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A Rare Case of Pregnancy Induced Lymphangioma of Vulva in A Primigravida with Singleton Pregnancy

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Abstract : *Acquired lymphangioma of vulva is popularly known as acquired or secondary lymphangioma circumscriptum of vulva but the more valid and descriptive terminology should be 'benign acquired lymphangiectasia of vulva'. Clinically, it presents with the surface lymph blisters seen as multiple, grouped, translucent papules, blisters and nodules chiefly over vulva and labia majora. Among its various underlying causes such as trauma, abdomino-pelvic surgeries, carcinoma of cervix, radiotherapy, tuberculosis, filariasis etc., only two cases of pregnancy induced acquired lymphangioma of vulva have been described in the previous literature. Herein we report a similar case in a 29 year-old primigravida in 7-month gestation, presenting with whitish, translucent, papules and blisters over oedematous vulva, labia majora and mons pubis since two months.*

Keywords : *Acquired lymphangioma, lymphangiectasia, lymphangioma circumscriptum, lymph blisters*

I. Introduction

Acquired lymphangioma is a rare benign hypertrophy and ectasia of dermal lymphatics following damage or obstruction to the deeper dermal lymphatics due to various underlying causes. [1] Clinically it presents as grouped, translucent, papules, nodules and vesicles appearing as warty lesion with or without lymphorrhoea. [2] Here we report a unique case of pregnancy induced acquired lymphangioma of vulva (ALV) in a primigravida with 7-month gestation period. To the best of our knowledge, only two cases of pregnancy induced acquired lymphangioma of vulva have been described worldwide.

II. Case Report

A 29-year-old primigravida with 7 months gestation came to our outpatient department presenting with multiple whitish papules, nodules and vesicles over oedematous vulva, labia majora and mons pubis and progressive pitting edema of both lower limbs since 2 months. The lesions were mildly pruritic & painful and had a tendency of oozing clear fluid on gentle manipulation. There was no history of trauma, long term medication, pelvic or abdominal surgery, sexually transmitted diseases, tuberculosis, filariasis, Crohn's disease or radiation treatment to the pelvis in the recent past.

On local cutaneous examination, multiple translucent, whitish, papules, nodules and vesicles were seen over vulva, labia majora and mons pubis arranged in groups exhibiting herpetiform appearance. The size of these lesions ranged from 2 mm - 2 cm in diameter. [Figure 1] Vulva and labia majora were edematous. The lesions were mildly pruritic and painful and mild tenderness was noted on palpation. On puncturing a translucent papule with a 29-gauge needle, oozing of clear fluid was seen. There were no inguinal or femoral lymphadenopathy. On general physical examination, both lower limbs were edematous extending from ankle to mid-portion of thigh. [Figure 2] There were no constitutional symptoms and patient was quite healthy looking.
On systemic examination, no abnormality was noted on obstetrical work-up and ultrasonography of abdominal & pelvic region. All routine laboratory parameters were within normal limits and tests for HIV(I & II), VDRL, HBsAg, HSV(I & II) and Mantoux tests were all negative.

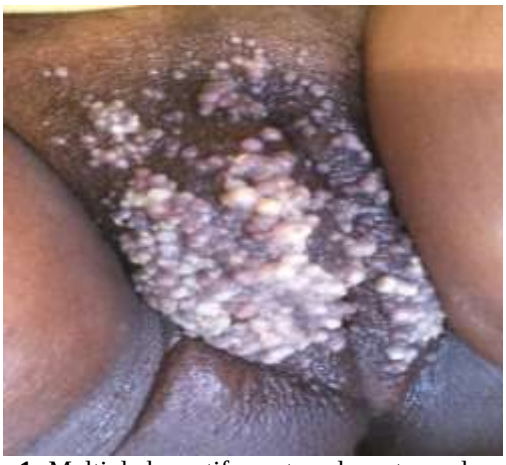

Figure 1: Multiple herpetiform, translucent papules**,** vesicles and nodules on oedematous vulva, labia majora and mons pubis

Figure 2: Non-pitting edema of both lower limbs

III. Histopathologic Examination

Histopathologic examination (HPE) of one of these fluid filled lesions revealed epidermal hyperkeratosis, focal acanthosis and multiple, variable-sized spaces representing dilated upper dermal lymphatic channels lined by flat endothelial cells, containing fibrinous material and red blood corpuscles. [Figure 3]

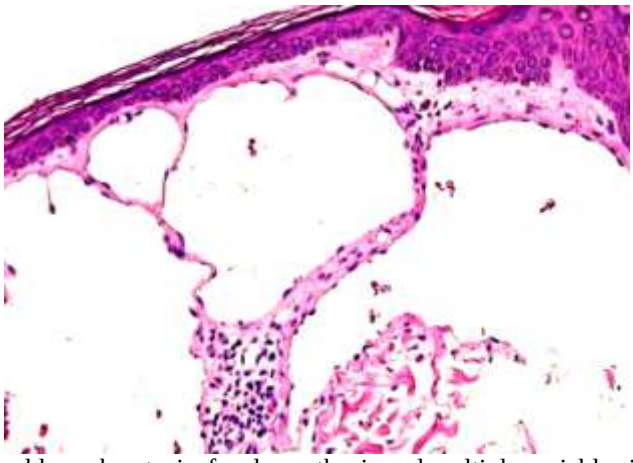

Figure 3 : Epidermal hyperkeratosis, focal acanthosis and multiple variable sized spaces representing dilated upper dermal lymphatic channels lined by flat endothelial cells containing fibrinous materials and RBC's. (H & E, X 40)

On the basis of clinical features and HPE, diagnosis of acquired lymphangioma of vulva was made. In the absence of getting consent of patient for any surgical manipulation, lesions were not removed and patient was sent to department of obstetrics and gynaecology for further evaluation with an advice to turn up regularly at one month interval to assess the progression or involution of the lesions.

IV. Discussion

Lymphatic anomalies include three entities ; lymphatic malformations, lymphangiomas and lymphangiectasia. [3] The increased endothelial cell turnover usually differentiates the 'true lymphangioma' from other two anomalies. True lymphatic malformations results from genetically determined congenital malformation of deep dermal and subcutaneous lymphatics leading to defective lymphatic drainage. Lymphangiectasia are not true tumors or malformations, but represent hypertrophy and distension of superficial lymphatic vessels resulting from obstruction of previously normal deep lymphatics. [1] Inspite of such distinct definitions, 'lymphangioma' is misleadingly used for lymphatic malformation and acquired lymphangioma for cutaneous lymphangiectasia. [4] Applying such terminologies, lymphangiomas are broadly divided into two forms: Congenital and acquired. Congenital form is again subdivided into three types: capillary (congenital 'lymphangioma circumscriptum' first used by Malcolm and Morris in 1889), cavernous and cystic (cystic hygroma). [5] Acquired lymphangioma is also known as acquired LC, secondary LC or cutaneous lymphangiectasia. [6]

Unlike congenital form which was first reported by Fox and Fox in 1878, acquired LC or cutaneous lymphangiectasia results from obstruction and/or damage of the deeper dermal lymphatics following trauma, recurrent cellulitis, surgery, cancer chemotherapy, radiotherapy, crohn's disease, carcinoma of cervix or vulva, tuberculosis, filariasis, STDs etc. [7,8] Apart from the basic differences in the aetiology of congenital and acquired forms, some differences in clinical features also exists. Congenital LC frequently involve axilla, shoulders, flanks, proximal limbs, oral mucosa, tongue and rarely perineum and vulva, whereas in acquired LC, lower limbs and genitalia (penis, scrotum and rarely vulva) are prone to such lymphangiectasia although cases of vulval LC or ALV due to pregnancy has been extremely rare as our case is just third such case. [6-9] Moreover, the superficial dermal lymphatics are more commonly affected in acquired LC rather the deeper one which are predominantly dilated in congenital LC. [8] Clinically, ALV or vulval lymphangiectasia presents as translucent, papules, vesicles or pseudovesicular nodules in the skin, which may ooze lymph spontaneously or after trauma.

ALV is best diagnosed clinically but on HPE, numerous flat CD31 & D2-40 positive endothelial cell-lined dilated lymphatics are characteristically seen in the papillary and mid-dermis. [10] They are filled with clear fluid and may contain red blood cells and fibrinous materials. The surrounding stroma have few scattered lymphocytes. The overlying epidermis is focally acanthotic and hyperkeratotic.

Differential diagnoses of ALV include primary LC of vulva, genital warts, herpes zoster, molluscum contagiosum, syringoma, leiomyoma, cellular angiofibroma, haemangioendothelioma, angiomyofibroblastoma, and aggressive angiomyxoma. [11-13] Histopathologic confirmation of the clinical diagnosis may be done to avoid misdiagnosis and wrong treatment.

There is no standard treatment protocol for primary or secondary LC of vulva. The treatment of such lesions is necessary to prevent secondary infection and to alleviate the swelling, pain and pruritus associated with it. Treatment modalities for LC of vulva which are reported in the literature include electrocoagulation, abrasive therapy, cryotherapy, CO2 and Nd:Yag lasers and surgical excision such as labiectomy, vulvectomy , mass excision, and wide local excision.[12] Recurrence is very common with non-surgical modalities done for deep LC, hence surgical excision is the preferred mode of treatment for deeper and larger LC. [7-9,12] In special situations such as for pregnancy induced LC of vulva, wait and watch principle may also be followed to see spontaneous resolution of the lesions which has been seen in previous such case. [13]

V. Conclusion

The possibility of misdiagnosis, difference in the management of primary and acquired vulvar lymphangioma and chances of secondary infections warrants the early diagnosis and management of ALV especially in pregnant women.

References

[1]. Mallett RB, Curley RK, Mortimer PS. Acquired lymphangiomata: report of four cases and a discussion of the pathogenesis. Br J Dermatol 1992; 126: 380–382.
[2]. Sharma R, Tomar S, Chandra M. Acquired lymphangiectases mimicking genital warts. Indian J Dermatol Venereol Leprol. 2002;68:166–7.
[3]. Enjolras O, Wassef M, Chapot R. Color Atlas of Vascular Tumors and Vascular Malformations. Cambridge: Cambridge University Press, 2007.
[4]. Verma SB. Lymphangiectasias of the skin: victims of confusing nomenclature. Clin Exp Dermatol 2009; 34(5): 566–569.
[5]. Brown JV, Stenchever MA. Cavernous lymphangioma of the vulva. Obstet Gynecol. 1989 May; 73(5 Pt 2):877-9
[6]. Verma S. Pregnancy-induced lymphangiectasias of the vulva. Int J STD AIDS 2008; 19(3): 211–212.
[7]. Amouri M, Masmoudi A, Boudaya S, Amouri A, Ben Ali I, Bouassida S, Guermazi M, Turki H (2007) Acquired lymphangioma circumscriptum of the vulva. Dermatol Online J 13:10
[8]. Singh N, Kumari R, Thappa DM. Vulval lymphangiectasia secondary to tubercular lymphadenitis. Indian J Sex Transm Dis. 2007;28:38–9.
[9]. Haneef NS, Ramachandra S, Metta AK, Haritha K. Lymphangiectasias of vulva. Indian Dermatol Online J. 2011;2:40–2.
[10]. Stewart CJ, Chan T, Platten M. Acquiraed lymphangiectasia ('lymphangioma circumscriptum') of the vulva: A report of eight cases. Pathology. 2009;41:448–53.
[11]. Bhat RM, Saldanha CS, Kambil SM, Dandakeri S. Cutaneous lymphangiectasia of the vulva secondary to tuberculosis. Indian J Sex Transm Dis 2012;33:35-7
[12]. Ghaemmaghami F, Karimi Zarchi M, Mousavi A (2008) Major labiaectomy as surgical management of vulvar lymphangioma circumscriptum: three cases and a review of the literature. Arch Gynecol Obstet 278:57–60
[13]. Khanna, U. and D'Souza, P. (2016), Acquired lymphangioma circumscriptum of the vulva in a twin pregnancy. Journal of the European Academy of Dermatology and Venereology, 30: 147–149. doi: 10.1111/jdv.12640