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Lymphangioma circumscriptum: clinicopathological spectrum of 29 cases

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INTRODUCTION

Lymphangiomas are malformations of lymphatic channels which may be localized or generalized, congenital or acquired. It is argued that lymphangiomas are true neoplasms and occur due to transformation of endothelial cells or stromal cells.\(^1\) They account for 4% of vascular tumors in children, about 50% presenting at birth without any familial history.\(^2,3\) They arise anywhere on skin, subcutaneous tissue or mucous membranes. The common sites include head and neck (including tongue), proximal extremities, trunk and buttocks. Abdominal viscera such as intestine, pancreas and mesentery as well as heart can be involved rarely. The classical types are: (a) Lymphangioma simplex (b) lymphangioma circumscriptum (c) cavernous lymphangioma and (d) cystic hygroma. Now they are broadly grouped into superficial and deep depending upon depth and size of lymphatic channels.

Lymphangioma Circumscriptum (LC) is placed in the first group while the second group comprises of cavernous lymphangioma and cystic hygroma.

The term lymphangioma circumscriptum was first coined by Morris et al. in 1889, presenting as multiple clusters of clear, pink or red vesicles which could be associated with bleeding or oozing of clear fluid, super infection with \textit{Staphylococcus aureus}, recurrent cellulites and lymphedema of the limb. The latter may cause subcutaneous thickening or cyst formation. Rarely association with Mafucci, Proteus and Cobb syndromes has also been reported.\(^4-6\) The usual presentation is at birth or early childhood but lesions can occur at any age. It has been reported to occur following radical hysterectomy and radiation therapy.\(^7\) Ultrastructure studies have shown differences between superficial and deep channels. The endothelial cells in superficial channels are more electron dense, flattened having partially multilayered basal lamina. In deeper channels cells are less electron dense, less flattened and have discontinuous basal lamina. These changes hint towards the fact that only superficial channels have abnormality.\(^8\)

The literature reveals only case reports of Lymphangioma Circumscriptum (LC) from Asian countries including Pakistan. There has been no larger epidemiological study from our country.

The aim of this study was to present the epidemiology and clinicopathological characteristics of cases diagnosed as lymphangioma circumscriptum.

ORIGINAL ARTICLE

**Lymphangioma Circumscriptum: Clinicopathological Spectrum of 29 Cases**

Saira Fatima, Nasir Uddin, Romana Idrees, Khurram Minhas, Zubair Ahmad, Rashida Ahmad, Naila Kayani and Muhammad Arif

**ABSTRACT**

**Objective:** To describe the clinicopathological spectrum of Lymphangioma Circumscriptum (LC).

**Study Design:** Observational case series.

**Place and Duration of Study:** Department of Pathology and Microbiology, AKUH, Karachi, from 2002 to 2012.

**Methodology:** All reported cases of LC were retrieved from medical record. Clinical and pathological features were noted. Frequency percentages were determined.

**Results:** There were 29 cases of LC predominantly males (62%). The mean age was 27.17 ± 15.5 years. The commonest sites was anal/perianal region (24%) followed by extremities (17%) and tongue, (14%). Vulval LC was seen in 3 patients. Two cases were described on scrotum. The lesions were most commonly suspected as viral warts, mole or polyp (in anal region). Vesicles with erosions and bleeding and localized growth were the usual clinical presentations. Four of the patients presented with swelling since birth. All were treated with surgical excision. Microscopic examination revealed acanthotic squamous epithelium with papillomatosis. The subepithelial region had collections of lymphatic channels composed of ectatic dilated vessels with serum and inflammatory cells in their lumina. The lymphatic channels were seen in deeper layers along with lymphocytic aggregates.

**Conclusion:** Lymphangioma circumscriptum is a malformation of abnormal lymphatic channels with feeding cisterns in subcutaneous tissue. It is a benign lesion usually occurring in anal/perianal region and confused with warts. Surgical excision is preferred mode of treatment.

**Key Words:** Cutaneous lymphangioma circumscriptum. Warts. Lymphatic malformation.

**INTRODUCTION**

Lymphangiomas are malformations of lymphatic channels which may be localized or generalized, congenital or acquired. It is argued that lymphangiomas are true neoplasms and occur due to transformation of endothelial cells or stromal cells.\(^1\) They account for 4% of vascular tumors in children, about 50% presenting at birth without any familial history.\(^2,3\) They arise anywhere on skin, subcutaneous tissue or mucous membranes. The common sites include head and neck (including tongue), proximal extremities, trunk and buttocks. Abdominal viscera such as intestine, pancreas and mesentery as well as heart can be involved rarely. The classical types are: (a) Lymphangioma simplex (b) lymphangioma circumscriptum (c) cavernous lymphangioma and (d) cystic hygroma. Now they are broadly grouped into superficial and deep depending upon depth and size of lymphatic channels. Lymphangioma Circumscriptum (LC) is placed in the first group while the second group comprises of cavernous lymphangioma and cystic hygroma.

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METHODOLOGY

A retrospective study was conducted and all the cases diagnosed as lymphangioma circumscriptum were retrieved from the surgical pathology files. There were 29 cases over a period of 13 years. Patient’s age, gender, site and size were recorded. Clinical presentation and physicians’ preliminary diagnosis were also noted. Microscopic characteristics were described in detail.

The data was double entered by two data entry operators in EpiData (version 3.2) and data entry errors were removed. The clean data then was converted into SPSS (version 19.0). Mean and standard deviation values were calculated for numerical variables i.e. age and tumor size of the patients. Frequencies and percentages were calculated for categorical variables i.e. gender and site.

RESULTS

The patients’ mean age was 27.17 ± 15.5 years. Minimum age was 4 years while maximum age was 66 years. There were 18 (62%) males and 11 (38%) females. The commonest sites in this study was anal/perianal region 7 (24%) followed by extremities 5 (17%) and tongue 4 (14%). Vulval LC was seen in 3 (10%) patients. Two (7%) cases each were described on scrotum, trunk and head and neck area. Site was not mentioned in 4 (14%) cases. The lesions were most commonly suspected as viral warts, mole or polyp in anal region. In one case, the clinical diagnosis was melanoma. Vesicles with erosions and bleeding and localized growth were the usual clinical presentations. Four patients presented with congenital swelling. Table I summarizes the cases according to their anatomical distribution, gender, age and size. In the region of anal canal, age at presentation was higher in males. Therefore, tumor size was also greater in males (2.68 ± 2.5 cm) as compared to females (2.20 ± 0.2 cm). In tongue, size was greater in males but in extremities tumors were larger in females.

Microscopic examination revealed acanthotic squamous epithelium with papillomatosis imparting warty appearance (Figure 1). The sub-epithelial areas had

<table>
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<th>Site</th>
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<th>Age SD</th>
<th>Size Mean</th>
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Figure 1: Acanthosis, papillomatosis and lymphatic channels in papillary dermis.

Figure 2: Markedly dilated lymphatic channels in papillary dermis showing lymphocytes and serum.
collections of lymphatic channels composed of ectatic dilated vessels with serum and inflammatory cells in their lumina (Figure 2). In areas where the channels were in close proximity with the epidermis, the lining had thinned out. The lymphatic channels were predominantly present in papillary dermis with some dilated spaces in deeper layers having larger caliber and thicker smooth muscle in their wall, extending up to the painted deep margin. Lymphocytic aggregates were present in the stroma.

**DISCUSSION**

The pathogenesis of lymphangioma circumscriptum was first described by Whimster in 1976.9,10 It was postulated that large muscle coated cisterns were present in subcutaneous tissue, disconnected from the normal network of lymphatic vessels. However, they are connected to dermal lymph channels. The vesicles result from the dilatation of lymphatics in papillary dermis caused by back pressure. Based on this pathogenesis, it was suggested that excision of subcutaneous cisterns while leaving overlying skin might offer a better cosmetic and acceptable option to the patient.

The endothelial cells of these channels stain with Ulex Europaeus. The markers which differentiate between vascular and lymphatic channels include D-240, Prox-1,11 vascular endothelial growth factor C and vascular endothelial growth factor 3.12 The latter two growth factors play important role in embryogenesis and increased lymphangiogenesis.

The reported cases usually presented at birth or early childhood, while in our study wide age range of presentation was seen. Four patients had congenital swellings, 3 on extremities and 1 on chest. Peachy et al. have further categorized cutaneous lymphangiomas into two classic and localized types differing in site, size, histological characteristics and symptoms.13 The classic form occurs at or shortly after birth, exceeding 1 cm² in size. Their sites of predilection being proximal extremities, axilla and chest. Localized form are less than 1 cm², affect any age group and any part of the body. In this study 4 (14%) cases presented at birth involving limbs and adjacent girdles, qualifying as classic lymphangioma circumscriptum.

In this study, perianal/anal region was the commonest site of lymphangioma circumscriptum, most of them having polypoidal and warty appearance. Bleeding PR was additional complaint in 3 cases. Tumours were larger (mean size 2.68 cm) in males so was the higher size. Their sites of predilection were in close proximity with the epidermis, the lining had thinned out. The lymphatic channels were predominantly present in papillary dermis with some dilated spaces in deeper layers having larger caliber and thicker smooth muscle in their wall, extending up to the painted deep margin. Lymphocytic aggregates were present in the stroma.

The treatment options include surgical excision, sclerotherapy, electrocoagulation, liquid nitrogen therapy and carbon dioxide laser therapy.18 The most preferred treatment for lymphangioma circumscriptum is surgical excision with complete removal of deep feeding cisterns thus, reducing the chance of recurrence.19 Lesions measuring greater than 7 cm have higher risk of recurrence.

**CONCLUSION**

Lymphangioma circumscriptum is a malformation of abnormal lymphatic channels with feeding cisterns in subcutaneous tissue. It is a benign lesion found mostly in anal/perianal region of males in the present study.

**REFERENCES**


