Scroto-perineal hidradenitis suppurativa complicated by giant scrotal elephantiasis

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INTRODUCTION

Scrotal elephantiasis has been a recognized complication of inguinal node irradiation, filarial infection, tumor involvement and surgical lymphatic destruction, but has rarely been reported in association with hidradenitis suppurativa (HS). HS, also known as acne inversa, is a chronic and often debilitating disease primarily affecting the axillae, inframammary regions and perineum. The location of the lesions may lead to social embarrassment and failure to seek medical treatment. Scroto-perineal HS complicated by scrotal elephantiasis is a distressing disease. Excisional surgery with reconstruction is the recommended treatment with a high likelihood of good outcome. We present a 38-year-old male patient with long-standing scroto-perineal HS complicated by giant scrotal elephantiasis.

CASE REPORT

A 38-year-old male patient (single, smoker) who had been working in the security sector and living in the eastern province of the kingdom of Saudi Arabia was admitted with massively enlarged scrotum. The condition first appeared 5 years previously, with scattered painful nodules on the scrotum and inner thighs. Although initially discrete, the nodules became progressively matted and swollen with purulent discharges. Before his presentation to our emergency department, the patient had been seen by a dermatologist and urologist and was given courses of oral antibiotics and topical ointments. Because of the lesion locations, the patient felt embarrassed and did not seek further medical treatment. The patient's scrotum gradually increased in size until he was walking with difficulty and ultimately became bedbound for the 2-year period prior to his admission.

Clinical examination revealed massive scrotal elephantiasis almost reaching the knees and totally engulfing the penis. Urine was noted as coming from a deep pit on the dorsal aspect of the scrotum. Testis and spermatic cords were not palpable. The skin of the suprapubic area, both groins, adjacent inner sides of the thighs, scrotum and scrotoperineum were covered with thick, indurated, scarred, non-pitting nodules, without...
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crepitus and with a few scattered sinuses that were discharging pus, resulting in poor hygiene and offensive smell [Figure 1]. Both axillae were also involved, but to a lesser extent.

All routine blood investigations were normal, with the exceptions of the microcytic hypochromic anemia, schistosomal and filarial tests, which were negative. *Staphylococcus aureas* could be cultured from pus swabs.

A computerized tomography scan of the chest, abdomen and pelvis showed marked extensive scrotal swelling with skin thickening and subcutaneous fat stranding, as well as pelvic and inguinal lymphadenopathy. Similar results were noted in both axillae, suggesting an infectious process.

A multidisciplinary investigative approach was employed before beginning definitive treatment. Dermatology, infectious diseases, psychosocial, plastic and colorectal surgeries were all consulted. Pre-operatively, two units of packed red blood cell were transfused and second-generation cephalosporin antibiotic treatment was initiated. Informed consent was obtained and an intensive care unit bed was assigned. Under general anesthesia and in the supine position, the patient initially underwent laparoscopic diverting colostomy followed by wide excision of the involved areas (suprapubic, groins, scrotum, inner thighs and premium) and a suprapubic catheter was inserted. Both spermatic cords and testis were found, with the preserved testis buried beneath the subcutaneous fat. The penis was degloved as its skin was involved in the infection; however, the corpora and urethra were preserved by careful dissection during surgery. A plastic surgeon was involved, including the use of a vacuum-assisted therapy machine followed by immediate dressing. After a few weeks, split-skin grafting was applied to the raw areas and skin cover was achieved. Complete excision of this condition is necessary in order to cure the disease. The disease involves the hairy and moist areas and the infection that accompanies it is deeper than perceived. Because of the chronicity of the wound, treatment options including flaps are restricted. The intention of the operation was to excise the scrotum and reconstruct it using appropriate modality. According to the reconstructive ladder, split-skin graft comes below the local and regional flaps. Also, because of the nature of the disease, local flaps from the involved areas were not an option without spreading disease further. A free flap would have been an overkill and fraught with difficulties. As a result of this, the simplest possible and most effective reconstructive option was used successfully.

The suprapubic catheter was removed, voided normally per urethra and the patient is due to have the colostomy closed. He is walking and experiencing a happy social life and positive self-image. Further plastic surgery is planned for ongoing axillary HS [Figure 2].

**DISCUSSION**

Scrotal elephantiasis is an unusual presentation and, although elephantiasis has been a recognized complication of inguinal node irradiation, filarial infection, tumor involvement and surgical lymphatic destruction, it has been only rarely reported in association with HS. HS is initially characterized by the presence of tender subcutaneous nodules, which over time may rupture, resulting in painful, deep dermal abscesses. The lesions may be rounded, but, unlike furuncles, do not usually exhibit pointing. After rupture, the lesions often extrude a purulent, foul-smelling discharge. As the disease process continues, fibrosis, dermal contractures and induration of the skin may occur. HS typically occurs in the axillary, inguinal, perianal, perineal, mammary and inframammary regions, with the axilla being the most commonly affected site. Perianal HS is associated with more debilitating outcomes. Long thought to be a disorder of apocrine origin, it is
now thought to be caused by follicular occlusion. Families with an autosomal dominant type inheritance have been reported. Hyperandrogenism is unlikely to play a role in HS. Bacteria are probably secondary colonizers, which may exacerbate HS, but are not considered primary etiologic agents. Poor hygiene, smoking and obesity are not primary causes of HS but are strongly associated with the disease and may exacerbate it. HS patients experience a quality of life worse than those with alopecia, mild to moderate psoriasis and several other dermatologic conditions, and results in economic and psychological disability in addition to physical limitation. Complications of scrotal HS include urethral strictures and fistulas, contractures and limb mobility limitations, cutaneous squamous cell carcinoma and anemia.

Death was reported in a male patient with perineal/perianal HS complicated by severe hypochromic anemia, hypoalbuminemia, hypergammaglobulinemia, amyloidosis and fistulas to the rectum, urethra, urinary bladder and peritoneum. There is no uniformly effective single therapy for HS. Therefore, clinicians will likely try an array of treatment modalities depending on the patient’s disease. For patients with extensive disease, wide excision can dramatically improve the patients’ quality of life. A similar case of scrotal elephantiasis secondary to HS was reported by Konety et al. (1969) and was managed with wide local excision and split-thickness skin grafting, yielding an excellent cosmetic and functional outcome.

For patients with extensive disease, a multidisciplinary approach is advised. Various surgical methods for treatment of HS have been described. Wide local excision with skin grafting, skin flap transfer, primary closure and fasciocutaneous and musculocutaneous flaps have all been applied for HS treatment according to the area affected. In our case, wide excision of extensive involved skin areas was necessary, making primary closure impossible. Split-skin grafting was applied resulting in a good cosmetic, functional and uncomplicated outcome [Figure 3]. Although rare, it is important to remember that this gross deformity, caused by profound scarring of the lymphatic, is a potential consequence of untreated, chronic inguinal HS, leading to elephantiasis that can result in severe psychological disturbances, social isolation and functional impairment. CONCLUSION

Scrotal elephantiasis secondary to HS is a challenging disease for both the patient and the physician. It is a chronic relapsing inflammatory skin process that can be extensive, causing both physical and psychosocial distress to the patient. A multidisciplinary approach with excisional surgery and appropriate reconstructive procedure is recommended to achieve a good outcome.

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How to cite this article: Alharbi B, Shlash A, Bedaiwi K, Hooti QA, Almohaisen A, Shlash S, Said M. Scrotal-perineal hidradenitis suppurativa complicated by giant scrotal elephantiasis. Urol Ann 2015;7:265-7.
Source of Support: Nil, Conflict of Interest: None.