Idiopathic Scrotal Elephantiasis

Mustafa Kürşat Evrenos, Merve Özkaya, Murat Yaman, Levent Yoleri
Department of Plastic, Reconstructive and Aesthetic Surgery, Celal Bayar University School of Medicine, Manisa, Turkey

INTRODUCTION

Lymphedema is a condition caused by extra-cellular fluids accumulating in subcutaneous compartments as a result of lymphatic obstruction. Etiologically it can be classified as primary (congenital) and secondary (acquired). Primary lymphedema can be seen to be associated with congenital lymphatic hypoplasia, as a component of a genetic syndrome such as Milroy’s disease or Meigs syndrome. Secondary lymphedema is usually seen following an obstruction. The most common cause of secondary lymphedema in the world is filariasis, but the number of lymphedema cases secondary to radiotherapy and lymph node dissections is also rapidly increasing. In males, genital lymphedema can be isolated to the penis or the scrotum, or affects both. This condition can be either idiopathic, or occur secondarily to a surgery, trauma, radiation, tumor or infection in lymph nodes where the major lymphatics of the genital region are drained. Once the flow of lymphatic fluid is disrupted, a pathophysiologic process that disrupts cellular migration in the immune system is initiated with the fluid accumulating in the distal tissues containing macromolecules and proteins. This results in cellular expansion, loss of elasticity, and a tendency to infection. Lymphatic fibrosis develops in the region. Thus, recurrent erysipelas, lymphatic cysts, and lymphuria, giant mass decubitus ulcers, and hygienic problems are frequently encountered in enlarged genital organs. Additionally, micturition difficulty and sexual dysfunction are fundamental issues impacting a reduced quality of life.

In this study we present a very rarely seen idiopathic case of isolated massive scrotal lymphedema. This case is of significance in that it demonstrates that an isolated lymphedema can grow into massive sizes and lead to partial systemic disorders even if the patient has not undergone surgery.

In April 2014 a 49-year old male patient applied to our clinic with a long-standing lump in the genital region. Patient anamnesis revealed a history of slow-progress for about 15 years. The swelling in the scrotal region had occurred in the absence of any tumor, infection, surgery, nor had the patient traveled abroad. The patient did not describe any accompanying pain and congenitally had one kidney. No particular aspects were identified in his familial history. The patient had applied to several external clinics since the beginning of the swelling, but had discontinued seeking medical help when no medical or surgical treatment was initiated. The patient had become bedridden when the slow-growing mass started to hinder his ability to walk or stand erect without support in the past five years.

In the physical examination the meatus of the penis could not be discriminated on the 70 x 60 x 40 cm scrotum. His testicles were not palpable. The skin on the left half of the mass had a pervasive verrucous appearance. To the anteroinferior of the lesion a necrotic zone of about 3 x 2 cm and hyperemia advancing towards the superior were observed and assessed in favor of cellulitis due to local temperature increase (Figure 1). The patient was found to be in good general health with stable vitals. No micturition problems were described. The patient’s laboratory findings reported deep anemia (hemoglobin: 6.6 g/dL). Hematologic examination reported anemia (hemoglobin: 6.6 g/dL). Hematologic examination reported an appearance consistent with bilaterally localized scrotal lymphedema. The testicles could not be evaluated. The patient was scheduled for reduction and reconstruction with local flaps. The patient was informed about the planned procedure and a possible orchiectomy, but he did not consent to the surgery and was voluntarily discharged following local infection control.

DISCUSSION

Lymphedema can be diagnosed based on a careful anamnesis and the findings from physical examination. Imaging methods corresponding to the clinical findings should be used to describe its pathophysiology and to determine the treatment plan. Even if MRI is not effectively used in identifying its etiology, pre- and postoperative edema bears importance in a comparative assessment. McDougal has suggested a new staging system in isolated external genital lymphedema. This system defines the cases by stages based on whether they are congenital or acquired, sporadic or inherited, and on the age of the patient at the time of its occurrence. Based on McDougal’s staging system, our case can be described as ‘idiopathic scrotal elephantiasis’. Morbid obesity and past urinary system infections which can trigger inflammatory processes have been blamed in etiology. Nevertheless our case did...
and by advancing the foreskin to the proximal.1,7,10,12 In scrotal lymphedema cases consist of chronic occurrences. In cases affecting the penis-although not a conclusive concept in treatment-penis reconstructions are performed using skin grafting7,9,10 and local flaps from surrounding healthy tissues. The patient, however, did not consent to a surgical procedure and was voluntarily discharged. As it stated in the article; patient did not accept surgery and sign informed consent surgery form and he was discharged at his own request after medical treatment.

Informed Consent: As it stated in the article; patient did not accept surgery and sign informed consent surgery form and he was discharged at his own request after medical treatment.

Figure 1. View of massive scrotal lymphedema in supine position of the immobilized patient

not present a morbid obesity profile. There were no precise data concerning past urinary system infections in our patient, however, his urine analysis and culture examination did not reveal infection. Surgery is the treatment in cases that present such massive occurrences. Except for two extensive series1,7 a report describing six cases5 and a number of case reports are available in the literature.6,8-16 Reports on isolated penoscrotal lymphedema cases consist of chronic occurrences. In cases affecting the penis-although not a conclusive concept in treatment-penis reconstructions are performed using skin grafting7,9,10 and local flaps from surrounding healthy tissues and by advancing the foreskin to the proximal.1,7,10,12 In scrotal reconstruction, skin flaps taken from the superior or the lateral are used following orchiopexy. In cases which this option is not possible, fasciocutaneous transposition or advancement flaps taken from medial thighs are used.9,16 Reported early postoperative complications include hematoma and opening of the wound, and long-term complications include chronic scrotal sinus.1,7 Given the size of the mass and the deep anemia of chronic disease secondary to frequent infections due to genital hygiene issues, surgery was the treatment proposed to our patient. While there are cases in the literature that report sedimentation and high CRP5, no cases are reported that describe concomitant deep anemia of chronic disease. In our case, the wound was stabilized following antibiotic therapy and regional debridement. The patient was informed about orchiopexy and a possible orchiectomy, and an operation was planned for reconstructing of the defect using flaps taken from the healthy surrounding tissues. The patient, however, did not consent to a surgical procedure and was voluntarily discharged.

Pervasive and massive scrotal lymphedema is a condition that can significantly impact the life quality of patients both physically and psychologically. As seen in our case it can also lead to severe secondary conditions. The increasing number of radiotherapy treatments and lymph node dissections performed today lead to the higher prevalence of this condition. These patients should be treated with a multidisciplinary approach by an experienced medical team including the plastic surgeon.

**REFERENCES**