Surgical Correction of Peno-Scrotal Elephantiasis Nostras Verrucosa

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Abstract: Peno-scrotalelephantiasis nostras verrucosa is debilitating syndrome consisting of long standing lymphoedema of the penis and scrotum of non-filarial origin, cutaneous features and occasional inguinal lymphadenopathy which precede the condition.
A 36 year old man presented with several years history of genital swelling that occurred after developing inguinal lymphadenopathy from suspected lymphogranuloma venerum. Patient suffered inter-departmental shutting with missed diagnoses of first lupus vulgaris and then condylomata acuminata of the penis and scrotum. Following series of incisional biopsy reports, initially unfolding lupus vulgaris, then condylomata acuminata and finally elephantiasis nostras verrucosa, patient had scrotectomy, fulguration of penile shaft lesion, orchidopexy and reconstruction of a neo-scrotum. Penoscorpul elephantiasis nostras verrucosa is scarcely reported in the literatures. Surgical correction is the main stay of treatment including excision of the scrotum, penile shaft skin and reconstruction of a neo-scrotum and penile shaft resurfacing.

Keywords: scrotal elephantiasis, neo-scrotum, elephantiasis nostras verrucosa.

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I. Introduction

Lymphoedema is the accumulation of protein rich lymphatic fluid in the tissue spaces following abnormality of the lymphatic drainage system. The abnormality may be congenital in origin referred to as primary lymphoedema or acquired called secondary lymphoedema. The abnormality may be at the nodal basin or the lymphatic system. The penis has two lymphatic systems namely, the superficial system and the deep lymphatic system. The superficial system drains the prepuce and the skin of the penis and empties into the superomedial nodes of the superficial inguinal nodes, while the deep lymphatics drain the glans and lying deep to the deep fascia and drain into the external iliac lymph nodes, then into the pelvic and para-aortic group of nodes. The scrotal lymph drainage is different from that to the testes. It drains into the superficial inguinal node of the same side of the body.

Adenopathies of the superficial inguinal nodes and or deep inguinal nodes may contribute to the development of peno-scrotal lymphoedema. The cause of the adenopathies may be due to infection with *chlamydia trachomatis* resulting in lymphogranuloma venerum, *mycobacteriumtuberculosis*¹, chronic perineal skin infection with or without subdermal abscesses ²,³, infestation by microfilarial, *Wuchereria bancrofti*, Brugia malayi and *Brugia timori*¹ ² other causes include iatrogenic cause, primary or secondary malignancies. Lymphoedema may affect the lower limbs-commonest site, upper limbs, the genitalia (penis, scrotum, labia), the face and other localized part of the body⁴.

Brunner⁵ graded lymphoedema into :
Latent: Subclinical, no clinically apparent lymphoedema
Grade I: Pitting oedema which more or less disappears on elevation of the limb.
Grade II: Non pitting oedema occurs which does not reduce on elevation
Grade III: Oedema with irreversible skin changes like fibrosis, papillae, fissuring

II. Case Report

Mr. A. E, a 36 years old truck driver, was referred from the Dermatology clinic to the Burns, Plastic and Reconstructive Outpatient clinic of the University of Calabar Teaching Hospital, Calabar. He presented with a history of peno-scrotal swelling of more than 10 years duration. The swelling preceded a history of hard boil-like lesion on his left groin. This progressively increase in size and number on the same side and subsequently...
involved the right inguinal region. The boil like lesions broke down with discharge of pus. The lesions on each side soon coalesced forming an ulcer with indurated surrounding. This was followed by peno-scrotal swelling and the appearance of rashes on the scrotum. The swelling was painful but not incapacitating except that he experienced decreased libido. The patient confirmed a history of multiple sexual partners. There was no fever, no associated lower urinary tract symptoms and no previous history of sexually transmitted infections.

Patient had visited a patient medicine shop for the above complaints where he was given Ampiclox capsules and Chymoral tablets. When patient did not have satisfactory outcome, he then presented at the General Surgical Outpatient where evaluation and biopsy suggested first lupus vulgaris and after several months of failed therapeutic trial of anti-Koch therapy, a revised diagnosis of condylomata accuminata was made. Many investigations were carried out including incisional biopsied specimens for confirmation.

Patient had therapeutic trail with anti-Kochs therapy but to no avail, then 5 fluorouracil/epinephrine gel; all these gave no remarkable improvement. Patient then left orthodox treatment for herbal treatment for a period of about six months. Following his return to the hospital, he was offered cryotherapy which he never had.

Based on the above he was referred to the Plastic team of the hospital. Patient had no child. Drank alcohol occasionally but did not use tobacco in any form.

Examination at the Burns & Plastic clinic revealed a young man obviously looking worried, not quite ill looking and had normal vital signs.

Other systems were unremarkable except the urogenital system which showed peno-scrotal swelling with multiple discrete dyschromic verrucous nodules and plagues on the entire scrotum, root and shaft of penis and the entire scrotum [Figure 1].

![Fig 1: preoperative clinical photograph](image)

Digital rectal examination showed good peri-anal hygiene, no obvious skin tags and no fissures. Patient had normal sphincteric tone and no masses per rectum. A repeat of the investigations were done. Full blood count and differentials, fasting blood sugar and urinalysis were all normal. A retroviral serology test and VDRL screening were non-reactive. A biopsy specimen requested by the Dermatologist confirmed diagnosis of *elephantiasis nostras verrucosa* of the penis and scrotum [see figure 3]. Patient was then offered fulguration of penile shaft lesion, scrotectomy, orchidopexy and reconstruction of neo-scrotum [Figure 2]. Patient became well and was happy post-operatively.
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Fig 2: postoperative clinical photograph
III. Discussion

Elephantiasis etymologically is coined from the Greek word *elephas*. Originally used to describe a kind of leprosy in which the skin took on the appearance of an elephant’s hide.

Elephantiasis is a grade III lymphoedema, a non-pitting oedema with irreversible skin changes like fibrosis, verrucous papillae, nodules, and fissuring. It is the late stage of the condition. *Elephantiasis nostras verrucosa (ENV)* is a chronic grade III lymphoedema characterised by cutaneous changes such as dermal fibrosis, hyperkeratotic verrucous plaques, nodules and fissuring resulting from chronic inflammation of the resulting lymphoedema.

The exact cause is unknown. However, it may manifest as a complication of many disease conditions. They include chronic bacterial skin infection, lymphogranuloma venereum, mycobacteria tuberculosis infection, syphilitic and mycotic infections as well as trauma, malignancies, irradiation, chronic venous stasis, and metabolic conditions such as thyroid dermopathies, systemic sclerosis.

The condition may affect any dependent part of the body with chronic lymphoedema. The lower limbs are most frequently affected. Other parts of the body that may be affected include upper limb, the abdomen, the face, the genitalia, the buttocks and the lower back. *Elephantiasis nostras verrucosa* of the penis and scrotum is a rare occurrence in our sub-region unlike filariatic elephantiasis, also known as *classic elephantiasis tropica*. The cause of the secondary lymphoedema here is lymphatic system obstruction by microfilarial parasites. The index patient had a history that was initially suggestive of *lymphogranuloma venerum*. Two serial biopsies by two subspecialties consecutively upheld the diagnosis of lupus vulgaris and condylomata acuminate respectively. A third histology report confirmed features of epidermal acanthosis and papillomatosis with few scattered koilocytes and wild dermal fibrosis. After defaulting treatment due to non-satisfaction of treatment outcome a third biopsy was requested before referral to Plastic clinic. The outcome of the histology report indicated feature of epidermal acanthosis, papillomatosis, in-addition to conspicuously dilated lymphatics in the papillary dermis along with deep dermal fibrosis. These features are consistent with the diagnosis of *elephantiasis nostras verrucosa*.

The management of this condition present a reconstructive challenge. It is aimed at improving cosmesis or treatment of the symptoms. For the index patient, the scrotal skin and subcutaneous tissues were involved in the pathology in addition to some part of the penile skin. Only the testicles and spermatic cords were preserved.

To improve cosmesis, patient had total scrotectomy and fulguration of the penile skin having hyperkeratotic verrucous nodules. A Neo-scrotum was reconstructed using unilateralpostero-medial thigh flap and orchidopexy was done to prevent torsion. The fulgurated lesions on the penis healed by re-epithelialization. The patient got well and was very satisfied with the outcome other treatment. Other treatmentmodalities that has been used successfully in the treatment of *elephantiasis nostras verrucosa* include excision and skin grafting, treatment with carbon dioxide laser therapy and shunt procedures in combination with either of the above.
IV. Conclusion

The occurrence of classical elephantiasis tropica caused by filariatic elephantiasis is common in the sub-Saharan Africa, whereas elephantiasis nostras verrucosa which is of non-filarial origin is rather rare. The condition present a reconstructive challenge. Management is aimed at improving patient’s quality of life including cosmetic appearance. The index patient suffered peno-scrotal elephantiasis nostras verrucosa. The management included scotectomy, orchidopexy and reconstruction of a neoscrotum. This greatly improved patient’s quality of life. Therefore, Surgery is the mainstay of management.

References
