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.

Department of Pathology and Microbiology, The Aga Khan University Hospital, Karachi.

Correspondence: Dr. Saira Fatima, B-4, Jumani Arcade, Gulshan-e-Iqbal # 14, Karachi.
E-mail: saira.fatima@aku.edu

Received: December 24, 2012; Accepted: August 07, 2015.

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 *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 clinicalpathological characteristics of cases diagnosed as lymphangioma circumscriptum.

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 I: Clinical summary of the cases with statistical analysis.

Site	Gender		n	Mean	SD
Anal canal	Male	Age	5	45.60	15.85
n=7 (24%)		Size	5	2.68	2.562
	Female	Age	2	32.50	10.60
		Size	2	2.20	0.282
Extremities	Male	Age	4	17	6.32
n=5 (17%)		Size	4	3.90	2.22261
	Female	Age	1	20.00	1
		Size	1	8	1
Tongue	Male	Age	2	16.50	14.849
n=4 (14%)		Size	2	1.1000	.14142
	Female	Age	2	17.50	3.536
		Size	2	0.90	0.848
Site unavailable	Male	Age	1	47	---
n=4 (14%)		Size	1	3	---
	Female	Age	3	14.67	4.933
		Size	3	7.33	2.88
Vulva	Male	Age	-	-	-
n=3 (10%)		Size	-	-	-
	Female	Age	3	33.33	8.386
		Size	3	4.1	5.15
Scrotum	Male	Age	2	36	8.485
n=2 (7%)		Size	2	2.2	0.424
	Female	Age	-	-	-
		Size	-	-	-
Chest	Male	Age	2	9.00	7.071
n=2 (7%)		Size	2	3.7500	1.76777
	Female	Age	-	-	-
		Size	-	-	-
Head and neck	Male	Age	2	29	22.6
n=2 (7%)		Size	2	2.55	1.484
	Female	Age	-	-	-
		Size	-	-	-

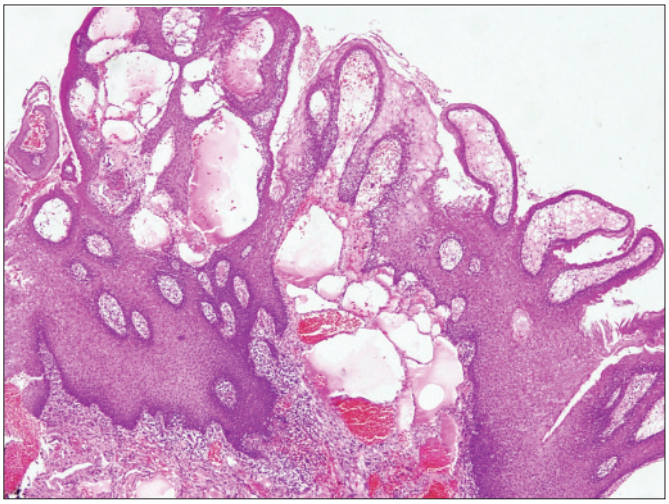

Figure 1: Acanthosis, papillomatosis and lymphatic channels in papillary dermis.

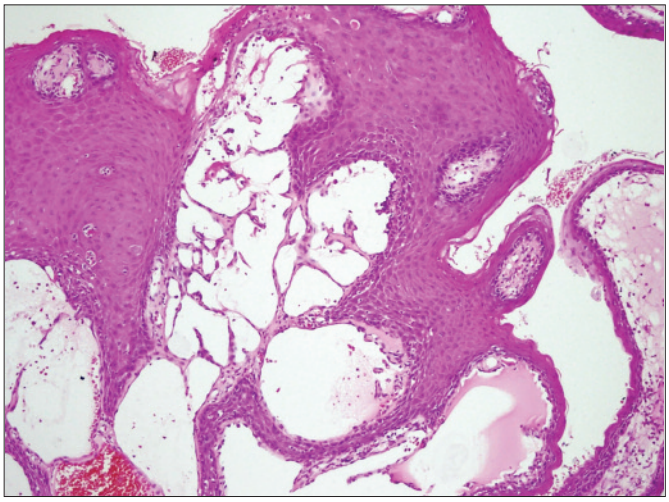

Figure 2: Markedly dilated lymphatic channels in papillary dermis showing lymphocytes and serum.

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 cm^2 in size. Their sites of predilection being proximal extremities, axilla and chest. Localized form are less than 1 cm^2, 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 age of presentation (mean age 45.60)

There were 3 cases of vulval lymphangiomas presenting through 3rd to 5th decades, mistaken as vulval warts. Only one of these lesions measured less than 1 cm in maximum dimension. There had been no prior history of surgery or radiation. Roy *et al*. described 11 cases of congenital LC of vulva.[13] The vulval lesions may clinically be misdiagnosed as genital warts, molluscum contagiosum or tuberculosis verrucosa cutis, hence, histological examination is mandatory for such lesions.[14]

Obstruction of lymphatic channels may be caused by inflammatory or infectious pathology e.g. tuberculosis, filariasis, lymphogranuloma venereum etc. Lymph nodes involved by a malignancy (primary or secondary) may obstruct the lymphatics by external pressure. These conditions are the underlying causes of localized or acquired lymphangioma circumscriptum. The cases in vulval and scrotal regions did not have positive history of tuberculosis in our study.

The complications of lymphangioma circumscriptum depending upon the location may include obstruction of airways and esophagus in case of oral lesions, secondary infections, cellulites and haemorrhage. Psychosexual abnormalities may develop in patients with genital lesions. A rare complication is development of lymphangiosarcoma occurring in cases where LC occurred at sites of previous radiotherapy.[15] A case of squamous cell carcinoma arising in lymphangioma circumscriptum has been reported.[16] Another tumor, Debska tumor (Papillary intralymphatic angioendothelioma) has also been described.[17]

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

1. Shetty DC, Urs AB, Rai HC, Ahuja N, Manchanda A. Case series on vascular malformation and their review with regard to terminology and categorization. *Contemp Clin Dent* 2010; **1**: 259-62.
2. Gigure CM, Bauman NM, Smith RJ. New treatment options for lymphangioma in infants and children. *Ann Otol Rhinol Laryngol* 2002; **111**:1066-75.
3. Gopal AP, Robertez AS. Cutaneous lymphangioma circumscriptum: frog spawn on the skin. *Int J Dermatol* 2009; **48**:1290-5.
4. Masood T, Al Hussain, Haroon TS. Proteus syndrome: a case report and review of literature. *J Pak Assoc Derm* 2000; **10**:64-9.
5. Child FJ, Werring DJ, Vivier AW. Proteus syndrome: diagnosis in adulthood. *Br J Dermatol* 1998; **139**:132-6.

6. Shim JH, Lee DW, Cho BK. A case of Cobb syndrome associated with lymphangioma circumscriptum. *Dermatology* 1996; **193**:45-7.

7. Schwab RA, Mc Collaough ML. Acquired vulval lymphangiomas: a sequel of radiation therapy. *Cutis* 2001; **67**:239-40.

8. Asano S, Endo H, Sagami S. An ultra-structural study of localized lymphangioma circumscriptum. *Arch Dermatol Res* 1978; **262**:301-9.

9. Narang T, Dipankar D, Dogra S. Lymphangioma circumscriptum and Whimster's hypothesis revisited. *Skinmed* 2011; **9**:123-4.

10. Whimster IW. The pathology of lymphangioma circumscriptum. *Br J Dermatol* 1976; **94**:473-86.

11. Sultan A, Dadras SS, Bay JM, Teng NNH. Prox-1, podoplanin and HPV staining assists in identification of lymphangioma circumscriptum of the vulva and discrimination from vulval warts. *Histopathology* 2011; **59**:1261-79.

12. Peachy RD, Lim CC, Whimster IW. Lymphangioma of skin: a review of 65 cases. *Br J Dermatol* 1970; **83**:516-27.

13. Roy KK, Agarwal R, Agarwal S, Kumar S, Malhotra N, Gopendru N. Recurrent vulvar congenital lymphangioma circumscriptum: a case report and literature review. *Int J Gynecol Cancer* 2006; **16**:884-943.

14. Aggarwal K, Gupta S, Kumar JV, Marwah N. Congenital lymphangioma circumscriptum of the vulva. *Indian Pediatr* 2009; **46**:428-9.

15. Amouri M, Masmoudi A, Boudaya S, Amouri A, Ben Ali I, Bouassida S. Acquired lymphangioma circumscriptum of the vulva. *Dermatol Online J* 2007; **13**:10.

16. Wilson GR, Cox NH, Mc Lean NR, Scott D. Squamous cell carcinoma arising within congenital lymphangioma circumscriptum. *Br J Dermatol* 1993; **129**:337.

17. Emanuel PO, Lin R, Silver L, Birge MB, Shim H, Phelps RG. Dabska tumor arising in lymphangioma circumscriptum. *J Cutan Pathol* 2008; **35**:65-9.

18. Dikmen Y, Terek MC, Akman L, Solmaz U, Zekioglu O, Akalin T. Congenital lymphangioma circumscriptum. *Int J Clin Oncol* 2008; **13**: 458-60.

19. Cavitcol, Hasdemir O, Yalcin E, Buyukasik O. Lymphangioma circumscriptum of the rectum: a case report and the review of literature. *J Chin Clin Med* 2008; **3**:159-61.