

Pregnancy and Vaginal Childbirth Followed by Reconstructive Vulval Surgery in a Woman with Giant and Isolated Vulval Elephantiasis

Rinnie Brar, Minakshi Rohilla*, Atul Parashar and Vanita Jain

Post-Graduate Institute of Medical Education and Research, Chandigarh, India

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*Corresponding author: minurohilla@yahoo.com

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Abstract

Isolated elephantiasis of the vulva refers to a gigantic swelling of the vulva without concomitant swelling of lower limbs. It is a rarely reported entity and its occurrence during pregnancy has been reported only once. This likely happens because of an insult to lymph node or lymphatic vessels draining the vulva which causes lymphatic obstruction from vulva. This causes lymphoedema which persists and gradually over time causes gigantic swelling of vulva. We report a case of an isolated vulval elephantiasis during pregnancy and we discuss the possible etiologies and management issues. This is the second case ever reported of isolated vulval elephantiasis during pregnancy. Our patient had a successful vaginal delivery followed by a satisfactory genital reconstruction at 8 months post partum and no recurrence thereafter.

Keywords: Vulva Elephantiasis; Plastic surgery; Vulval surgery Esthiomene.

Introduction

Isolated elephantiasis of the vulva refers to a gigantic swelling of the vulva without concomitant swelling of lower limbs. Genital elephantiasis is a common result of lymphatic obstruction caused by filariasis, lymphogranuloma venereum, granuloma inguinale, carcinomas, lymph node dissection, irradiation and tuberculosis.[1] It is a rarely reported entity and its occurrence during pregnancy has been reported only once. [2] This likely happens because the primary insult (infection, radiation or surgery at the vulva etc.) resolves; however, lymph node or lymphatic vessel damage causing obstruction to lymph drainage of vulva persists which gradually over time causes lymphoedema and subsequent swelling of vulva. We report a case of an isolated vulval elephantiasis during pregnancy and we discuss the possible etiologies and management issues. Our patient had a successful vaginal delivery followed by a satisfactory genital reconstruction at 8 months post partum and no recurrence thereafter.

Case Report

A 20 year old primigravida at 22 weeks of gestation presented to gynecology outpatient wing with the complaint of a large mass at the vulva. This vulval mass was very small in size before pregnancy which gradually increased in size as her pregnancy progressed. There was no history suggestive of tuberculosis, chyluria, swelling of thighs, legs or feet, genital ulceration or bubo formation, multiple sexual partners or partner with multiple sexual contacts, similar lesion at any other site of the body or any similar lesion in her partner.

Her general physical examination was unremarkable. She had a body-mass-index of 23 kg/m², blood pressure of 110/80 mmHg, pulse rate of 84 per minute. There was no evidence of cervical, axillary or inguinal lymphadenopathy,

or any features of micronutrient deficiency. Her abdomen was soft, non tender, and uterus was palpated to a size corresponding to 22 weeks of gestation. Bilateral lower limb examination was normal with no swelling, edema or ulceration. Local examination of the perineo-vulval region revealed 12 x 10 cm fleshy, non-tender growth with irregular surface arising from both labia merging centrally and involving the clitoris. There was no involvement of urethral or peri-anal region (Figure 1a and 1b). Gynecological examination revealed healthy vagina and cervix on speculum examination. On per-vaginal bimanual examination, cervix was normal and gravid uterus of size approximating 22 weeks was palpated.



Figure 1: (a) Large mass arising out of and involving bilateral labia minora causing vulval elephantiasis, labia majora appear uninvolved; (b) Same mass lifted up and opened depicting normal vaginal orifice underneath.

On evaluation, preliminary blood investigations including hemogram, renal and liver function tests were normal. Serological testing for HIV was negative. STI (Sexually transmitted diseases) testing for Lymphogranuloma venereum (LGV), genital warts, donovanosis and syphilis returned negative. Tuberculosis was ruled out on TB-Interferon gamma release assay (IGRA). Microscopy from blood drawn at midnight was performed on three consecutive nights and no microfilariae were found. Additionally, antenatal investigations including thyroid function tests, hemoglobin electrophoresis, glucose tolerance test, Level II sonography were normal. The mass was biopsied and sent for histopathological examination which reported fragments of fibrocollagenous tissue lined by stratified squamous epithelium. There were no acid fast bacilli (AFB) to suggest tuberculosis. There was no evidence of any dysplasia.

A provisional diagnosis of vulval elephantiasis of an unknown etiology was made. A meeting was held with the woman and her partner for discussion of the plan for pregnancy and delivery. Anticipated complications and management during antenatal period, labour and delivery were discussed. The woman opted for expectant management of the mass with close observation till delivery and reconstructive surgery later in the post partum period. There were no obstetric complications in the remainder of her pregnancy. Vulval mass remained static in size on follow up.

Spontaneous labour and leakage settled in at 37 weeks of gestation, intravenous labour analgesia was provided and her labour progressed uneventfully. A right medio-lateral episiotomy, 45° from the midline was given in the second stage of labour after lifting up the labial mass. Vaginal delivery was conducted successfully delivering a healthy baby boy weighing 2.7 kgs with APGAR scores of 8 and 9 at 1 and 5 minutes respectively. There were no maternal injuries and the episiotomy was repaired in layers.

At 6 weeks and at 6 months post partum she presented with slight reduction in the size of the elephantiasis. However, total resolution of the same was not attained. Excision of the vulval mass and vulval reconstruction was planned and performed under regional anaesthesia at 8 months post-partum in liaison with the department of plastic surgery (Figure 2a and 2b). The final histopathology report confirmed the mass to be a soft tissue growth lined by stratified squamous epithelium with underlying fibro-collagenous stroma containing dilated lymphatics with no AFB or dysplasia. The patient had a second uneventful pregnancy in the 3rd post-operative year delivering a healthy baby girl of 2.8 kgs vaginally at term. There was no recurrence of the vulval mass in the second pregnancy or during subsequent follow up over 2 years.



Figure 2: (a) Pre-operative photograph depicting regression in the size of the mass, however, complete resolution was not achieved; **(b)** Immediate post-operative photograph following reconstruction surgery depicting normal vulval anatomy.

Discussion

Our case had isolated vulval elephantiasis, which was first brought to medical attention during pregnancy. Work up for the etiology of the same revealed nothing contributory. There have been few cases of non pregnant women with such isolated vulval elephantiasis reported in the literature where no etiology could be found. In cases where etiology could be elucidated, most cases of elephantiasis were secondary to filariasis, an infection which is common in the tropical countries. Filarial worms (*Wuchereria bancrofti* and *Brugia malayi*) lodge in the lymphatic system causing blockage of lymphatic drainage. Filariasis is diagnosed by the presence of microfilariae on blood films or by demonstration of positivity to circulating filarial antigen (CFA). [3, 4] In our case, both tests (blood smears and CFA) returned negative, ruling out filariasis. In the only other case reported describing pregnancy with vulval elephantiasis, the cause was active filariasis as microfilariae were demonstrated circulating in the blood. [2] Similar lymphatic obstruction due to lymph node tissue destruction by tuberculosis, vulval radiation or surgery has also been reported to cause elephantiasis.[5] In our case, there was no history of vulval radiation or surgery. Active tuberculosis was also ruled out by serological assay and histopathological findings. Histopathological findings suggestive of tuberculosis include caseating granulomas, lymphocytic infiltration with giant cells and acid fast bacilli positivity, which were not seen in our case. [6] Some STIs which cause mass lesions at vulva may appear like lymphatic elephantiasis to the untrained eye such as LGV, genital warts and granuloma inguinale. [7] In our case, all these infections were ruled out by physical and histopathological examination. Neoplastic nature of the mass was also ruled out by histopathological examination of the mass.

So, after exclusion of these causes, the likely etiology of vulval elephantiasis in our case was postulated to be either congenital lymphoedema or lymphatic destruction due to some infection in the past, as there was no evidence of any active infection. Further conclusive evidence of lymphatic obstruction could have been obtained by performing lymphoscintigraphy; however, our patient refused it due to concerns of fetal exposure to nuclear compounds. [8]

Regarding the management of our case, we discussed various options with the woman in detail. Option of excision of the vulval lesion and genital reconstruction during pregnancy was also given. Benefits of earlier relief in dragging pain and heaviness at the perineum, and increased chances of a successful vaginal delivery were explained. However, our patient refused this option due to risk of recurrence and risk of anesthesia during pregnancy.[9] In the previously reported case of vulval elephantiasis in pregnancy, excision of the vulval lesion was performed at 5 weeks of gestation; however, lymphedema recurred to pre-operative proportions by 12 weeks of gestation.[2] Our patient opted for conservative management for vulval elephantiasis to avoid surgery and anesthesia during pregnancy. Risk of increase in the size of the elephantiasis during pregnancy which may make vaginal delivery unfeasible was explained. At term, thorough counseling of the couple regarding mode of delivery was done. Benefits of vaginal delivery over elective caesarean section were explained. However, risks associated with vaginal delivery such as accidental extension of episiotomy into the lesion, perineal tears, hemorrhage or rupture of mass was also explained.

Our patient chose to continue her pregnancy unintervened with respect to vulval mass and had a successful vaginal delivery with an optimal maternal and fetal outcome. Subsequent excision of the vulval mass was done after her puerperal recovery. Successful surgical removal of mass with genital reconstruction has been reported in many cases of non-pregnant women with such large vulval masses to improve cosmesis and to reduce chances of secondary infection due to pressure ulcerations. [5, 10] It seems best to postpone the excision of vulval mass during pregnancy to avoid risk of abortion, risk of recurrence and anaesthetic exposure to fetus.

Take home message: Successful vaginal delivery is possible in cases with gigantic vulval elephantiasis during pregnancy depending on the size and location of the mass. The management plan of such a case requires comprehensive counselling of the woman and her partner to achieve optimal perinatal outcome. A thorough clinical examination, careful follow up, planning of delivery and successful excision of vulval mass subsequently is of paramount importance for the management of giant vulval elephantiasis in pregnancy.

Disclosure

Written informed consent was taken from our patient.

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