

Lymphangioma circumscriptum of the vulva: Report of a rare case

Anuradha Sinha, Jyoti Prakash Phukan, Shilpa Jalan, Subrata Pal

Department of Pathology, Bankura Sammilani Medical College, Bankura, West Bengal, India

ABSTRACT

Lymphangioma circumscriptum (LC) is a form of lymphangioma characterised by benign dilation of lymphatic channels, which affects the skin and subcutaneous tissues. The most common sites of LC are mucosa of mouth, tongue, groin, axilla, trunk and proximal region of extremities. Vulva is a rare site of LC. In this report, we are presenting a case of LC of vulva occurring in a 60-year-old female without any obvious reason. The patient presented with multiple genital wart-like papular lesions in the vulva. Biopsy of lesion reveals LC. She was treated with vulvectomy and showed no sign of recurrence till date.

Key Words: Lymphangioma, lymphangioma circumscriptum, vulva

INTRODUCTION

Lymphangioma circumscriptum (LC) is a benign lymphatic malformation characterized by dilation of lymphatic vessels in the skin and subcutaneous tissue.^[1] These abnormal lymphatic malformations do not communicate to the normal lymphatics.^[2] The exact cause of LC is unknown. It can be congenital or acquired due to damage of lymphatic vessels of various etiologies. LC is commonly found in chest, mouth, axilla and tongue.^[1] Vulva is an uncommon site for LC. Till date, only a few cases of LC of vulva have been reported in the literature. We intend to report a case of LC of vulva in a 60-year-old female presented with multiple warty papular lesions of vulva.

CASE REPORT

A 60-year-old woman attended to obstetrics and gynecology outpatient department of our medical college with multiple

asymptomatic wart-like lesions over vulva and mons pubis. There was no history of burning, pruritus or associated pain. According to the patient, the lesions had developed since 4-5 years. There was no history of pelvic surgery, prior chemotherapy, gastrointestinal disease or tuberculosis. On examination, multiple warty lesions are seen ranging from 3-10 mm in size in the vulva and mons pubis. Abdominal and pelvic ultrasonography (USG) were done and were normal. Routine blood examination was normal except mild leucocytosis. With a clinical differential diagnosis of wart, molluscum contagiosum and lymphangioma, biopsy was taken from lesion and sent to us for histopathological evaluation.

Histopathological examination revealed acanthosis and hyperkeratosis of epidermis with numerous dilated lymphatic channels containing eosinophilic proteinous material in the epidermis and papillary dermis [Figure 1]. Thus a diagnosis of LC was confirmed. Patient underwent surgical procedure with removal of vulva and involved area with 3 cm in depth. After surgery, the sample was sent to pathology department for further examination [Figure 2].

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Sinha A, Phukan JP, Jalan S, Pal S. Lymphangioma circumscriptum of the vulva: Report of a rare case. J Mid-life Health 2015;6:91-3.

Address for Correspondence: Dr. Jyoti Prakash Phukan, Department of Pathology, Bankura Sammilani Medical College, Kenduadihi, Bankura - 722 102, West Bengal, India.
E-mail: drjyotiphukan@yahoo.co.in

Access this article online

Quick Response Code:



Website:
www.jmidlifehealth.org

DOI:
10.4103/0976-7800.158968

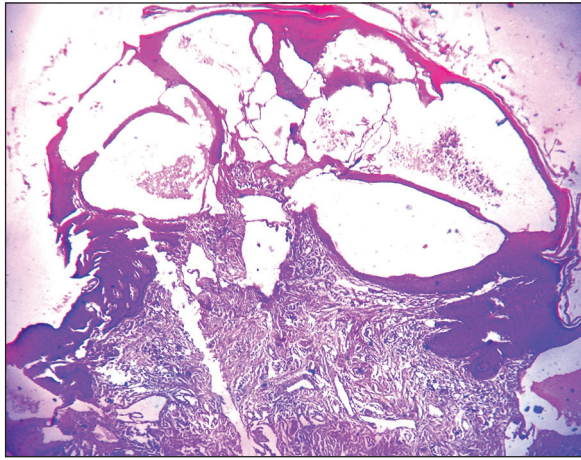


Figure 1: Histopathological section of the lesion showing typical features of lymphangioma circumscriptum with acanthosis and hyperkeratosis of the epidermis along with numerous dilated lymphatic channels in the epidermis and papillary dermis containing eosinophilic proteinaceous material. [H and E, × 100]

Histopathological examination once again confirmed the diagnosis of LC and the resected margins were clear. There was no evidence of malignancy. The patient was followed up for 1 year and there was no sign of recurrence during this period.

DISCUSSION

Lymphangiomas are congenital malformations of the lymphatic system that usually involve skin and subcutaneous tissue.^[3] Lymphangioma can occur at any age but majority are seen in children. They can be divided into superficial (Lymphangioma circumscriptum), deep (Lymphangioma cavernosum) and cystic (Cystic hygroma) varieties. LC is a distinct form of lymphangioma that is localized to the skin and subcutaneous tissue. It was first described by Fox and Fox in 1879 as lymphangiectodes and later on renamed to lymphangioma circumscriptum by Morris in 1889.^[3] Clinically it manifests as pseudovesicles, which bluge on the skin and hyperkeratosis may give rise to warty appearance.^[4] They contain a clear, yellow or pink coloured fluid depending on the extent of blood content. Histology of LC shows dilated lymphatics in the epidermis and the papillary dermis. They are lined by flat endothelial cells and the dilated lymphatics contain red and white blood cells.^[2]

The exact aetiology of LC is unknown. However various growth factors such as vascular endothelial growth factor-C (VEGF-C) and VEGF-D and their receptors on the lymphatic endothelial cells may have a role in the mechanisms that controls the development of LC.^[5] Various classification systems are used to classify LC, out of which the classification made by Peachey *et al.*, is widely used.^[6] He divided LC into two main forms: Classic and localized.



Figure 2: Gross resected specimen showing multiple warty popular lesions in the vulva

The classic form usually appears at or soon after birth and usually involves proximal limbs. The classic form of LC is thought to derive from muscular lymphatic cisterns which have failed to segment during embryonic development.^[7] They usually appears as vesicular and do not progress into warty plaque. On the contrary, localized form is seen in any age and has no site predilection. Again, LC can be divided into congenital and acquired form. Congenital LC stems from local malformation of lymphatics and manifest at birth or before 5 years of age.^[8] Acquired forms of LC occurs secondarily due to obstruction of lymphatics and can manifest at any age. The acquired form occurs more commonly in the vulvar region than any other regions of the body and the most common causes includes pelvic surgery, radiation therapy, infection such as tuberculosis, Crohn's disease etc.^[9] Although our case seems to be an acquired case,; we do not find any etiological association. The lesions were present for 4-5 years and history including investigations fails to pin-point any associated cause.

Vulva is an uncommon site for LC. As the clinical presentation of LC may vary from pseudovesicles to nodules or wart-like lesions, correct clinical diagnosis is usually not possible. Most common differential diagnosis includes genital warts, herpes zoster, molluscum contagiosum and even leiomyoma.^[10] Biopsy and histopathology provides the correct diagnosis. Apart from cosmetic concerns, LC may cause some complications that includes swelling and pain in vulva, recurrent cellulitis and psychosexual dysfunction.^[11] Malignant transformation of LC is rare and usually related to radiotherapy which has been previously performed at the sites of LC.^[3]

There is no standard therapy for LC. As there is no medical treatment, the traditional treatment is surgical removal. However, surgery may sometimes fail and early recurrence is common.^[7] Other treatment options include sclerotherapy,

cryotherapy, superficial radiotherapy, pulsed dye laser, intense pulse light and electrocautery.^[7,12] Other treatment modality includes vaporization with CO₂ laser and it usually yield acceptable cosmetic results.^[13] Treatment with Nd: YAG laser has also been tried with good outcomes.^[14] Newer treatment options also include electrodesiccation of the lesion which has been tried following failure of pulsed dye laser with significant improvement.^[15] However, we opted for the traditional surgical removal after explaining to the patient, as we do not have newer treatment facilities. Fortunately, there is no evidence of recurrence after one year of follow-up.

Lastly, we have reported this case because of its rare site of presentation. Thus, although rare; LC should be kept in mind in the differential diagnosis of any warty lesions involving vulva.

Financial support and sponsorship

Nil.

Conflict of interest

There are no conflicts of interest.

REFERENCES

1. Esquivias Gómez JI, Miranda-Romero A, Cuadrado Vallés C, Bajo del Pozo C, Sánchez Sambucety P, Martínez Fernández M, *et al.* Lymphangioma circumscriptum of the vulva. *Cutis* 2001;67:229-32.
2. Whimster IW. The pathology of lymphangioma circumscriptum. *Br J Dermatol* 1976;94:473-86.
3. Patel GA, Siperstein RD, Ragi G, Schwartz RA. Zosteriform lymphangioma circumscriptum. *Acta Dermatovenerol Alp Panonica Adriat* 2009;18:179-82.
4. Buckley DA, Barnes L. Vulval lymphangiectasia due to recurrent cellulitis. *Clin Exp Dermatol* 1990;21:215-6.
5. 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;16:930-4.
6. Peachey RD, Lim CC, Whimster IW. Lymphangioma of skin. A review of 65 cases. *Br J Dermatol* 1970;83:519-27.
7. Lai CH, Hanson SG, Mallory SB. Lymphangioma circumscriptum treated with pulsed dye laser. *Pediatr Dermatol* 2001;18:509-10.
8. Sharami H, Ghaemmaghami F, Yarandi F, Milani F, Alizadeh N. Vulvar lymphangioma circumscriptum: A case report. *Acta Med Iranica* 2004;42:456-60.
9. Mu XC, Tran TA, Dupree M, Carlson JA. Acquired vulvar lymphangioma mimicking genital warts. A case report and review of literature. *J Cutan Pathol* 1999;26:150-4.
10. Gönül M, Cakmak SK, Soylu S, Kiliç A, Gündüz H, Gül U, *et al.* Lymphangioma circumscriptum of the vulva: A case report. *Acta Derm Venereol* 2009;89:180-1.
11. Vlastos AT, Malpica A, Follen M. Lymphangioma circumscriptum of the vulva: A review of the literature. *Obstet Gynecol* 2003;101:946-54.
12. Kudur MH, Hulmani M. Extensive and invasive lymphangioma circumscriptum in a young female: A rare case report and review of the literature. *Indian Dermatol Online J* 2013;4:199-201.
13. Huilgol SC, Neill S, Barlow RJ. CO (2) laser therapy of vulval lymphangiectasia and lymphangioma circumscriptum. *Dermatol Surg* 2002;28:575-7.
14. Sasaki R, Negishi K, Akita H, Suzuki K, Matsunaga K. Successful treatment of congenital lymphangioma circumscriptum of the vulva with CO₂ and long-pulsed Nd: YAG lasers. *Case Rep Dermatol* 2014;6:1-4.
15. Emer J, Gropper J, Gallitano S, Levitt J. A case of lymphangioma circumscriptum successfully treated with electrodesiccation following failure of pulsed dye laser. *Dermatol Online J* 2013;19:2.