peri-anal region and are hence mistaken for ano-genital warts. In the neck, they are quite common in posterior triangle of the neck and they are stereotypically soft and fluctuant. They are less common in anterior triangle, however when they become large they are likely to cause dysphagia and respiratory distress. [6,7]

Lymphangioma circumscripta is characterised by presence of vesicles which

are either discrete or grouped resembling frog eggs. Vesicles contain clear fluid but occasionally the color may vary from red to blue black due to presence of blood because of secondary hemorrhage from surrounding tissue. In some cases, the surface may be warty due to hyperkeratosis. Increased girth of limb reported in some cases of LC especially in superficial vesicles which have associated deeper component. Lymphangioma of the anterior aspect of tongue cause macroglossia. [8,9] Some patients having LC may have distressing symptoms in form of pain, pruritus, and exudation. Rare complications include infection, hemorrhage and malignant transformation.

Lymphangiography and ultrasonography are helpful in diagnosis; MRI has been used for evaluating the hidden deep component of lymphangioma circumscripta. Histopathological appearance of lymphangioma circumscripta is characterized by solitary and grouped dilated cystic spaces in the papillary dermis. Cystic spaces are lined by endothelial cells. In deeper dermis and sub cutaneous fat, dilated lymphatics with thickened muscular walls are seen. It has been postulated that muscular lymphatics in the deep dermis and subcutaneous tissue are aberrant in nature and not in continuity with the normal lymph conducting pathway. Retrograde transmission of pressure from these lymphatics causes dilation of superficial lymphatics resulting in formation of the vesicle.

The main indicator for therapy in LC is cosmetic. If the disease is asymptomatic, it is better to follow "wait and watch" approach. Among the various modalities for the treatment, surgical excision remains the treatment of choice especially for the lesions confined to superficial dermis. Various other treatment modalities include X-rays, radiotherapy, cryotherapy, sclerotherapy, cautery, argon laser, CO2 laser, pulse Dye laser. [10,11] Prognosis is good in most of the patients except in cases of large tumors obstructing the airway. A few cases of

squamous cell carcinoma arising from lymphangioma circumscriptum have been reported. [12]

CONCLUSION

Lymphangioma Circumscriptum is a rare proliferative disorder of lymphatic vessels. Surgical treatment remains the mainstay of treatment, although recurrence rates are high.

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