Recurrent Unilateral Vulval Elephantiasis: A Case Report

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Received May 2013; revised and accepted September 2013

Abstract

Genital elephantiasis is caused by a variety of infective and non infective causes leading to blockage of lymphatic. We are presenting a rare case of recurrent unilateral vulval elephantiasis which has recurred after initial reconstructive surgery. A 38 year old female presented with vulval swelling and on examination there was gross unilateral vulval enlargement. FNAC (Fine needle aspiration cytology) and biopsy were contributory for diagnosis. Patient was started with antibiotics and daily dressing was done till the infection was subsided and the patient was planned for reconstructive surgery.

Keywords: Vulval Elephantiasis, Filariasis and Reconstructive Surgery

Introduction

Elephantiasis is one of the oldest, bizarre and most crippling disorder that has a long history and worldwide distribution. Genital elephantiasis is caused by a variety of infective and non infective causes leading to blockage of lymphatics. Esthioneme is a Greek terminology used to describe elephantiasis which means to eat and carries an idea something gnawed, eroded or ulcerated (1).

Filariasis is endemic in tropics and subtropics. Wuchereria bancroftii, which accounts for 90% of filarial infections and is estimated to infect 120 million people in the tropics (2). Out of infected people1:3 live in India (3) and 1 billion of people are at the risk of infection (4). The order of involvement of lymphatic filariasis is lower limb, trunk, breast, upper limb and genitelia. In India Bihar has highest endemicity (around 17%), Kerala (around 15.7%) and utter predesh (around 14.6%) (2).

We are presenting a rare case of unilateral vulval elephantiasis which has recurred after initial reconstructive surgery. Genital deformity due to filarial aetiology in female is very rare. Recurrence after initial reconstructive surgery is very rarely reported.

Case history

A 38 year old female patient presented with swelling in vulval region since 2 years. Patient complains of increase in the size of the swelling from past 2 years and swelling has reached to present size. Her menstrual history was normal. Para1, living 1 delivered by Lower segment caesarean section 15 years back. No history suggestive of tuberculosis. No past history of genital ulcer, inguinal swelling, surgery or irradiation.

The swelling was small in size of around 6*7 cm in 2002 and the swelling was excised and diagnosis of vulval elephantiasis was made and pelvic reconstructive surgery was done. Reports of which are not available but according to patients history her external genital were of normal size fallowing surgery.

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On examination patient was afebrile and no lymphadenopathy and her gait was unusually wide and large, hypertrophied, pendulous, multilobed mass of hypertrophied tissue hanging down and obstructing the vulval cleft was seen on left side (fig 1). On standing position swelling was hanging up to upper 1/3rd of thigh. Overlying skin was hard and thickened and showed extensive rugosities. The overlying skin was chronically infected with areas of ulceration and pigmentation. There was foul smelling odour due to infection. On palpation it was firm irregular and none pitting. No inguinal lymphadenopathy or any scars and sinuses in that area. On systemic examination no abnormality detected.

FNAC (fine needle aspiration cytology) done outside 2 months before showed hypocellular smear with lymphocytes on a proteinecious background. Biopsy showed lesion composing of interlacing band of fibroblasts and collagen and perivascular collection of lymphocytes and mononuclear cells were present (fig 2). The stroma showed myxoid changes and foreign body granulomas were present and features suggestive of superficial chronic inflammation. ZN staining was negative. PAS staining showed negative for fungal hyphae. Buffy coat examination no filarial was seen. Filarial antigen assay was positive. Her ESR was 50mm in first hour. Mantoux test was negative. Pelvic ultrasonography was unremarkable. Smear from the secretion showed mixed bacterial flora.

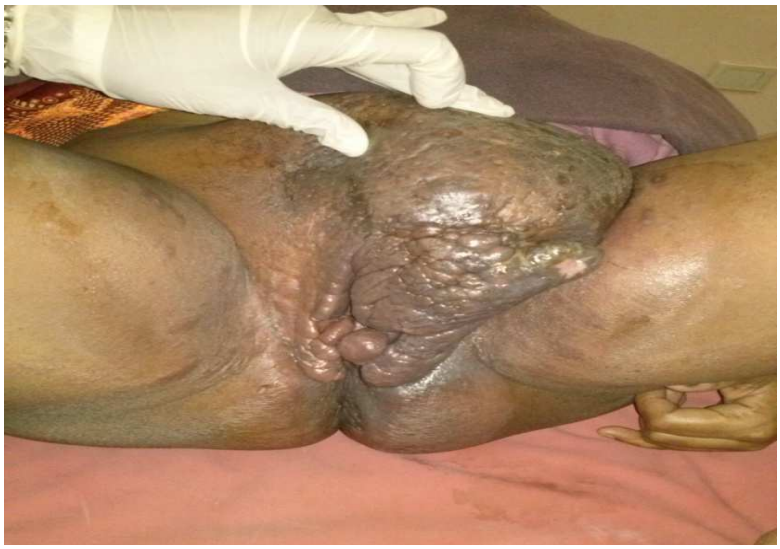

Figure 1: Unilateral vulval elephantiasis

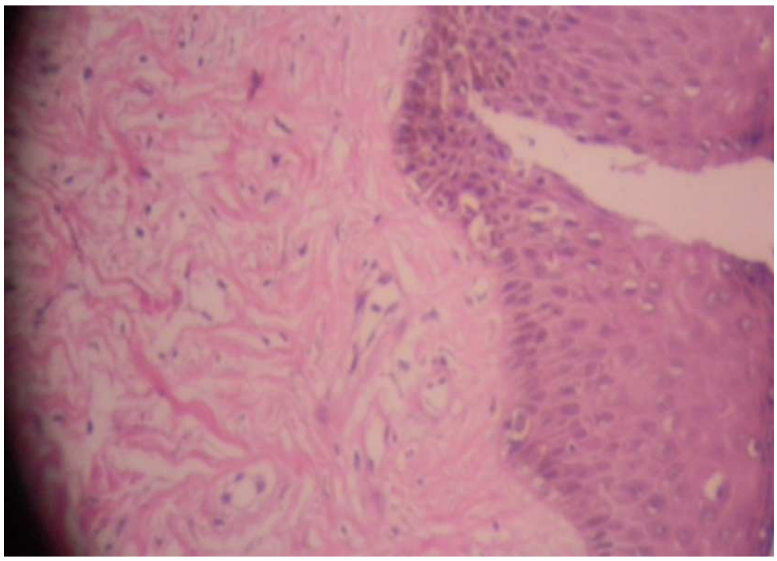

Figure 2: Biopsy report showing lymphatic infiltration and proliferation of fibrous tissue

Patient was started with antibiotics and daily dressing was done till the infection was subsided and the patient was planned for reconstructive surgery (excise the swelling and reconstruction of external genital) but the patient refused surgery due to personal reason.

Discussion

The term elephantiasis was first described by Celsius (30BC-50AD) (3). Lymphatic filariasis external genitelia are very rare and it has been reported as sporadic case reports even though it was first described in 1673 (5). Largest study on these cases was from India in 1980 where they have described 25 case reports (4).

Elephantiasis of vulva roughly affects not more than 1-2% of total case of filarial elephantiasis (6). Causes include filariasis, lymphogranuloma venerum, donovanosis and tuberculosis, it can also occur fallowing radical hysterectomy, pelvic lymphadenectomy and radiation.The main differential diagnosis include fibro epithelial polyp, fungal infection, granuloma inguinale, carcinoma and irradiation (7).

Management of vulval elephantiasis include multidisciplinary approach involving gynaecologist, dermatologist and plastic surgeon. Aim of the management is to reduce the swelling, restore the normal shape and sexual function and to prevent inflammatory episodes.

Diethylcarbamazine (DEC) in three divided doses of 6 mg/kg/day for 12 days kills adults and microfilaria. Good skin care and prompt treatment of the bacterial infections is very important in management. Surgical management includes excision of skin and subcutaneous tissue of involved part and closing the defect with adjacent normal skin. If the defect is very large split thickness skin graft or bilateral super medial thigh flaps can be used. Excision therapy for vulval elephantiasis is quiet satisfactory (8).

This case is presented because of its rarity and it recurrence after initial reconstructive surgery. Vulval filariasis in most of the times is a clinical diagnosis. Neither the micro filarial is seen in peripheral smear nor the filarial worm demonstrated in the tissue section. Only clue towards diagnosis in our case is positive filarial antigen assay.

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