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Elephantiasis Nostras Verrucosa. Excision with full-thickness skin grafting of the penis, scrotum, and perineal area

Nathan Judge, Ali Kilic

University of Alabama at Birmingham, Division of Plastic Surgery, Birmingham, AL 35294, USA.

Corresponding author:

Ali Kilic, MD

Assistant Professor

University of Alabama at Birmingham

Division of Plastic Surgery

510 20th St South FOT

Birmingham, AL 35294

E-mail: *akilic@uabmc.edu*

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Abstract

Background: Elephantiasis nostras verrucosa is a rare cutaneous complication of chronic lymphatic obstruction. It is most commonly caused by bacterial infection, trauma, neoplasia, obesity, and venous stasis.

Main observations: In this report, we describe a case of elephantiasis nostras verrucosa involving the scrotum and perineal area in a 32-year-old. The lesions were excised, and full-thickness skin grafting of the penis, scrotum, and perineal skin was performed.

Conclusion: This case demonstrates the efficacy of excision with full-thickness skin grafting of the penis, scrotum, and perineal area in a patient with elephantiasis nostras verrucosa confined to the scrotum and perineal region. (*J Dermatol Case Rep*. 2016; 10(2): 32-34)

Introduction

Elephantiasis nostras verrucosa (ENV) is a rare condition that arises in the setting of chronic, non-filarial lymphedema.[1] Common etiologies of the lymphedema include bacterial infection, trauma, neoplasia, obesity, and chronic venous stasis.[2] Chronic lymphatic obstruction leads to dermal fibrosis and enlargement of the affected body part. Cutaneous changes seen in ENV include non-pitting edema, hyperkeratosis, papillomatosis, and verrucous lesions.[3] In this report, we describe a case of ENV involving the scrotum and perineal region.

Case Report

The patient is a 32-year-old man with a past medical history significant for von Willebrand disease and hypertension, who presented to clinic with a 10-year history of multiple scrotal and penile skin lesions, scrotal elephantiasis, and chronic drainage (Fig. 1). His past surgical history was significant for splenectomy and cholecystectomy. The patient denied any trauma, surgery or infection to groin area. A diagnosis of elephantiasis nostras verrucosa was made based on the results of a skin biopsy.

Complete removal of scrotal, partial penile skin lesion excision, and right inguinal orchiectomy were performed, along with a full-thickness skin graft of the penis, scrotum, and perineal skin. The post-operative period was uneventful, and the skin graft took very well (Fig. 2).

Discussion

Elephantiasis, the massive enlargement of a body part, results from chronic lymphedema and is sub-classified based on the etiology of the initial lymphatic obstruction.[1] The most common form is elephantiasis tropica, which is due

Figure 1

The patient presented with multiple scrotal and penile skin lesions, scrotal elephantiasis, and chronic drainage.

Figure 2

Postoperative 1 month. Diseased scrotal and penile skin is removed and the defect is skin grafted.

to filarial infection. The term "elephantiasis nostras" was initially introduced by Castellani in 1934 to differentiate filarial elephantiasis from elephantiasis of bacterial origin.[2] "Nostras" means "of our region" (temperate zone), and "verrucosa" describes the verrucous cutaneous changes seen in the later stages of elephantiasis nostras.[1] Although elephantiasis nostras was originally described as due to recurrent bacterial infection, the definition has since been expanded to include all non-filarial forms of elephantiasis.[3]

The pathogenesis of ENV involves chronic lymphatic obstruction, which causes lymphostasis and accumulation of protein-rich fluid in the dermis. The protein-rich fluid induces fibroblast proliferation and blunts local immune responses.[3] Fibroblast proliferation leads to fibrosis of the dermis and subcutaneous tissue, and impaired local immune response predisposes to recurring lymphangitis. Inflammation associated with recurrent lymphangitis promotes further fibrosis.[3]

ENV can occur in any location in which chronic lymphostasis is present. The most common sites of involvement are the lower legs and feet, but it may also rarely involve the face, ear, upper extremities, abdomen, and scrotum.[3] Early in the course of the condition, recurrent lymphangitis is accompanied by enlargement of the affected area and pitting edema.[4] Progressive fibrosis leads to chronic non-pitting edema, and the epidermis develops a cobblestone-like appearance.[5] In the late stages of the condition, verrucous lesions develop and give the skin a mossy appearance.[5] Histological characteristics of ENV include pseudoepitheliomatous hyperplasia and dermal fibrosis.[3] The diagnosis of ENV is made based on clinical history, physical examination, and biopsy findings.

Management of ENV is difficult, and there is recurrence in the vast majority of cases. To date, there have been no large, long-term studies of ENV treatment, and there is no standard of care.[3] The primary treatment goal is reducing lymphatic obstruction.[3] Conservative options include weight reduction and use of compression stockings.[3] Several case reports have suggested that oral retinoids may have some efficacy in improving cutaneous changes and reducing lymphedema.[7] Surgical intervention is often a viable option for ENV cases refractory to conservative and medical therapy.[7] Surgical debridement has been shown to produce some esthetic improvement; however, it does not correct the underlying lymphatic obstruction.[3] Lesion excision has been successful for some large ENV lesions, but recurrence is nearly inevitable.[7-9]

Conclusion

This case is significant because we were unable to find any published case reports of elephantiasis nostras verrucosa confined to the scrotum and perineal area. The patient did very well after the surgery and was pleased with the result.

References

1. Routh HB. Elephantiasis. *Int J Dermatol*. 1992; 31: 845-852. PMID: 1478760.
2. Kalafi A, Vasaghi A. Elephantiasis nostras verrucosa of lower limb: a case report. *Acta Med Iran*. 2014; 52: 786-787. PMID: 25369015.
3. Sisto K, Khachemoune A. Elephantiasis nostras verrucosa: a review. *Am J Clin Dermatol*. 2008: 9: 141-146. PMID: 18429642.
4. Castellani A. Elephantiasis nostras. *J Trop Med Hyg*. 1934; 37: 257-264.
5. Rowley MJ, Rapini RP. Elephantiasis nostras. *Cutis*. 1992; 49: 91-96. PMID: 1563290.
6. Castellani A. Researches on elephantiasis nostras and elephantiasis tropica with special regard to their initial stage of recurring lymphangitis (Lymphangitis recurrens elephantogenica). *J Trop Med Hyg*. 1969; 72: 89-97. PMID: 5769718.
7. Pitcher AA, Pagan CA, Small K, Otterburn DM. Excision of elephantiasis nostras verrucosa lesions in a patient with hereditary lymphedema: case report and review of the literature. *J Foot Ankle Surg*. 2015; 54: 747-750. PMID: 25441846.
8. Manupati S, Ahmed FZ. Elephantiasis nostras verrucosa secondary to congestive cardiac failure. *BMJ Case Rep*. 2016 Mar 4; 2016. pii: bcr2016214720. doi: 10.1136/bcr-2016-214720. PMID: 26944379.
9. Hotta E, Asai J, Okuzawa Y, Hanada K, Nomiyama T, Takenaka H, Katoh N. Verrucous lesions arising in lymphedema and diabetic neuropathy: Elephantiasis nostras verrucosa or verrucous skin lesions on the feet of patients with diabetic neuropathy? *J Dermatol*. 2016; 43: 329-331. PMID: 26281791.