Lymphoedema Vulva

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Introduction

Lymphoedema 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, organisation of the oedema fluid and sclerosing fibrosis of the subcutaneous tissue giving rise to the firm, non pitting 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. We present a case of lymphoedema vulva due to underlying tubercular aetiology and aggravated iatrogenically.

Case report

A 22 year old unmarried girl presented with asymptomatic gradually increasing multiple warty growths and swelling of the vulva of six months duration. She denied history of sexual exposure, irregular fever, weight loss, chronic cough or any digestive symptoms. Her periods were regular. Four years earlier she had developed multiple painless swellings in the neck and groin. The neck swellings burst spontaneously and healed gradually. The groin swellings on the left side were given multiple incisions by a village quack which slowly healed. General examination revealed an averagely built and nourished female with matted cervical, axillary and inguinal lymph nodes. There was no pallor, icterus or pedal oedema. Local examination revealed multiple, firm, hyperkeratotic glossy papules and swelling of vulva, multiple healed scars over left inguinal region (Fig.1) along with matted lymphnodes and a discharging sinus in the right groin. Respiratory system, abdomen and gynaecological examination were normal.

She was clinically diagnosed as a case of lymphoedema vulva and confirmed histologically by a skin biopsy from the lesion which showed acanthosis, multiple dilated lymph vessels lined by a single layer of endothelium in upper dermis, downward growth of epidermis, a lympho-plasmocytic inflammatory infiltrate and absence of any giant cells or atypical cells (Fig.2). Fine needle aspiration cytology (FNAC)

of lymph node was inconclusive. Polymerase chain reaction (PCR) for mycobacterium tuberculosis was negative. Biopsy of cervical lymph node showed destruction of lymphnode architecture, epitheloid cell granulomas and Langhan's type of giant cells, consistent with tubercular lymphadenitis (Fig. 3). Other investigations were erythrocyte sedimentation rate of 84mm/1st hour, negative blood test for microfilariae, normal ultrasound study of abdomen and pelvis and a negative enzyme-linked immunosorbent assay (ELISA) test for human immunodeficiency virus antibody. She was finally diagnosed as lymphoedema vulva with tubercular lymphadenopathy and put on anti tubercular therapy (ATT). After eight months of standard ATT the sinus over the groin healed, lymphnodes regressed, but lymphoedema of the vulva persists and plastic / reconstructive surgical consultation is being sought.

Discussion

Lymphoedema can be either primary (Milroy's disease/Praecox /Tarda) or secondary due to filariasis, tuberculosis, malignancy or iatrogenic. Fibrosis of the lymphatics leading to esthiomene following incision and drainage procedure of inguinal lymph nodes in

Fig. 1 : Firm, hyperkeratotic glossy papules and swollen vulva (20 x H & E stain).

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Fig. 2 : Skin biopsy showing multiple dilated lymph vessels lined by a single layer of endothelium in upper dermis with acanthosis and downward growth of epidermis (50 x H & E stain).

lymphogranuloma venereum (LGV) bubos was common before the advent of antibiotics. The iatrogenic causes are radiotherapy and radical mastectomy. More often lymphoedema is a result of treatment, either surgical excision of draining lymphnodes and / or radiotherapy [3]. Lymphoedema of the vulva can be a postoperative complication after vulvectomy and inguino femoral lymphadenectomy [4].

Vulval lymphoedema can be a rare extra-intestinal manifestation of Crohn's disease [5,6] but the ulcer in such cases shows epitheloid and giant cell granulomas. All these features were absent in this case. Clinically it may mimic genital warts [7] which was ruled out. Lymphangioma circumscriptum [8] 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 [9] but lacks true thick walled vessels. Though our case was clinically and histopathologically confirmed to be lymphoedema of the vulva yet the primary cause was difficult to establish. There was no clinical/ epidemiological evidence to suggest LGV. Filariasis was ruled out since there was no associated swelling of the legs or lymphangitis and blood was negative for microfilaria. PCR for mycobacterium tuberculosis was negative. However the lymph node biopsy established the tubercular aetiology. Lymphangiography was not considered at this stage since it is generally reserved for preoperative evaluation [3].

To conclude, this case was primarily a manifestation of tubercular lymphnode infection causing fibrosis, destruction and blockage of bilateral inguinal lymphnodes

Fig. 3 : Cervical lymph node biopsy showing epitheloid cell granulomas and Langhan's giant cells (50 x H & E stain).

leading to bilateral lymphoedema of the vulva and secondarily aggravated iatrogenically by multiple incisions of the left inguinal lymph nodes.

Conflicts of Interest

None identified

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