Elephantiasis nostras verrucosa associated with chronic constipation in a low-weight patient

Zayıf bir hastada kronik konstipasyon ile ilişkili elefantiyazis nostras verrükoza

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Abstract

Elephantiasis nostras verrucosa (ENV) is a chronic, progressive disease closely associated with obesity. We report a weak 51-year-old male patient with a long history of constipation due to Hirschsprung’s disease, a decrease in venous return due to prolonged abdominal distention during ileus attacks, and eventually a severe ENV clinic. Abdominal tomography of the patient shows that the expanded column segments compressed the iliac veins.

Keywords: Elephantiasis, stasis dermatitis, constipation, Hirschprung disease, social withdrawal

Öz

Elefantiyazis nostras verrükoza (ENV), obezite ile yakından ilişkili kronik, ilerleyici bir hastalıktır. Burada, Hirschsprung hastalığı nedeniyle uzun zamandır kabızlığı olan hasta ileus atakları sırasında uzun süren abdominal distansiyon gelişmesi nedeniyle venöz dönüş azalma ve nihayetinde ciddi düzeyde ENV kliniği gelişti. Hastanın abdominal tomografisinde genişletilmiş kol segmentlerinin iliac venleri sıkıştığı görülmektedir.

Anahtar Kelimeler: Elefantiyazis, staz dermatitis, kabızlık, Hirschprung hastalığı, sosyal geri çekilme

Introduction

Elephantiasis nostras verrucosa (ENV) is a rare, chronic, progressive, relapsing disease characterized by non-filarial lymphedema. ENV, seen mainly with bilateral leg involvement, is a disorder characterized by papules and verrucous lesions. Obesity and soft tissue infection are the preponderant risk factors of ENV. Herein, we described a severe ENV case arising from chronic constipation despite low-weight, which is the first case in literature to our knowledge.

Case Report

A 51-year-old male patient was admitted with profoundly malodorous, hyperkeratotic, purulent, painful lesions on the legs, especially marked on the left leg. Patient who did not care for personal hygiene had been brought to our hospital by his relatives after the patient’s legs began to smell malodorous. His complaints initiated at the age of 37 years with swelling and wounds on the left leg. Similar complaints of the right leg occurred over time. The patient had been treated with systemic and topical antibiotics hitherto when his complaints...
of pain, swelling, wound appeared. He had undergone Hirschsprung’s disease. His medical history was negative for filariasis, familial Milroy’s disease, surgery, radiation, neoplasia. He had never been abroad. He wasn’t obese with body mass index (BMI) at 16.6 kg/m\(^2\). In the examination, there was diameter difference between the legs, non-pitting edema on the bilateral lower limbs covered with yellow-brown, thick hyperkeratosis and cobblestone-like appearance under the peeled crusts, especially substantial over the nearly whole extensor aspect of left leg while there were similar plaques with peripheral ecchymosis on the medial malleolus and middle of the leg on the right side (Figure 1). There was no reflection with the Wood’s light examination carried out to exclude \textit{Pseudomonas} pyoderma. The remainder of the examination was normal.

We conducted multiple punch biopsy, tuberculosis, fungal and bacterial cultures from the skin due to considering ENV, blastomycosis-like pyoderma, tuberculosis verrucosa cutis, chromoblastomycosis, verrucous/squamous cell carcinoma in the differential diagnosis. Intravenous antibiotic (ampisilin-sulbaktam) was administered empirically just after admission. In addition, potassium permanganate bath mitigated his pain and bad smell within one week and the patient’s thick yellow-brown crusts intermittently underwent debridement (Figure 2).

A comprehensive blood examination did not show any abnormalities other than sedimentation (48 mm/hr normal range: 0-20 mm/hr) and C-reactive protein (8.2 mg/dL normal range: 0-5 mg/dL). Occult blood in the stool was negative as well. The area of induration was measured at a diameter of 10 mm. \textit{Staphylococcus aureus}, sensitive to aminopenicillins, was isolated from culture obtained purulent lesion areas. No fungus and tuberculosis bacillus were grown.

Examination of histopathologic sections revealed orthokeratosis, irregular acanthosis, focal parakeratosis, mild spongiosis in the epidermis as well as acute and chronic stasis findings such as extravascular erythrocytes and hemosiderin-laden macrophages, fibro-myxoid stroma around proliferating vascular structures in the dermis. There was no dermal invasion and atypical mitosis. Periodic acid-Schiff and Gomori methenamine silver staining showed no fungal agent (Figure 3). Although deep vein thrombosis and arterial pathology were not detected in the arterial and venous doppler ultrasonography, all of the deep venous structures, particularly on the left, had venous insufficiency findings and tortuous varicose veins were observed. We diagnosed the patient with ENV based on clinical, histopathological manifestations. Complaints involved in ENV are flared up during periods when the patient had long-term constipation. We speculated that the primary cause of venous insufficiency may be long lasting episodes of decreasing venous return due to the frequent recurrent abdominal distension. Indeed, abdominal computed tomography (CT) showed that the sigmoid colon was 25x160 mm and all colon segments were substantially dilated. Expanded column segments compressed the stomach, bladder and bilateral iliac veins (Figure 4).

A thorough interview revealed that the patient had been socially inactive for a long time due to his skin lesion and frequent stomachache. He had not shown his legs to even the closest relatives and refused to go to the hospital. The patient was diagnosed with Hirschsprung’s disease at 20 years of age and had been treated 31 years ago. But thereafter his constipation problem has not improved because of possible secondary stricture after the operation or incomplete surgical margin. Still, he had sought no medical advice hitherto. We could observe that a severe ileus clinic developed in the patient who could defecate once every 2 weeks and had difficulty in gas extraction; his abdominal distention progressed over time and he had severe abdominal pain attacks during admission to our clinic.

The patient whose complaints regressed by antibiotherapy, except for ileus was referred to general surgery after abdominal CT since he had no defecation for more than 2 weeks, vomiting, intermittent fever and colic abdominal pains. The patient who improved by a conservative approach in the department of general surgery refused a surgical intervention to solve his constipation. So, general constipation recommendations, enema and laxative treatments were arranged and he was discharged. Informed consent was obtained.

![Figure 1. Bilateral lower limbs covered with yellow-brown, thick hyperkeratosis and cobblestone-like appearance under the peeled crusts, in particular, marked on the left side and diameter difference between the legs](image1)

![Figure 2. 7th day of treatment (crusts continue to be removed)](image2)
Discussion

ENV is characterized by papules, verrucous lesions which deform the affected area with time. Although the etiopathogenesis of ENV remains unclear, the most important risk factor is morbid obesity and soft-tissue infection and chronic venous insufficiency. Whatever the etiology of elephantiasis is, the skin lesions are formed by a common pathogenesis. There is a vicious cycle in which the secondary bacterial infection and chronic venous insufficiency feeds each other.

According to his suggestion, recurrent secondary infections due to disturbed lymphatic drainage will cause the process to a vicious cycle as in the present case. The affected tissue thickens by lymphatic pooling, and the skin barrier feed each other.

Here, we presented a patient with extended constipation due to Hirschsprung’s disease. Constipation resulting in expanded sigmoid colon can produce venous insufficiency and even ENV development. In addition, we underscored that the concurrent social isolation in our patient was another possible risk factor of ENV. To our knowledge, this is the first such case in the literature.

Interestingly, psychiatric conditions characterized by social withdrawal such as social anxiety disorder, depression are risk factors for ENV because of propensity to venous stasis by prolonging immobilized duration and contributing to constipation. We think that it is the invisible part of the iceberg that our patient has also a social withdrawal, which is another factor contributing to the progression of the disease.

ENV is a rare entity which can result from chronic constipation. The presence of the other risk factors can make the picture even worse. Here, we presented a patient with extended-constipation due to Hirschsprung’s disease. Constipation resulting in expanded sigmoid colon can produce venous insufficiency and even ENV development. In addition, we underscored that the concurrent social isolation in our patient was another possible risk factor of ENV. To our knowledge, this is the first such case in the literature.

Ethics

Informed Consent: It was obtained.
Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: Ç.T., I.G.İ., S.A., PÇ., H.M.E., Concept: Ç.T., H.M.E., Design: Ç.T., I.G.İ., S.A., PÇ., H.M.E., Data Collection or
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