Vulvar Elephantiasis of Filarial Origin: A Case Report

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ABSTRACT

Lymphedema can be defined as swelling of soft tissues which is the result of accumulation of protein rich interstitial fluids caused by a low output failure of lymph.1 This leads to proliferation of fibroblasts and mast cells, organization of the edema fluid and sclerosing fibrosis of the subcutaneous tissue giving rise to the firm, nonpitting and irreversible swelling. Hyperkeratosis, verrucous and condylomatous changes are features of long-standing lymph stasis and are collectively termed 'elephantiasis'.2 The legs, arms and genitalia are the commonly involved. Elephantiasis can be filarial or nonfilarial in origin. Filariasis results from infection with Wuchereria bancrofti and Brugia malayi. While Wuchereria infection has only been noted in humans, Brugia infection has been noted in both man and animals. Filarial elephantiasis of female genitalia is extremely uncommon; a rough estimate of its incidence would not be more than 1-2% of total cases of filarial elephantiasis.3 We present a case of vulval elephantiasis due to underlying filarial etiology.

CASE REPORT

This case report describes a 40-year-old lady para 3 + 0 who came to our center with complaints of mass in genital area with associated dragging pain and difficulty in coitus, difficulty in walking and psychological distress for last 3-5 years. No history of chronic cough, chyluria or any lymphadenopathy. She was 68 kg, attained menarche at 13 years, a healthy lady with all normal systemic examinations. All previous pregnancies were uneventful resulting in normal vaginal delivery and live births. No history of lymphedema in any family member.

On investigation, laboratory findings were hemoglobin 11 g/dl, leukocyte count 6,400 cells/mm³, neutrophilia of 80%, absolute eosinophil count was 200. Patient was hepatitis B positive (HBsAg).

On examination, there were two nontender irreducible huge well-defined bossilated vulval mass, right and left measuring 45 × 38 cm and another 22 × 20 cm in size, respectively (Fig. 1). There were multiple nodular swellings on outer surface on each mass.

Figure 1. Preoperative picture showing two nontender irreducible huge well-defined bossilated vulval mass, right and left measuring 45 × 38 cm and another 22 × 20 cm in size, respectively.
Excision of both masses with vulvoplasty was done in single stage (Figs. 2 and 3). There were two large bossilated mass weighing 15 and 8 kg, measuring 48 × 40 × 10 cm and 26 × 25 × 7 cm in sizes, respectively with multiple nodular swellings on outer surface measuring 2.5 × 0.5 cm in sizes. Postoperatively the patient was kept on amoxiclav and amikacin prophylaxis. The postoperative period was uneventful.

Key histological features were stratified squamous epithelium showing acanthosis and hyperkeratosis, underlying stroma showed dermal fibrosis and collagenization with mixed inflammatory infiltrate in the form of eosinophils, polymorphs, lymphocytes, macrophages and plasma cells (Fig. 4).

Discussion

Vulval lymphedema can be caused by radiotherapy and as a postoperative complication after vulvectomy and inguinofemoral lymphadenectomy. Vulval lymphedema can be a rare extraintestinal manifestation of Crohn’s disease but the ulcer in such cases shows epitheloid and giant cell granulomas. Clinically, vulval lymphedema may mimic genital warts. Lymphangioma circumscriptum was ruled out since it has an early onset at birth with initial presentation of thin translucent vesicles, which may become verrucous later on. Microscopically, it may mimic aggressive angiomyxoma but lacks true thick walled vessels.

As per recommendations most lymphedema patients can be treated with a combination of limb elevation, a high quality compression garment, complex decongestive physical therapy and compression pump therapy and if necessary surgery. Surgical techniques for correcting lymphedema may be excisional or physiological. Surgical treatment is used only in extreme cases in order to reduce the weight of the affected organ, to help minimize the frequency
of inflammatory attacks, to improve cosmesis, and to potentially reduce the risk of secondary angiosarcoma.12,13 Our patient came with complaint of vulval swelling with inability to walk, with difficulty in coitus. Excision of mass with vulvoplasty was done. Excision results in better quality-of-life and improved cosmesis, which make the patient perform better her sexual activities and daily needs (Fig. 5).

REFERENCES


No Mental Harm from HRT in Early Menopause

Hormone replacement therapy (HRT) in early menopause caused no apparent harm to patients’ cognitive function, according to long-term follow-up of participants in clinical trials. (Source: Medpage Today)

Uterine Bleeding: ACOG Updates Guidelines

The American College of Obstetricians and Gynecologists has issued updated guidelines for treating abnormal uterine bleeding caused by ovulatory dysfunction. The new guidelines were Published in the July issue Obstetrics & Gynecology. (Source: Medscape)