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Vulval lymphangiectasia secondary to tuberculosis: a case report and review of literature

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Abstract: Cutaneous lymphangiectasia, also called as acquired lymphangioma, is a benign cutaneous disorder involving the dermal and subcutaneous lymphatic channels. The cutaneous lesions of lymphangiectasia can range from clear, fluid-filled blisters to smooth, flesh-colored nodules. Coexisting lymphedema is present in most patients with acquired lymphangioma. It can rarely occur on the vulva with less than 30 cases reported so far. We describe a 40-year-old nulliparous woman who came with multiple raised lesions over the vulva. She gave history of getting treated for abdominal tuberculosis in the past. On local genital examination, the patient had multiple, small nodular lesions involving both sides of the vulva. Multiple linear scars were present over the vulva bilaterally. The patient underwent vulval growth excision biopsy. Histopathological examination of the specimen confirmed our diagnosis.
Keywords: Cutaneous lymphangiectasia, acquired lymphangioma, lymphangioma circumscriptum

INTRODUCTION

Lymphangiectasis is superficial lymphatic dilatation which occurs as a consequence of lymphatic damage by an external cause, leading to obstruction of local lymphatic drainage. Lymphangiectasis are also termed as acquired lymphangiomas. Patients usually present with numerous translucent vesicles in a chronic lymphedematous area. Some authors apply the terms acquired lymphangioma and lymphangioma circumscriptum interchangeably. In both conditions, the typical cutaneous lesions are groups of small translucent vesicles, often compared with frog spawn. Although both share similar clinical and histologic features, the authors believe that they are two distinct entities. Acquired cutaneous lymphangiectasia (ACL), also called as acquired lymphangioma, is a rare condition in which dilated lymphatic channels arise following damage to previously normal deep lymphatics, whereas lymphangioma circumscriptum (LC) is used when lymphatic channel dilation occurs because of congenital malformations of the lymphatic system involving the skin and the subcutaneous tissues. Although clinically and histologically they resemble each other, differentiation is possible on the basis of history because lymphangiomas are present since birth or early childhood, while acquired cutaneous lymphangiectasia develop later and are associated with various causes [1,2]. There are less than 30 cases of vulval lymphangiectasia reported in the literature[3].We describe a case of acquired cutaneous lymphangiectasia of the vulva secondary to tuberculosis.

CASE REPORT

A 40-year-old female presented with history of multiple raised lesions over the vulva since 5 years. The number and size of lesions have gradually increased over time. No history of discharge and pain in the lesions. She was otherwise healthy and there was no history of constitutional symptoms. 15 years ago, she was treated for bleeding per rectum and was diagnosed with intestinal tuberculosis and received antitubercular treatment. She did not undergo any surgery or radiation therapy in the past. Her menstrual cycles were regular. She is married for 20 years and has never conceived. Patient was worked up for infertility after treatment for tuberculosis and was found to have bilateral tubal block on hysterosalpingography and was advised invitro fertilization but patient deffered the treatment in view of financial issues. On local genital examination, there were multiple, small, non tender nodular lesions measuring around 5×4 mm involving both sides of the vulva including bilateral labia majora. No discharge was seen from the lesions(Fig-1). The lesions were firm to hard in consistency. Vulval edema was absent. Multiple linear scars were present over the labia majora bilaterally. There was no regional lymphadenopathy. Complete analysis on blood, liver and renal function tests and urinalysis were within normal limits. Mantoux test was negative. Gynecological workup and

ultrasonography of the pelvic organs revealed no abnormality. CECT revealed multiple calcific lymph nodes of approx size 5-6 mm in the periportal, mesenteric and pre aortic region. Terminal ileal loop was thickened, caecum was contracted and pulled up. Ileocaecal angle was obtuse. Right labia majora was edematous. Findings were suggestive of edematous right labia majora with ileocaecal Koch's. Screening tests for human immunodeficiency virus and hepatitis B virus were nonreactive. The patient underwent excision biopsy of the vulval lesions under local anesthesia. Histopathological examination of the biopsy specimen revealed dilated lymphatics and overlying acanthotic epidermis. With the above findings, we came to a diagnosis of lymphangiectasia of vulva.

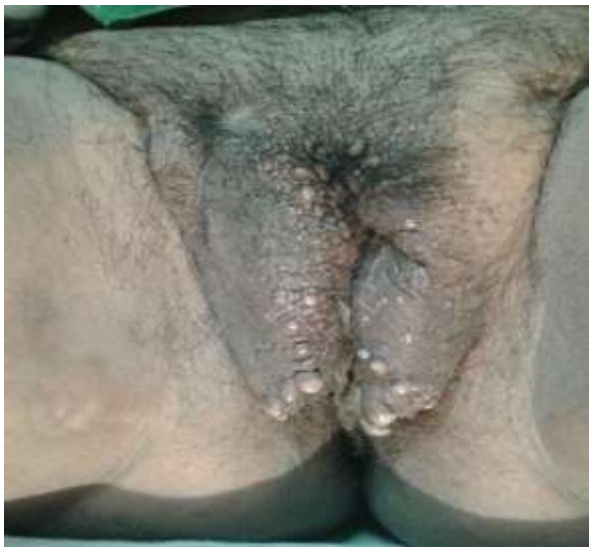

Fig-1: Vulval lymphangiectasia multiple non tender nodular lesions

DISCUSSION

Vulval lymphangiectasia is a rare disease characterised by dilatation of superficial lymphatics. Although lymphangiectasia has been reported in the literature with increased frequency in the past 2 decades, the disease remains rare. The disease affects the age group 22–75 years with mean age 48.5 years [4]. No racial or sexual predominance has been reported.

Lymphangiectasia is seen following surgery or radiotherapy for carcinoma of the cervix or vulva, tubercular inguinal lymphadenitis, or Crohn's disease of the vulva [1,2,3]. Rarely, lymphangiectasias may occur in pregnancy and spontaneously regress with childbirth [5].

The pathogenesis of lymphangiectasia is not known; however, the vesicles associated with lymphangiectasia are suggested to represent saccular dilatations of local superficial lymphatics. These vesicles develop secondary to increased intralymphatic pressure as a result of accumulation of lymph in the superficial vessels caused by damage to previously normal deep lymphatics thereby causing lymphedema.

Vulvar lymphangiectasia can be asymptomatic or may present with pruritis, burning, or pain over the vulva. Clinically, lymphangiectasia is characterized by thin-walled translucent vesicles filled with clear colorless fluid, which may be scattered or grouped like frog spawn. Sometimes the vesicles may be blood tinged and sometimes smooth flesh colored nodules can occur[2,3,6]. Our patient had multiple small nodules seen over labia majora bilaterally. Rarely, the lesions can have a firm hyperkeratotic appearance[3,6]. This variation in the morphology is due to gradual tissue organization, probably enhanced by the presence of lymphedema or recurrent cellulitis[3,7]. Coexisting lymphedema is present in most patients with acquired lymphangioma. They are often misdiagnosed as herpes, genital warts, or molluscum contagiosum[2,3,4].

Histologically, dilated lymphatic channels lined by single layer of endothelial cells are present in the papillary and reticular dermis. Involvement in the deeper dermis is rare. The overlying epidermis may display varying degrees of hyperkeratosis, acanthosis, and papillomatosis. Vulvar lymphangiectasia has to be distinguished from lymphangioma circumscriptum (LC). LC is a congenitally derived hamartoma with early onset of the lesions. Histologically, LC tends to have more extensive involvement of the deep dermis and subcutis. Lymphangiectiasis lack the subcutaneous muscle-coated cisternae characteristic of lymphangioma circumscriptum. Other conditions such as mucin secreting metastatic adenocarcinoma mimicking acquired lymphangioma, benign lymphangioendothelioma, syringoma, and condyloma should be ruled out[1].

Diagnosis and treatment of the lesions are important because lymphangiectasis are often complicated by pain, copious fluid drainage, and recurrent attacks of cellulitis. The diagnosis is mainly clinical, aided by histopathological finding of dilated lymphatics in the dermis. Treatment for lymphangiectiasis is important because of the risk that ruptured vesicles may provide a portal of entry for infection, thus cleansing with topical antibacterial is advisable. Reduction of underlying lymphedema is important. Daily compression bandage yields good results, but such a measure is difficult in sites like vulva[3]. Many surgical treatment modalities have been advocated such as electrodesiccation, lasertherapy, sclerotherapy, cryotherapy, and surgical excision. Excisional surgery eliminates the abnormal subcutaneous lymph vessels and cisterns and corrects the aesthetic appearance of the edematous vulva[8]. Lesion recurrence is frequent but resection can be

repeated several times[9]. Examination of the frozen section of the lateral and deep margins of the excised tissue helps to reduce the reccurence[1]. Carbon dioxide laser therapy vaporizes the lesion superficially and seals the underlying lymph vessels and diminishes recurrence. It can be repeated if any recurrences occur[8]. However, carbon dioxide laser ablation may lead to pain, aggravation of the lesions, and keloid formation. Sclerotherapy with OK-432 is a new medical treatment especially effective in macrocystic lesions[10]. Propranolol, as a new modality of treatment is safe in children and may be an important alternative in the treatment of congenital lymphangiomatosis[11]. However, its role in lymphagiectasia has not been studied. Our patient did not have any significant complaints and opted for conservative management. Lymphangiectasia has a good prognosis because it responds well to treatment. The prognosis of diffuse lymphangiomatosis is poor if the condition is resistant to standard therapies.

Lymphangiectiasis pose no potential for malignant transformation. 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.

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