Lymphangioma Circumscriptum Of Vulva Due To Tuberculosis

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Citation

D Pal, D Moulik. *Lymphangioma Circumscriptum Of Vulva Due To Tuberculosis*. The Internet Journal of Gynecology and Obstetrics. 2008 Volume 10 Number 2.

Abstract

We report a case of a 17-year old female patient presented with huge swelling of the vulva following tubercular lymphadenitis of the inguinal region. The case was diagnosed as acquired lymphangioma circumscriptum of the vulva due to destruction of the inguinal lymph nodes by tuberculosis. The vulval swelling did not respond after antitubercular chemotherapy and required a cosmetic surgery. The role of surgery is emphasized in this case.

INTRODUCTION

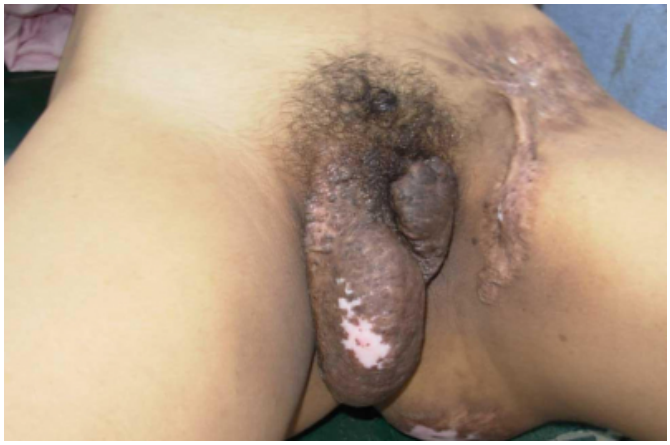
Lymphangioma circumscriptum is the end result of a variety of obstruction to the lymphatic system affecting the extremities and genitalia [1] . In India it is the common sequel of filariasis, lymphogranuloma venereum and rarely Donovanosis, carcinoma or tuberculosis. [1,2] It occurs sometimes after radical hysterectomy, pelvic lymphadenectomy, chron's disease and radiotherapy for carcinoma cervix [3] . Tubercular etiology of this entity is very rare and only few cases are reported in the English literature [3,4,5] .

CASE REPORT

A 17-year old unmarried female patient from rural background presented with asymptomatic diffuse swelling of the vulva for last 3 years. It started 5 years back when she developed multiple swellings in the left inguinal region. Initially she was treated by a village doctor and the swelling turned into a large abscess in the left inguinal region subsequently which ruptured spontaneously. On examination there was a large ulcerated area in the left inguinal region with copious serous discharge. Right inguinal lymph nodes were matted and tender. No other group of lymph nodes were palpable.

Figure 1

Figure 1: Photograph showing vulval swelling with depigmented skin with scar of healed abscess in the left inguinal region



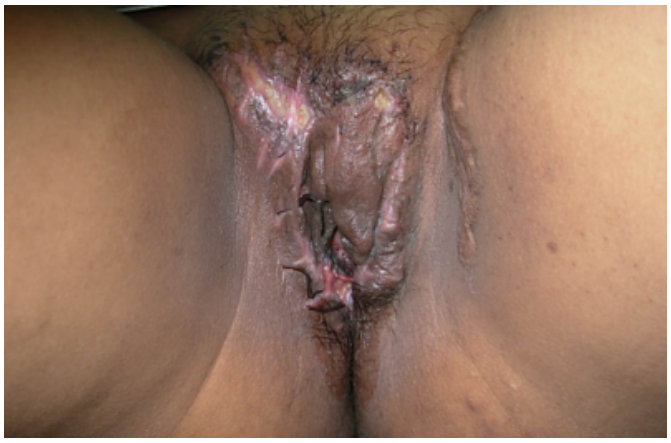
The vulva was diffusely edematous with overlying skin studded with papulo-vesicles with watery secretion at that time with loss of pigmentation on the vulval skin (Fig.1). The labia minora on inner aspects showed lymphangiactasis and a few vegetative lesions. Urethral orifice, vagina and anal orifices were normal. Biopsy from the vulva showed features of lymphedema with gross fibrosis and tissue smear was negative for tuberculosis. She had a normal haemogram with an erythrocyte sedimentation rate 110 mm in first hour. She was normoglycemic with normal renal biochemical parameters. Night blood for microfilaria was negative. Pus culture showed growth of E.coli in ordinary culture. Z-N staining from the pus yielded M. tuberculosis and AFB culture also showed growth of M. tuberculosis. Smear from

the ulcer did not show any Donovan bodies. Mantoux test was positive for tuberculosis with 16mm X 16mm induration and erythema. Chest X-ray was noncontributory. Pelvic ultrasonography was unremarkable.

Ant tubercular drugs were started with rifampicin, INH, pyrazinamide and ethambutol for two months and the last two drugs were continued for another four months. The ulcer started to heal within 2 weeks of therapy and on 3 months follow up ulcer healed completely but her vulval swelling increased in size inspite of AT drugs. Since the patient was young with an extensive lesion we surgically removed the involved area with an elliptical incision on both sides by a wide excision of 3 cm diameter in depth with suturing of the overlying skin which led to complete recovery with a good cosmetic appearance (Fig. 2).

Figure 2

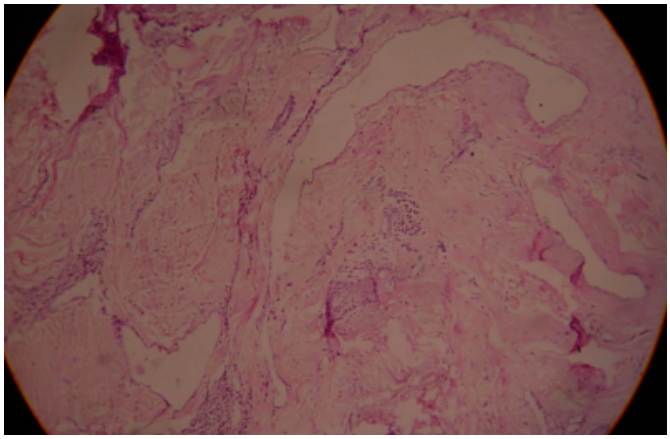
Figure 2: Postoperative appearance of vulva after one week of operation



Histopathology suggested multiple dilated lymph vessels in the papillary dermis lined by a single layer of endothelial cells with inflammatory infiltrate in the papillary dermis suggestive of lymphangioma circumscriptum.(Fig. 3) Till one year follow up there is no recurrence.

Figure 3

Figure 3: Histopathology from vulval swelling showing dilated lymphatics in a fibrocollageneous stroma (H & E X 400)



DISCUSSION

Lymphangiomas are very rare proliferations of the lymphatic system. Congenital cases are found after birth or early childhood but acquired cases affects young and reproductive age group, and are associated with physical disability and extreme mental anguish. Acquired lymphangioma circumscriptum of the vulva is induced by impaired lymphatic flow. Obstruction of lymph flow causes lymph stasis and subsequent proliferation of fibrous tissue resulting in lymph edema and elephantiasis. Obstruction of lymph flow occurs directly from tubercular lymphangitis, destruction of lymph glands by disease process or by constriction of lymph vessels from the surrounding fibrous tissue. In filariasis and LGV the lymph edema is secondary to lymphangitis whereas in granuloma inguinale it is due to the pressure of scar tissue on lymphatics [2] . In this case absence of tubercular histology from vulva rules out direct infiltration of tuberculosis in vulva. Clinical course of the disease points towards the destruction of the lymph nodes by tubercular abscess and constriction of the remaining lymph vessels by fibrosis as the causative factor for lymphangioma circumscriptum of vulva in this case.

There is no medical treatment in lymphangioma circumscriptum [3] . Preferred treatment is complete surgical excision [3,4,5] which gives good cosmetic appearance. Recurrences are not uncommon unless deep lymphatic cisterns are adequately removed [4,5] . Vaporization with a CO2 laser is an alternate modality recommended recently for lymphangioma circumscriptum of vulva with an acceptable cosmetic result [6] but therapeutic failure after laser therapy has also been reported [4] . In young patients surgical excision

is the best option where cosmetic aspect of the treatment is also important and there is possibility of keloid formation later on after laser therapy [3].

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