Case Report

Cutaneous lymphangiectasia of the vulva secondary to pulmonary tuberculosis: a case report

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ABSTRACT

Cutaneous lymphangiectasia of the genitalia represent dilatation of upper dermal lymphatic’s following damage to previously normal deep lymphatics, is an uncommon condition and only few cases have been reported following pulmonary tuberculosis. Here we are describing a case of 75 years old female patient, who came with multiple translucent papulovesicular lesions over the vulva, labia majora, minora and upper thigh from twelve year duration, with watery discharge since eight years. She had a history of pulmonary tuberculosis twenty years ago. The patient underwent punch biopsy of the lesion over the vulva and histopathological examination confirmed our diagnosis as lymphangiectasia of the vulva. She was managed conservatively with anti-tubercular drugs and got relief in watery discharge and skin lesions.

Keywords: Lymphangiectasia of vulva, Pulmonary tuberculosis, Acquired lymphangioma

INTRODUCTION

Lymphatic malformations are mostly congenital, whereas the acquired cutaneous lymphangiectasias (ACL) arise due to obstruction of deeper lymphatic vessels secondary to other aetiology.¹ Acquired cutaneous lymphangiectasia is a rare benign cutaneous disorder characterized by dilatation of dermal and subcutaneous lymphatic channels. The cutaneous lesions of lymphangiectasia can range from clear, fluid filled blisters to smooth, flesh-coloured nodules, sometime coexisting lymphedema is also present. ACL of the vulva is very rare and less than 30 cases have reported in the literature.² In this study we present a case report of 75 years female patient diagnosed as Lymphangiectasias of the vulva secondary to pulmonary tuberculosis.

CASE REPORT

A 75 years old female presented with complaints of multiple raised lesion over the vulva and watery discharge per vaginum for eight years. No significant past history except four months anti-tubercular drugs for pulmonary tuberculosis twenty years back.

On local genital examination, there were single and grouped non tender, non-pruritic, translucent papulovesicular lesions involving both sides (right and left side) of the vulva, including bilateral labia majora, labia minora, and thighs with clear watery discharge from the vagina. There was vulvul edema but no regional lymphadenopathy (Figure 1).
Multiple translucent and papulovesicular lesions involving both sides of the vulva, including bilateral labia majora, labia minora, and thigh with vulvul edema.

Gynecological work-up, routine blood investigations, ultrasonography of the abdomen and pelvic organs revealed no abnormality. Screening tests for human immunodeficiency virus, VDRL, hepatitis B virus, chlamydia and filariasis serology were nonreactive. Chest x-ray was suggestive of healed pulmonary tuberculosis.

Histopathological examination of punch biopsy specimen revealed thin-walled and ectatic lymphatic channels in the superficial dermis. The dermal papillae and the dermis showed proliferating, congested capillary sized blood vessels, melanophages, and diffuse as well as perivascular mild inflammatory infiltrate, finding consistent with the diagnosis of lymphangiectasia of the vulva (Figure 2).

An extensive search for an aetiology of this disease was initiated, but to fail. As tuberculosis is one of the common causes of lymphangiectasia and our patient had history of inadequate chemotheraphy for pulmonay tuberculosis in the past, empirically anti-tubercular (ATT) drug therapy was started. There was complete recovery from watery discharge, lesions decreased in size and number within one month of starting ATT so continued on the same treatment to achieve full resolution of symptoms on completion of treatment.

DISCUSSION

Acquired cutaneous lymphangiectasia (ACL) is also known as lymphangiectasis, acquired lymphangioma, benign lymphangiomatous papules. Although lymphangiectasia has been reported in the literature with increased frequency in recent past, but still the disease remains as under reported as they are often misdiagnosed as herpes, genital warts, or molluscum contagiosum.

ACL can occur in conditions associated with destruction of lymphatic channels such as infections like filariasis, tuberculosis, erysipelas, lymphogranuloma venereum, Crohn’s disease, surgical or radio therapeutic procedures, trauma, female genital mutilation, keloids, scleroderma and lymphatic obstruction associated with neoplasia (carcinoma of the cervix or vulva). The pathogenesis of lymphangiectasia is not known; however, Lymphangiectasis arise following damage to previously normal, deep lymphatic vessels. The lymphatic vessels of the superficial dermal plexus drain a fixed area of skin through the vertical collecting lymphatics to the deep plexus. Damage to the deep lymphatic vessels leads to back-pressure and dermal backflow, with subsequent dilatation of the upper dermal lymphatics. Lymph blisters of the penis or scrotum (and less commonly vulva), leaking lymph or chyle, may be seen in primary lymphedema. In such circumstances the lymphangiectasia represents reflux of lymph or chyle from incompetent pelvic lymphatic’s and is usually associated with lymphedema of one or both lower limbs.

Acquired cutaneous lymphangiectasia is an unpleasant but benign condition. It is characterized by thin-walled translucent vesicles filled with clear, colorless fluid, which may be scattered or grouped like frog spawn. Sometimes these vesicles may be blood tinged and smooth flesh coloured nodules can occur. Rarely, the lesions can have a firm hyperkeratotic appearance. This variation in the morphology is due to a gradual tissue organization, probably enhanced by the presence of lymphedema or recurrent cellulitis.

Histologically, dilated lymphatic channels are present in the superficial and mid-dermis; few dilated lymphatics are seen in the deep dermis. The overlying epidermis may display varying degrees of hyperkeratosis, acanthosis, and papillomatosis, and it may appear to enclose the ectatic lymphatic channels. These dilated lymphatic channels may contain scattered lymphocytes and red
blood cells, imparting a purplish tinge to the lesion. The large endothelium-lined spaces filled with eosinophilic protein-like material in the upper dermis. These endothelial cells have been found to be CD31 and D2-40 positive, but CD34 negative.5

Vulvar lymphangiectasia has to be distinguished from lymphangioma circumscriptum (LC), a congenitally derived hematoma by histology. Other conditions such as mucin secreting metastatic adenocarcinoma mimicking acquired lymphangioma, benign lymphangioendothelioma, syringoma, and condyloma should be ruled out.5

Diagnosis and treatment of the lesions are important because they may be associated with pain, chronic oozing and infection, occasionally leading to cellulitis. The diagnosis is mainly clinical, aided by histopathological finding of dilated lymphatics in the dermis. Treatment should be directed towards the aetiology and aimed at reduction of underlying lymphedema and control of infection.3 Daily compression bandage yields good results, but such a measure is difficult in sites like vulva and scrotum. Ablative modalities such as surgery, electrocautery, cryosurgery and argon laser surgery can be used for treatment for residual lymphangiectasia but are associated with frequent recurrences unless the deep lymphatic cisterns are adequately treated. Sclerotherapy with OK-432 is a new medical treatment, especially effective in macro cystic lesions. In cases where infection is responsible, treatment should be instituted as early as possible to lessen the damage to the lymphatics and relieve lymphatic obstruction.

Lymphangiectasia pose no potential for malignant transformation, however follow-up care is essential for early treatment of recurrence. Lymphangiosarcoma (Stewart-Treves syndrome) may occur in chronic edematous limbs, and early detection is critical. The prognosis for patients with diffuse lymphangiomatosis is poor if the condition is resistant to standard therapies.8

Learning points

1. Detailed dermatologic and histopathological examinations should be performed on patients presenting with genital organ (vulval) papular lesions for early recognition and appropriate treatment of vulval lymphangiectasia.
2. Pulmonary tuberculosis is one of common cause for lymphangiectasia, so an extensive search should be carried out for this when no other definitive cause found and the patient should be treated with anti-tubercular drugs, as in our case.
3. Lymphangioma circumscriptum must be ruled out when considering the diagnosis of ACL.

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