Giant Scrotal And Penile Elephantiasis Of Idiopathic Etiology: A Case Report

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Citation

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Abstract

Background: Scrotal lymphedema is condition though common in endemic
filariasis regions in Africa and Asia but rare out side these region. It is of variable origin in the western world.

Case presentation: We present a case of a 30-year-old man with massive elephantiasis of the scrotum and penis. The patient underwent subtotal scrotectomy saving penis, testes and spermatic cords and followed by scrotal reconstruction with adequate cosmetic and functional outcome.

Conclusion: In this report we discuss a rare case of giant scrotal and penile elephantiasis, reflect on the etiology and the diagnostic and therapeutic approaches. Surgery can be successful even in giant scrotal elephantiasis.

INTRODUCTION

Lymphedema of penis and scrotum is rarely seen in countries in which filariasis is not endemic. The abnormal accumulation of lymphatic fluid in subcutaneous tissue of penis and scrotum could lead to swelling, pain, dysuria, and sexual dysfunction (impotency and erectile dysfunction). Lymphedema may be idiopathic or secondary to inflammation, surgery, malignancies, trauma, radiation, hypoproteinemia, and other medical disorders.

Primary lymphedema i.e. congenital elephantiasis is an extremely rare condition. Patients develop edema at adolescence without restriction to the external genitalia. In hereditary elephantiasis of the Meige type, lymphedema of the external genitalia occurs due to malformation of lymphatic vessels.

Lymphatic obstruction is limited to penis and scrotum and is not seen in adjacent organs such as lower extremities, abdomen, and buttock. [1] Regardless of the cause of scrotal and penile elephantiasis, this disease can lead to physical and spiritual weakness and its treatment is difficult particularly in the aged. [2]

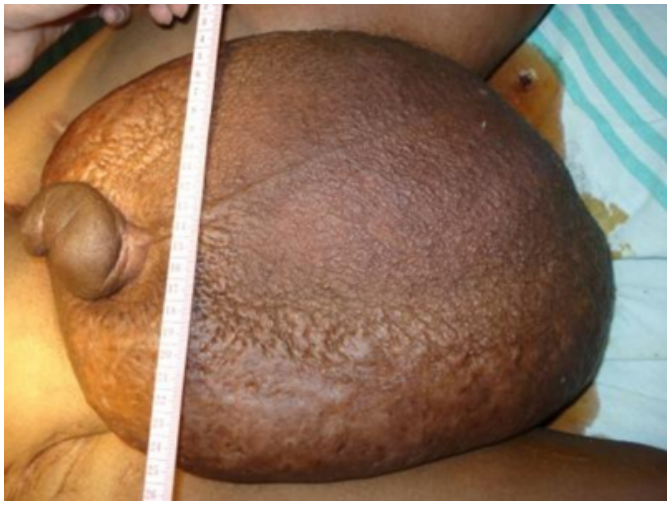
CASE HISTORY

A 30-year-old men was referred to our hospital because of painless swelling of penis and scrotum (fig. 1). He reported a history of swelling since six year, which had been intensified during the past two year. No history of irradiation, surgery, trauma, or infection was reported.

On examination, the patient had a massively enlarged scrotum extending up to his knees. The huge solid verrucous scrotal mass of 30 × 30 × 25 cm made it impossible to differentiate the anatomic structures. The scrotal skin was thickened and edematous hiding the penis. (Figure1). The penis was also edematous and disfigured and urethral orifice emerged as a deep pit on the anterior surface of the mass. No inguinal adenopathy was found. Mild lower extremities edema was present. Other systems were normal. The testes and cords were not palpable, but no abnormalities were shown by ultrasonography.

Figure 1

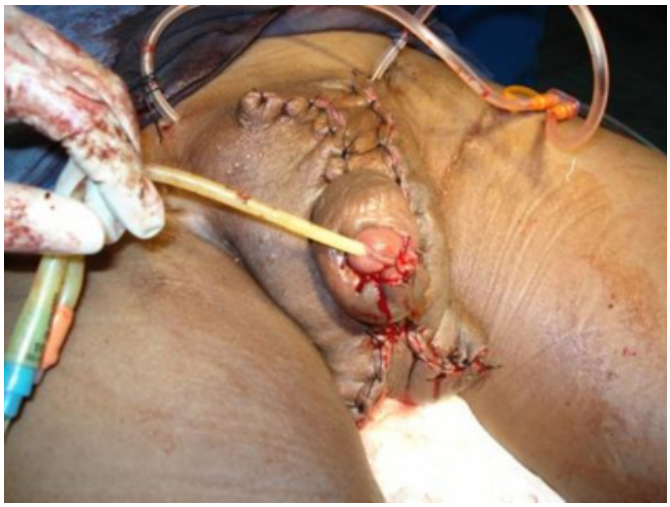
Figure 1: Giant scrotal and penile lymphedema showing edematous and thickened scrotal skin with disfigured penis



Ultrasonography of testes and lower urinary tract were normal. CBC, Urine routine and microscopy, ESR and blood biochemical tests were normal. Serologic study was negative for filariasis. With the diagnosis of idiopathic elephantiasis of penis and scrotum, surgery was planned in which extensive debridement of the involved tissue, scrotoplasty by the use of skin flaps of 1/3 of the posterior scrotum, and Z plasty by applying extra skin of penis region to repair its cover were to be performed (fig. 2). The excised scrotal tissue weighed 9 kg. Grossly, the specimen contained multiple fluid-filled cysts. Histopathologic examination showed nonspecific chronic inflammation with areas of epidermal thickening and dermal fibrosis. Wound was healed primarily and suture was removed on 14th postoperative day.

Figure 2

Figure 2: Extensive debridement with scrotoplasty.



DISCUSSION

Genital elephantiasis is mostly developed in tropical regions and is a rare condition outside regions endemic for Chlamydia trachomatis or Wuchereria bancrofti. Degreef believes that about 20% of males in tropical regions develop penile and scrotal elephantiasis. [3]

Lymphedema has two types: Primary and secondary. Primary lymphedema is subdivided into three categories: 1. congenital-inherited (Milory syndrome), 2.praecox (with early onset), and 3.tarda (with late onset) [1]

Secondary lymphedema has four subtypes: 1.obstructive (secondary to neoplasm, radiation, surgical intervention, mechanical trauma, and chemical agent's injection), 2.Inflammatory (parasitic, bacterial, and fungal infections), 3.Phlebitis, and 4.Angioneurotics. [4]

Penile and scrotal lymphedema mostly occurs following an infection or as a reaction to trauma. Idiopathic lymphedema is rarely seen and is caused by a primary obstruction of lymphatic vessels of scrotum. [1]

No effective medical treatment has been introduced; however in patient with spotty occurrence of genital lymphedema may respond to prolonged course of fluoroquinolones Different surgical methods for the treatment of chronic genital lymphedema have been reported in the literature. Two main methods are as follows:

1. Physiologic methods or lymphangioplasty through which lymphatic discharge from involved regions to new lymphatic channels is obtained.

2. Lymphangiectomy with reconstructive surgery. [2]

Lymphangioplasty is used in the cases of recurrent lymphedema; however, this method can not be successful in the cases of chronic fibrosis or lymphedema caused by radiation because of the lack of appropriate lymphatic channels. [2]

Lymphangiectomy includes the removal of superficial lymphatic network, which is located above the Buck's fascia which is derived from median raphae and prepuce lymphatics. These lymphatics drain to superficial posterior lymphatic channels. A deeper system is located beneath the Buck's fascia and is drained into deep inguinal lymph nodes.[4]

This method of drainage leads to the success of this surgical method. It is essential to remove involved skin and subcutaneous tissue completely (reduction scrotoplasty) to prevent lymphedema recurrence followed by reconstructive surgery of penis and scrotum.[5,6] The posterior scrotal skin usually uninvolved and use as a source of skin flap for reconstruction of scrotum. Different surgical techniques are used in lymphangiectomy and repair of penis and scrotum which include Jourdan and Meller,[1] Dlepech,[2,5] Larrey,[5] Cadogan and Anderson,[7] Raghaviah,[8] Vaught,[9] Dandapat,[3,5] Morey,[10] Apesos,[2] and Malloy.[1]

Surgical complications of elephantiasis or genital lymphedema include hemorrhage, hematoma, urethral injury, infection, painful erection, decrease of sensation, and scar in suture line. These complications could be reduced by using a proper incision, use of Z plasty instead of longitudinal suture, separating of testes and cord by an external incision in scrotum before taking any measure, and removal of involved tissue.[2,3]

CONCLUSION

Genital lymphedema is a rare syndrome mostly caused by Chlamydia trachomatis or Wuchereria boncrofti. It can be a part of the constellation of idiopathic lymphedema of lower extremity. We presented a patient with giant scrotal lymphedema due to idiopathic cause and discuss diagnostic and therapeutic approach. In the case presented here extensive excision of elephantoid tissue saving penis, spermatic cord and testes was performed with adequate cosmetic and functional results.

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