An Unusual Case Report of Genital Elephantiasis Nostras Verrucosa

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ABSTRACT:
Elephantiasis nostras verrucosa is an unusual cutaneous manifestation of chronic nonfilarial lymphedema. It features warty, leathery, or verrucous hyperkeratotic plaques with a cobblestone appearance on nonpitting edematous skin. This condition predominantly affects the lower extremities. Herein, we present a case of elephantiasis nostras verrucosa with an uncommon presentation in the genitalia, accompanied by a superficial fungal infection.
Key words: Genitalia, Elephantiasis nostras verrucosa

Introduction

Chronic lymphedema, whether of primary (congenital) or secondary (acquired) origin, can eventually lead to massive swelling and verrucous skin thickening. This condition is known as elephantiasis nostras verrucosa (ENV), elephantiasis verrucosa nostrum, or simply elephantiasis nostras[1]. It manifests with hyperkeratosis and papillomatosis of the epidermis, alongside superimposed hyperkeratotic papulonodules that render a distinct cobblestone-like skin appearance[2]. ENV can cause persistent deformity, functional impairment, localized skin and tissue infections, septicemia, and the rare Stewart-Treves syndrome, a cutaneous angiosarcoma with a grim prognosis[3]. We report a case of an elderly female who developed genital ENV as a secondary complication of intraabdominal surgery concomitant with radiotherapy.

Case report

A 72-year-old Thai woman presented with gradually progressive, well-defined, confluent erythematous plaques with a cobblestone appearance on the perineum, gluteal cleft, and inner thighs over 5 years. She described mild pain and itching at the affected sites. Eight years prior, she was diagnosed with middle rectal adenocarcinoma, which had invaded the myometrium of the uterus (stage T4N1M0). She underwent low anterior resection with transabdominal hysterectomy and concurrent chemotherapy and radiotherapy. Three years later, she gradually developed radiation proctitis, chronic diarrhea, and the skin lesions. There was no history of exposure to tropical infections, recurrent cellulitis, erysipelas, soft tissue infection, lymphangitis, or familial lymphedema.

Physical examination revealed bilateral, malodorous, well-defined, macerated, verrucous, and erythematous plaques with a cobblestone appearance. These plaques, covered by yellowish crusts, were located on the perineum, gluteal cleft, and inner thighs (Figure 1). No lymphadenopathy or lower limb edema was observed. From the patient's history and physical examination, the differential diagnoses include lymphatic filariasis, chromoblastomycosis, papular mucinosis (lichen myxedematosus), papillomatosis cutis carcinoides, and Stewart Treves syndrome.

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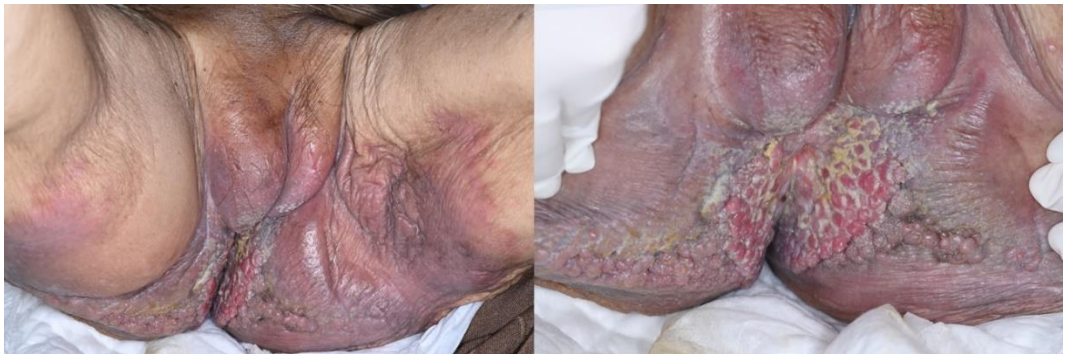

Figure 1 A 72-year-old female patient with classic skin findings of genital elephantiasis nostras verrucosa, including an admixture of plaques, papules, and nodules with a cobblestoned and mossy appearance on the perineum, gluteal cleft, and inner thighs of the patient

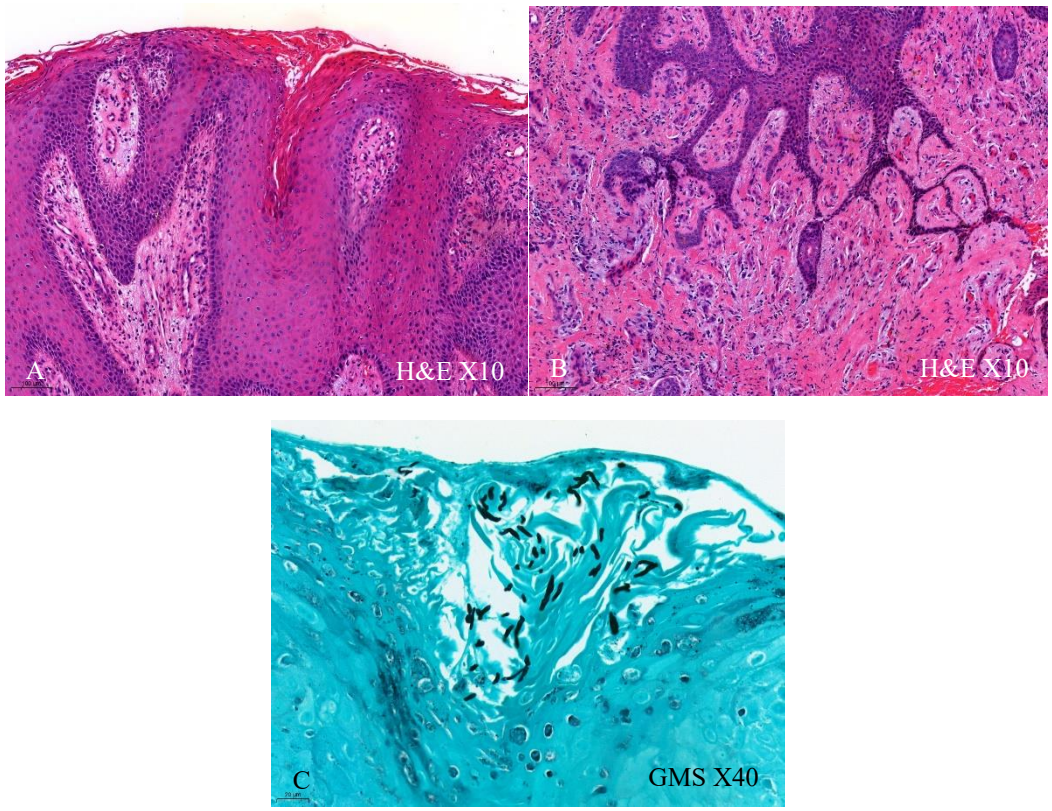

Figure 2 (A-B) Histopathology findings of markedly acanthotic epidermis with papillomatosis (pseudoepitheliomatous hyperplasia) and the presence of proliferative capillary vessels in the papillary and upper reticular dermis. Dermal fibrosis with increased stromal fibroblasts was present. Epidermal dysplasia was not identified. (C) Gomori methenamine silver (GMS) highlighted budding yeasts with pseudohyphae in parakeratosis. (A, B: H&E X10; C: GMS X40)

The initial potassium hydroxide examination of the lesions revealed oval budding yeasts with pseudohyphae. An incisional skin biopsy of the perineal lesion demonstrated a markedly acanthotic epidermis with papillomatosis (pseudoepitheliomatous hyperplasia) and proliferative capillary vessels in the papillary and upper reticular dermis (Figure 2A-B). Dermal fibrosis with increased stromal fibroblasts was also observed. No epidermal dysplasia was noted. These findings were suggestive of lymphedema. Gomori methenamine silver staining highlighted budding yeasts with pseudohyphae in parakeratosis (Figure 2C). Tissue culture for fungus identified *Candida albicans*. The patient was diagnosed with genital ENV concurrent with a superficial fungal infection. Computed tomography of the abdomen indicated evidence of a previous low anterior resection with a patent colorectal anastomosis, enterocolic anastomosis, and side-to-side jejunojejunostomy. A few subcentimeter lymph nodes were observed in the paraaortic and both iliac regions, measuring up to 0.6 cm in size.

The therapeutic goals were to alleviate discomfort, prevent physical disability, and monitor for rectal cancer recurrence. Conservative treatments involved using compressive dressings and maintaining cleanliness and dryness in the affected area. To treat the superficial fungal infection, topical clotrimazole cream and oral fluconazole 200 mg weekly for 4 weeks were prescribed until clinical and mycological improvement was achieved. Concurrently, the underlying malignancy was closely monitored through measurements of serum carcinoembryonic antigen levels, computed tomography imaging, and colonoscopy. No evidence of recurrence was observed.

Discussion

ENV is a severe and rare cutaneous manifestation of nonfilarial chronic lymphedema[1]. Patients often have a protracted history of edema before the onset of fibrosis, hyperkeratosis papules, and verrucous lesions[3]. ENV is often observed in dependent body parts, particularly the lower extremities[1,3]. However, any region with chronic lymphedema, including the upper extremities, abdomen, and scrotum, can be affected[1,3]. Filariasis and cancer treatment are the most prevalent causes of secondary lymphedema cases[1]. The literature has documented only a few cases of genital ENV[4,5]. Our patient's atypical genital ENV resulted from surgical and radiation interventions for advanced-stage rectal adenocarcinoma. Impaired lymphatic drainage following surgical lymph node dissection or excision, coupled with radiation therapy, can lead to the loss of dermal lymphatic vessels and nodal fibrosis, obstructing lymphatic regeneration[1,6]. Lymphedema after oncological treatments usually manifests as unilateral limb swelling, but gynecological cancer surgeries can cause bilateral swelling[7]. If not properly managed, lymphedema can progress to complications such as chronic ulcers, deformity, infection, and even secondary malignancies such as Stewart-Treves syndrome[3].

For ENV diagnosis, a comprehensive medical history, physical examination, and tissue biopsy to verify histopathological characteristics are essential[3,8]. A lesion biopsy is necessary to exclude other etiologies or concurrent diseases such as cutaneous malignancy or lymphangiosarcoma (Stewart-Treves syndrome). It is crucial to thoroughly evaluate patients suspected of having ENV to firmly exclude the presence of such conditions[3,9].

Treatment of ENV is challenging. Initial management to decrease lymph accumulation includes skin hygiene, limb elevation, weight reduction, compression dressing, mechanical massage, bandages, compression stockings, and pneumatic pumps. Nonresponders to conservative therapies may require surgical

intervention[3,10]. In our patient with genital ENV, mechanical compression was not feasible. Hence, the therapy goals were to ease discomfort, prevent physical impairment, eradicate the superimposed infection, and closely monitor rectal cancer recurrence. The patient continues to utilize conservative measures, but minimal skin improvement has been observed. Additionally, patient education and psychosocial support are paramount[3].

Conclusion

We highlight a case of ENV in an elderly female occurring in an uncommon area, the genitalia. This significant condition developed due to persistent lymphedema. While a thorough history and physical examination are often sufficient for diagnosing ENV, histopathology and other imaging investigations may be necessary to exclude other disorders. Disease management remains challenging. Treatment should focus on reducing lymphostasis, which will improve cutaneous changes and prevent recurrent infections.

Disclosure

The authors report no conflicts of interest in this work.

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