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IP Journal of Diagnostic Pathology and Oncology

Journal homepage: https://www.jdpo.org/

Case Report

Acquired lymphangioma circumscriptum of the vulva in a middle aged woman with chronic hidradenitis suppurativa – A case report

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ARTICLE INFO

Article history:
Received 23-01-2023
Accepted 20-02-2023
Available online 29-03-2023

Keywords:
Hidradenitis suppurativa
Lymphangioma circumscriptum
Vulva

ABSTRACT

Lymphangioma circumscriptum (LC) is a distention of lymphatic channels present in skin and subcutaneous tissues. LC is either primary (usually present at birth or develops in early childhood) or secondary due to diminished lymphatic outflow. It is commonly prone to develop in oral cavity, tongue, proximal regions of arms and legs, groin, axilla, and trunk. Primary vulvar involvement is unusual. Vulvar LC is a rare untoward outcome of hidradenitis suppurativa (HS), which causes significant physical and emotional discomfort. We report a case of acquired LC of vulva as a sequlae of HS in a 42 years old female who presented with painful papules, edema and thickening of the vulva with history of foul-smelling discharging sinuses. The patient was managed surgically by wide excision and reconstructive surgery by Plastic surgery unit.

1. Introduction

Acquired lymphangioma or lymphangiectasis rarely involve vulva.[1] Most commonly acquired genital lymphedema occurs due to radiation or surgical treatment for cervical cancer, infective causes (acute cellulitis, filariasis, and genital tuberculosis). Lymphedema in context to chronic Hidradenitis suppurativa is infrequent.[1,2] HS is a chronic inflammatory disorder characterized by recurrent abscesses, draining sinus tracts, scarring, and blockage of lymphatic channels and accumulation of lymph in the interstitial space. The most characteristic clinical presentation is of multiple small raised bleb-like lesions from which watery fluid may ooze. A 42 years old female presented in skin OPD with a history of painful papular lesions, edema and foul smelling pus discharging sinuses of vulvar region. The patient was treated surgically with wide excision and skin grafting.

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2. Case Report

A 42-year-old female presented in skin OPD with complaint of painful erythematous swelling with thickened skin and foul-smelling discharging sinuses, over vulval region for the past one year (Figure 1). There was no history of swelling in lower limb, any abdominal surgery or any intrauterine device, pulmonary tuberculosis or diabetes. Patient was referred to Plastic Surgery unit where she underwent reconstructive surgery with local wide excision of edematous vulvar skin followed by skin grafting. Grossly, skin covered soft tissue received in histopathology section measuring 23x10x9cm. The external surface showed multiple pus discharging sinuses along with pustules (Figure 2). Cut section showed solid and partially cystic areas containing purulent material.

Microscopically, epidermis showed pseudoepitheliomatous hyperplasia. Papillary dermis revealed numerous ectatic lymphatics filled with eosinophilic proteinaceous material (Figures 3 and 4).

https://doi.org/10.18231/j.jdpo.2023.009

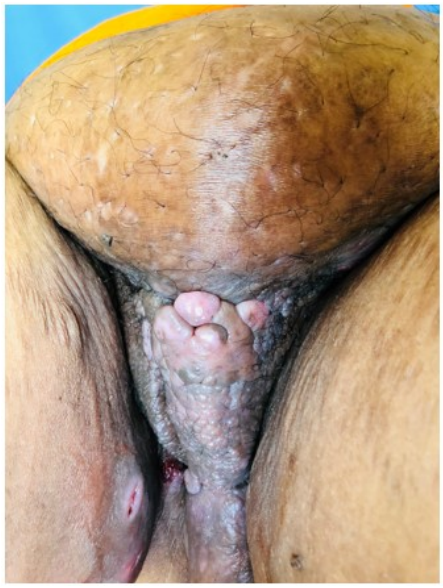

Fig. 1: Clinical image of vulva showing papulonodular lesion and sinus opening

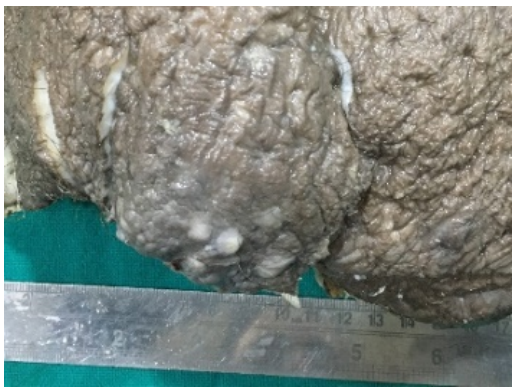

Fig. 2: Gross specimen of vulva showing pustules

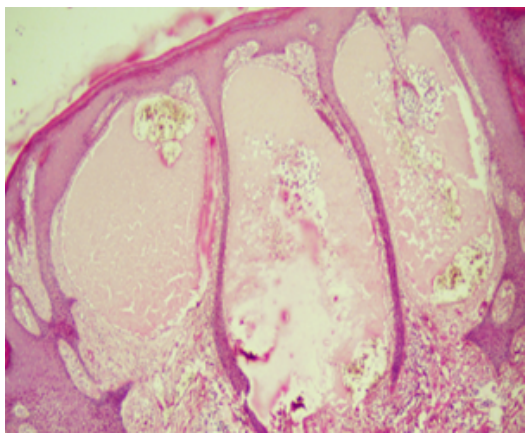

Fig. 3: Papillary dermis showing numerous ectatic lymphatics filled with eosinophilic proteinaceous material (H&E)

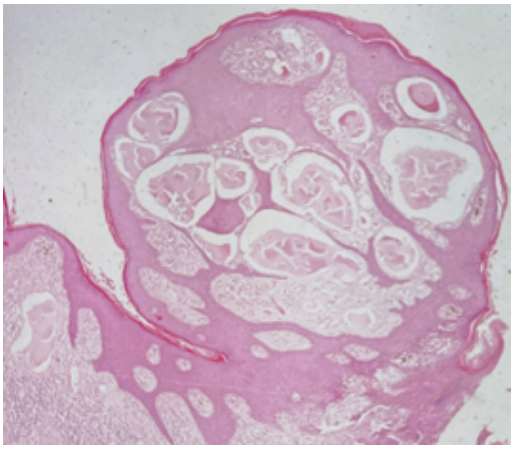

Fig. 4: Numerous ectatic lymphatics lined by flattened endothelial cells in superficial dermis (H&E).

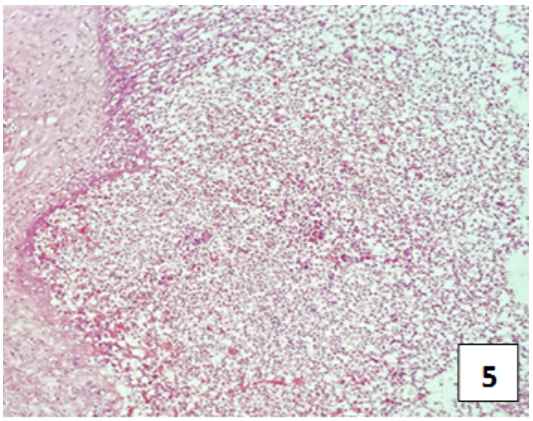

Fig. 5: Sinus showing purulent exudative material (H&E)

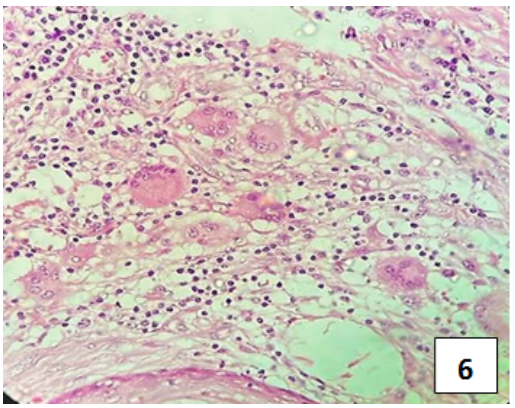

Fig. 6: Chronic inflammation with multinucleated foreign body giant cell reaction

Areas of neutrophilic exocytosis and multiple sinuses lined by granulation tissue and containing neutrophilic microabscesses were seen (Figure 5). Reticular dermis and subcutaneous tissue showed diffuse extensive lymphoplasmacytic infiltration and multinucleated foreign body giant cell reaction (Figure 6). Focally, destruction of sebaceous and skin adnexal structures were also noted.

3. Discussion

Hidradenitis suppurativa (HS) is a chronic, inflammatory skin lesion, which tends to recur, may cause disfigurement, contractures and obstruction of superficial lymphatics vessels with collection of lymph in the interstitial space. Vulvar LC is known to develop as a complication of Hidradenitis suppurativa (HS).[3] Lymphedema can develop in either a widespread or localized manner.[4] It has been hypothesized due to recurrent, chronic inflammation and scarring, subcutaneous lymphatic channels are hindered from draining into the common lymphatic system completely. Thus leading to cumulation of lymph in the superficial interstitial tissue and furthermore smooth muscle around these lymphatic channels contract and causes vesicle formation.[5]

HS generally affects women between 30–58 years, with an average age of 46 years. However, acquired LC is frequently reported in slightly older women as compared to typical onset of HS, which is commonly reported in second and third decade of life. This is because of the fact that lymphatic obstruction occurs due to sequlae of longstanding HS.[3,6]

Clinically, in HS patient presents with history of recurrent multiple pus discharging sinuses, scarring and semitranslucent skin-colored papules which coalescing into plaques, and bear a resemblance to gelatinous mass of 'frog spawn' on the surface of skin.[1,5] It was traditionally known as HS polyposa.[7]

Radical surgery and /or radiation treatment for cervical cancer, infective diseases (acute cellulitis, sexually transmitted diseases, genital tuberculosis, erysiples, lymphogranuloma venereum),chronic inflammatory lesion; Crohn's disease, bilateral varicose veins in the lower extremities, are the pathogenic factors for acquired LC.[1]

Chu et al.[4] employed the term "verrucous lymphostasis" to include both the generalized lymphedema of the abdomen and localized verrucous papules, plaques and nodules secondary to lymphedema on the buttocks, groin and labia majora in their five case series of HS.

Few cases of vulvar HS related acquired LC resembles human papilloma-related condyloma due to fluid filled vesicles along with concurrent lymphedema, which may expand to adjoining abdomen and thigh skin. Rarely, these lesions may have hyperkeratotic and warty appearance, which can mimic verrucous carcinoma.[1,5] Various studies have shown that, there is a 50% increased chances of malignancy patients with chronic HS. Especially, in HS patients the relative risk for squamous cell carcinoma is 4.6 and mostly confined to the perineal or buttock areas.[3,8]

Histopathologic examination revealed thickened, hyperkeratotic overlying epidermis with elongated rete ridges. The superficial dermis displayed lymphangiectasia, which is characterized by miltiloculated cystic spaces filled with lymphatic fluid and lined by endothelium.[1,4]

Among 33 cases with genital lymphedema, Rosenweig et al.[9] observed that most common cause of lymphatic obstruction to be metastatic malignancy of inguinal nodes, radiation therapy, and surgical excision of the lymph nodes. It is possible that HS is a rare cause of lymphedema.

The treatment of choice in patients with acquired vulvar lymphedema is excision of excess tissue followed by split-thickness skin grafting has been reported to be successful.[1]

4. Conclusion

Vulva is the rare site for lymphangioma circumscriptum. It is important to consider LC among differential diagnoses of various papular/warty lesion of genital, in order to address the cosmetic and psychosexual implications and to offer adequate treatment.

5. Source of Funding

None.

6. Conflicts of interest

There are no conflicts of interest.

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Cite this article: Gupta N, Joseph P, Behl L, Mehak, Uppal S. Acquired lymphangioma circumscriptum of the vulva in a middle aged woman with chronic hidradenitis suppurativa – A case report. *IP J Diagn Pathol Oncol* 2023;8(1):46-49.