Acquired lymphangioma of vulva secondary to radiotherapy to carcinoma of cervix; a case report and literature review

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Introduction

Acquired or secondary lymphangioma is a rare condition characterized by simple dilatation of surface lymphatic vessels, due to damage to the deep lymphatics by multitude of causes. Acquired lymphangioma has to be differentiated from lymphangioma circumscriptum (LC) which is a developmental defect of the deep dermal and subcutaneous lymphatics. Both conditions clinically and histologically resemble each other. However, the differentiation is possible on the basis of clinical history, because lymphangiomas are present at birth or early childhood; while acquired cutaneous lymphangiectasia develops later. Here we describe a case of acquired cutaneous lymphangioma of the vulva, secondary to radiotherapy to carcinoma of cervix.

Case report

A 50 year-old female presented with a history of multiple raised skin lesions over the external genitalia of one year duration. These were initially asymptomatic, later copious foul smelly discharge was noted. There was occasional pain and itching.

She has a past history of moderately differentiated squamous cell carcinoma of cervix 8 years ago. For this she had undergone a radical hysterectomy and radiotherapy. She also had bilateral obstructive uropathy as a complication of radiotherapy. For this she had undergone insertion of bilateral JJ stents and taken a course of prophylactic antibiotics.

On clinical examination, multiple vesicles were noted over right labia majora and mons pubis (Figure 1). Few of them were firm and verrucous. There was no tenderness. Inguinal lymph nodes were not enlarged.

H and E section of the skin biopsy showed hyperkeratotic, acanthotic epidermis with broad rete ridges. There were sub epidermal cystic dilatations lined with endothelium containing amorphous eosinophilic material and a few lymphocytes (Figure 2).

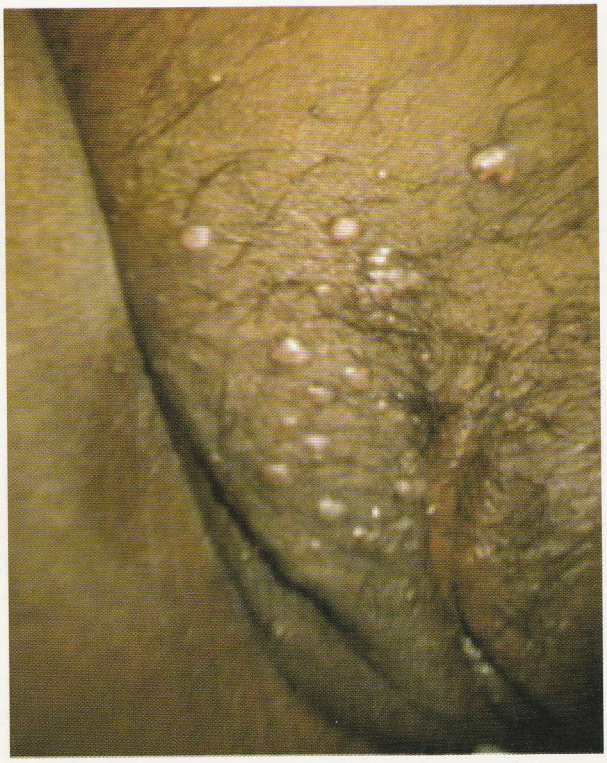

Figure 1.

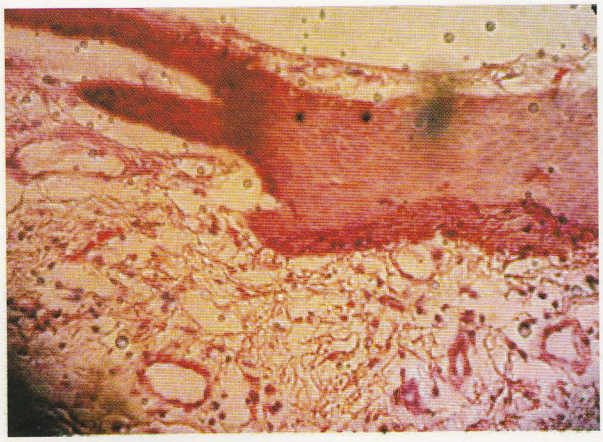

Figure 2.

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Ultra sound study of the lesion showed dilatation of superficial dermal lymphatic channels and oedema of the skin of right side of mons pubis. Her biochemical investigations were normal. Screening tests for human immunodeficiency virus and hepatitis B virus was negative.

This patient was treated with multiple sessions of cryotherapy.

Discussion

Lymphangioma is a lesion composed of dilated lymphatic channels, resulting from defective lymphatic drainage due to congenital or acquired causes. Congenital lymphangiomas (lymphangioma circumscriptum) represent malformation of deep dermal and subcutaneous lymphatics with secondary dilatation of superficial lymphatics. Acquired lymphangiomas or lymphangiectasia represent dilatation of superficial lymphatics, resulting from obstruction of previously normal deep lymphatics secondary to radiation[3], extensive surgery, trauma, keloid, scrofuloderma, pregnancy, scleroderma, neoplasia,and infections such as filariasis, tuberculosis[4], recurrent erysipelas[3,5], lymphogranuloma venereum[1,2,6].

Normally, 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 backflow, with subsequent dilatation of the upper dermal lymphatics[2,7].

Vulvar lymphangiomas are rare and are usually reported following surgery or radiotherapy for carcinoma of the cervix or vulva, tuberculous inguinal lymphadenitis, or Crohn's disease of the vulva[1,2,6].

Clinically, lesions can be asymptomatic, pruritic, burning or painful. The lesions are characterized by translucent vesicles, which may be scattered or grouped like frog-spawn. Some hyperkeratotic lesions mimic genital warts[6,8,9]. They can be misdiagnosed as herpes or molluscum contagiosum[2,6,10].

Lymphangiomas are characteristically known to be arising approximately 7-15 years after lymph node dissection and radiotherapy of the genitalia[11].

The disease affects females aged 22-75 years with mean age 48.5 years[10]. Our patient also developed lymphangioma approximately 7 years after the surgery and radiotherapy.

Histopathological findings of lymphangioma are confirmatory. These include hyperkeratosis and acanthosis of the epidermis with multiple dilated lymphatics in the papillary dermis. Few dilated lymphatics can be seen in the deep dermis. Scattered lymphocytes in the stroma may be seen. The lymphatic endothelial cells have been found to be positive for CD31. However D2-40 is the most specific immune marker for lymphatic endothelium and CD34 is negative[12]. Histological features of our patients are compatible with above classical histological finding. However, specific immunohistochemistry was unavailable for our patient.

As complications, squamous cell carcinoma[13] recurrent cellulitis[3, 5], psychosexual dysfunction and lymphangiosarcoma[20] may develop in long-standing cases. We planned to follow up our patient to detect such complications.

Treatment is aimed at reduction of underlying lymphedema and control of infection[6]. In cases where there is infection, treatment should be instituted as early as possible to minimize further damage to the lymphatics[2].

Daily compression bandaging is difficult in sites like vulva though it gives good results in treating lymphangiomas of other sites[6].

Excisional surgery, carbon dioxide laser[13], cryotherapy[3], electrocoagulation, and sclerosing agent injections are major treatment modalities. Excisional surgery eliminates the abnormal subcutaneous lymph vessels as well as cisterns and corrects the appearance of edematous vulva[11]. Lesion recurrence is frequent with surgery, but resection can be repeated several times without adverse effects[14].

Various lasers such as Pulsed dye and Erbium lasers can be used[15]. Carbon dioxide laser vaporizes the lesion superficially and it seals the underlying lymph vessels and diminishes recurrence. The laser therapy can be repeated if any recurrences occur[11]. However, carbon dioxide laser ablation may lead to pain, aggravation of the lesions, and keloid formation[8].

Serpier H *et al* proposed cryosurgery which provides real benefits with a minimal trauma when treating patients with vulval lymphangioma[3]. Our patient is treated with multiple cryotherapy sessions.

Sclerotherapy with OK-432 is a new medical treatment especially effective in macrocystic lesions[16,17].

Propranolol, as a new modality of treatment, has been successfully used and is safe in children and

may be an important alternative in the treatment of this disease in infants[18].

In conclusion, acquired vulval lymphangioma is a rare benign lesion composed of superficial dilated lymphatics. It is important to understand broad spectrum of differential diagnosis, different clinical courses and multiple risk factors of this condition. Prompt treatment should be instituted to minimize further damage to the lymphatics by infection and long standing lymphoedema. It is mandatory to follow up these patients, because transformation to squamous cell carcinoma and lymphangiosarcoma have been reported.

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