Elephantiasis Nostras Verrucosa: A Case Report of Effective Management with Complete Decongestive Therapy

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Elephantiasis nostras verrucosa describes the cutaneous changes including dermal fibrosis, hyperkeratotic, verrucous, and papillomatous lesions that occur after chronic secondary, nonfilarial lymphedema. The treatment goal is to reduce lymph stasis and avoid further complications. We report a case of elephantiasis nostras verrucosa without adequate care before effectively managed by simple treatment. Other differential diagnoses and non-invasive complete decongestive therapy (CDT) were also discussed. (Dermatol Sinica 23: 228-232, 2005)

Key words: Elephantiasis nostras verrucosa, Lymphedema

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INTRODUCTION

Elephantiasis nostras verrucosa describes a rare group of cutaneous changes including dermal fibrosis, hyperkeratotic, verrucous, and papillomatous lesions that occur after chronic secondary, nonfilarial lymphedema.\(^1\) We report a patient with elephantiasis nostras verrucosa who was successfully treated by elastic bandage compression.

CASE REPORT

An 86-year-old man came to us in September, 2004 because of recalcitrant lesions with non-pitting edema of both lower legs and dorsal feet. Beginning six months earlier, he developed multiple vesicles on bilateral dorsal feet up to lower legs with severe itching. Multiple flesh-colored papules ensued and gradually became woody cobblestone-like lesions.\(^{\text{Fig. 1}}\)

For two years, he had been wheelchair-bound with minimal ambulatory activity due to glaucoma-related poor vision of 20 years and severe pain from longstanding bilateral ingrowing toe nails. When the skin lesion became weeping with foul smelling, the family took him to several institutions and was wandering from one service to another without noticeable improvement. He denied history of trauma, febrile illness, surgery, foreign travel and family history of lymphedema.

We admitted him to our inpatient unit and performed work-ups. The blood smear revealed no filariasis or peripheral eosinophilia. Duplex ultrasound examination of bilateral lower legs failed to disclose obvious reflux or obstruction of superficial and deep venous system. Chest X-rays and tumor markers were negative or within normal limits. Lymphoscintigraphy didn’t show lymphatic system obstruction or abnormality in bilateral lower limbs and lower abdominal cavity. Skin biopsy taken from lower leg demonstrated various degrees of hyperkeratosis and acanthosis with flattened rete ridge.\(^{\text{Fig. 3A}}\) The chronic edematous change and wide-spaced collagen with focal fibroplasia was significant in dermis. There were numerous ectatic and dilated lymphatic capillaries.\(^{\text{Fig. 3B}}\) We elected to manage his problems with external graduated compression and applied short-stretch bandages from distal feet to above knees for whole day and changed daily. We also encouraged him to elevate legs, and increase ambulation. In addition, erosions with lymphorhea were treated with topical Neomycin-Bacitracin ointment and steroid ointment on cobblestone-like lesions for pruritus. Within three days, the edema of bilateral lower legs improved dramatically and

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Fig. 1

(A) Dorsal feet swelling with serous discharge at right foot. Severe onychomycosis with ingrowing toe nails are noted.\(^{\text{B}}\) Profound, generalized papillomatous lesions at bilateral calves.\(^{\text{C}}\) Erosions with wooden thickened skin.\(^{\text{D}}\) Close view of cobblestone-like lesions with yellowish lymphorhea of right calf.

Fig. 2

(A) After treatment for 10 month, onychomycosis of toe nails improved a lot.\(^{\text{B}}\) The cobblestone-like lesions disappeared.\(^{\text{C}}\) Lower limbs return to usual size and texture.\(^{\text{D}}\) Some post-inflammatory hyperpigmentation and mild xerosis of right calf.
serous discharge diminished. The skin changes of his legs continued to improve. We also removed his ingrowing nails and treated onychomycosis. Follow-up visit 10 months after discharge from hospital, his legs remained in good condition and the quality of life was much better for him and his family. (Fig.2)

**DISCUSSION**

The most important observation in our case is that dermatology, with simple regimen, effectively manages a recalcitrant condition and significantly improves the qualities of life of our patient as well as his family. For 6 months, he wandered through various medical and surgical services and often was told "not much can be done". Dermatology should play more active roles in treating elephantiasis nostras verrucosa.

Elephantiasis nostras verrucosa vividly describes the cutaneous changes of chronic lymphedema: marked edema, thickening of the skin, with cobblestone-like and verrucous appearance. It occurs mostly on bilateral lower limbs, and occasionally on ear, lip, penis, and abdomen.\(^2\)\(^-\)\(^10\) Progression of lymphedema, primary or secondary, increases protein concentration in the tissue and results in chronic inflammation with subsequent cell proliferation. (Table 1.)\(^11\) It leads to fibrosis in the edematous tissue and dilatation of the afferent lymph vessels.\(^12\) The pressure generated by lymphatic contractions constitutes the main force for lymph flow.\(^13\) Uptake of lymph is facilitated by local arterial pulsation, skeletal muscle contraction, and intrinsic, intermittent tissue movement.

In order to give proper treatment, clinical history (hereditary, infection, trauma, surgery history, or radiation), physical examination (non-pitting edema and vesicles or bullae related to chronic lymphedema\(^14\)) and laboratory data should be reviewed carefully. Skin biopsy, Duplex-Doppler examination (including venous insufficiency or vascular malformation), lymphangiography, lymphoscintigraphy\(^15\),\(^16\) and even computed tomography, MRI (to rule out lymph system obstruction or malignancy)\(^17\) are helpful for making diagnosis.

What causes lymphedema in our patient? The work-up was negative for lymph system obstruction or malignancy. He had diabetes, glaucoma and ingrowing nails without adequate treatment for years. Due to prolong immobilization with secondary inflammation process, the vicious cycle further caused severe lymphedema and lymphorhea.

The goal of treating elephantiasis nostras verrucosa is to reduce lymph stasis. Complete decongestive therapy (CDT) plays a key role. First introduced by Foldi and Foldi\(^18\) and further modified by Casley-Smiths,\(^19\) CDT consists of

![Fig. 3](image)

(3) Histology showed hyperkeratosis, various degrees of acanthosis with elongated and flattened rete ridges. (H&E X100)(B) Numerous ectatic lymphatic channels at upper dermis. (H & E, 400X)

**Table 1. Primary and secondary lymphedema**

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<th>Primary</th>
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<tr>
<td>Congenital: Milroy’s disease</td>
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<td>Idiopathic: Praecox, Tardum, Variant with yellow nails and pleural effusions</td>
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<th>Secondary</th>
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<tr>
<td>Infection: Bacteria (Streptococcus infection), Filariasis, Bartonella</td>
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<tr>
<td>Trauma</td>
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<tr>
<td>Surgery</td>
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<tr>
<td>Malignancy</td>
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<tr>
<td>Fibrosis: Radiation, Stasis, Localized myxedema, Panniculitis, Idiopathic or pharmacological crotoperitoneal fibrosis</td>
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two parts (Table 2.) Phase I includes manual lymph massage and drainage, proper motion exercises, elevation of affected limbs, nail care, and use of multilayered short stretch graduated bandages or stockings. The phase II emphasizes daily-wearing fitted compression garments, self-manual lymph massage and special designed exercise to reduce edema. The ideal bandaging system provides high working pressures and relatively low resting pressures to safely remove lymphedema without compromising microcirculation. The working pressure produces when the muscle pump works against the resistance of the bandage, as when exercising. Inelastic and short stretch bandages are better because they can provide higher working pressure and greater muscle pump efficiency than other bandage garments. Conversely, because of the low resting pressure (pressure exerted when the muscle is inactive and relaxed), compression bandages may be worn 24 hours with good patient compliance. The pressure induced by compression is defined by Laplace’s law: \( P = \frac{T}{R} \). The pressure (P) exerted by an elastic bandage is proportional to the tension of the bandage (T) and the inverse of the radius of the skin surface area (R). Therefore, pressure on ankle should be monitored carefully due to its smallest radius that bears higher pressure. CDT has been widely available in Europe for many years and was introduced to other parts of the world as well, especially for breast cancer patients who developed lymphedema after radiation or surgical therapy.

In our case, the phase I therapy started with the care of skin and nail to eradicate infection. The next step was to use multilayered short stretch bandages compression. Consistent compression evidently reduces limb size, and pressures should not be greater than 60 mmHg, as suggested by some authors, to avoid injuring lymphatic vessels. However, there is no consensus on the most appropriate amount of compression that should be provided. It is important to keep the balance between the effect and patient's compliance. We adjust the bandage pressure day by day according to patient's current condition and make sure it is not too tight or too loose. Short stretch bandages that are applied at full stretch are less likely to overcompress the limbs. We applied the bandages in a straight spiral at full stretch with 50% overlap on patient's bilateral lower limbs from feet to above knees. (Fig. 4) Compression should be completed using rolled pads over concave areas, as retromalleolar gutters. Afterwards, the phase

<table>
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<th>Table 2. Complete decongestive therapy</th>
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<td><strong>Phase I: Treatment – 1 to 4 weeks</strong></td>
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<tr>
<td>• Meticulous skin and nail care</td>
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<td>• Manual lymphatic drainage</td>
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<td>• Low-stretch multilayer bandaging</td>
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<td>• Physical therapy in bandages</td>
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<tr>
<td><strong>Phase II: Maintenance</strong></td>
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<tr>
<td>• Meticulous skin and nail care</td>
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<tr>
<td>• Low-stretch multilayer bandages worn overnight</td>
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<tr>
<td>• Prescribed exercises in bandages [Surgical support garments (30-50mm Hg) for ongoing control]</td>
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Fig. 4

(A) Check pulse of dorsal pedis first, than keep foot at over-extension position. (B) Wrap bandage completely over the foot, then angle the bandage upward toward the ankle at extension position. (C) According to Laplace’s law, the pressure on ankle shouldn't be too tight. Use one finger to make some space for wrapping. Rolled pads may be used over concave areas. (D) Keep the bandage in a straight spiral at full stretch with 50% overlap on patient's leg and continue on up to above knee.
II basically focuses on the maintenance. Wearing compression stockings with adequate exercise until the edema resolved. In fact, the dynamic pressure of bandage compression is difficult to monitor, and it may vary due to experience and training. The compression hosiery can provide more stabilized graduated pressure and easy for self-use. It is mandatory to educate patient and family for home management and continuation of treatment. Our patient kept wearing compression hosiery daily for 4 months and stopped in January, 2005. Currently, his legs are still in good conditions and he takes aerobic exercise one hour everyday. Comparing with drug or surgical intervention, CDT is non-invasive, inexpensive and satisfactory.

REFERENCES
Multiple Painful Erythematous Papuloplaques and Pustules on Limbs with High Fever in a 59-year-old Woman

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CASE REPORT

A 59-year-old woman was admitted to our infectious disease section due to acute onset of multiple painful erythematous skin rash and high fever (>38.5°C) 10 days ago. The laboratory examination revealed leukocytosis (33550/Cumm) with neutrophilia (76%), elevated ESR (43mm/hr) and increased CRP (4.50mg/dL).

We were consulted because the high fever and skin rash persisted without improvement after 2 days of antibiotics treatment. The clinical examination showed multiple tender erythematous papuloplaques with pustules formation on bilateral palms, knees and lower legs (Fig 1, 2). An incisional biopsy from palmar area was performed (Fig 3). Besides, recurrent oral aphthae for 1 year and poor-healing perineal ulcers were complained by the patient (Fig 4).

Moreover, both viral and bacterial cultures of the blood and skin tissue were negative and immunoglobulin, complement and tumor markers levels were all within normal limits.
DIAGNOSIS: Behçet’s Disease with Sweet’s Syndrome-like Presentation

DISCUSSION

Behçet's disease (BD) is an inflammatory disease of the vascular and mucocutaneous system that primarily affects young adults. The disease is characterized by recurrent oral ulcers, genital ulcers, and skin lesions, as well as retinal vasculitis and aseptic meningitis. The disease is often associated with other systemic manifestations such as arthritis, uveitis, and neurological involvement.

In the given case report, the patient presented with a Sweet’s syndrome-like presentation. Sweet’s syndrome is a febrile neutrophilic dermatosis associated with monoclonal gammopathy, often seen in patients with lymphoproliferative disorders. The patient also had a positive pathergy test, which is a characteristic feature of BD.

The patient was diagnosed with BD andSweet’s syndrome (SS) based on the typical clinical features and the presence of pathergy. SS is characterized by the sudden onset of fever, rash, and arthritis. The rash is typically tender and may be associated with periarteritis nodosa (Pancoast’s syndrome).

The differential diagnosis for the patient's presentation includes other vasculitides such as leukocytoclastic vasculitis (LCV), which can cause skin lesions similar to those seen in SS. LCV is characterized by a leucocytic response in the vessels of the skin and mucous membranes.

The treatment for SS typically includes systemic corticosteroids, and in severe cases, other immunosuppressive agents such as methotrexate or ciclosporin may be used.

REFERENCES