CASE REPORT

Elephantiasis nostras verrucosa: a rare complication of lower limb lymphoedema

Neoh Kar Keong,^{1,2} Angelina Tang Siing Ng,¹ Pubalan Muniandy,³ Wong Voon Fei^{1,2}¹Department of Internal Medicine, Sibul Hospital, Sibul, Sarawak, Malaysia²Faculty of Medicine, SEGI University³Department of Dermatology, Sarawak General Hospital, Kuching, Sarawak, Malaysia**Correspondence to**
Dr Neoh Kar Keong,
kkneoh@yahoo.com

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SUMMARY

We reported a case of a 49-year-old man who had chronic lymphoedema of bilateral lower limbs for 30 years, but he did not seek treatment. His disease was complicated with irreversible changes of elephantiasis nostras verrucosa and had recurrent admissions due to infection. He was not keen for surgical intervention. This case report illustrated a rare and non-reversible complication of a common skin disease, lymphoedema and also the importance of identifying and treating it early.

BACKGROUND

Elephantiasis nostras verrucosa (ENV) is a rare and progressive dermatological disease characterised by a thickened and fibrotic skin. It is a non-filariasis chronic lymphoedema, causing disfigurement of the extremities, and it will lead to recurrent infections and disabilities. There is no standardised guideline and treatment to date. By writing this up, we hope to raise awareness among the health practitioners that treating lymphoedema in early stage is important to prevent long-term complications.

CASE PRESENTATION

We described a case of a 49-year-old obese Chinese man with no known medical illness who had been under close observation for the bilateral lower limbs swelling and disfigurement for 2 years. The lesions started 30 years ago, initially described as skin thickening, worsening into nodules and thick plaque formation, but he did not seek treatment.

Clinical examination revealed that his bilateral lower limbs were severely disfigured, swollen with a cobblestone appearance up to the distal third of the tibia, non-pitting in nature, with ulceration and crusting over the exposed skin surface (figure 1). There was lost of sensation over the hyperkeratotic skin of bilateral lower limbs.

The lower limbs had worsened recently and were complicated with a foul-smelling serous discharge (figure 2). A skin biopsy showed a benign fibroepithelial polyp with some reactive changes with no features of malignancy. His blood parameters and septic workups were normal, and a skin culture showed a mixed growth.

His functional status was severely impaired with a loss of ankle function, and required walking frames for ambulation. The lower limbs disfigurements had restricted his daily activities and he became unemployed and remained single. Limited activities had in turn worsened his lymphoedema.

OUTCOME AND FOLLOW-UP

He was admitted to hospital for six times in the past 2 years for recurrent cellulitis and lower limb pain. He was treated with multiple courses of antibiotic, including cloxacillin, amoxicillin-clavulanate and cefuroxime but showed no improvement. He was advised for an amputation of bilateral lower limbs in view of recurrent infections with a loss of ankle function. He refused to have any surgical intervention.

DISCUSSION

Lymphoedema is described as the accumulation of tissue fluid in the interstitium secondary to impaired lymphatic drainage. ENV is an extreme complication of chronic lymphoedema.

Lymphoedema occurs when the lymphatic load exceeds the transport capacity of the lymphatic system, and the excess fluid accumulates in the interstitium. Impaired lymphatic drainage will lead to a progressive enlargement and hypertrophy of the gravity dependent area. Recurrent infection and inflammation in turn worsen the vicious cycle of lymphoedema.

Lymphoedema can be classified into:

A. Primary lymphoedema: congenital (onset <2 years old), lymphoedema precox (onset between 2 and 35 years old) and lymphoedema tarda (onset after 35 years old). Primary lymphoedema shows an association with chromosomal abnormalities and familial distribution.

B. Secondary lymphoedema: consequence of disruption or obstruction of lymphatic drainage as a result of surgery or disease process. The common examples are blockage of lymph node secondary to lymph node dissection or neoplastic disease, surgery, direct trauma, radiation induced fibrosis and recurrent infections.¹

Moffatt's study in UK revealed that the crude prevalence of chronic lymphoedema in the UK community is 1.33/1000, in which it is more common in women than in men (2.15 vs 0.47/1000) and becomes more prevalent with increasing age (5.4/1000 in age >65 years).²

Lymphoedema is closely associated with malignancy. Cormier³ found out that the incidence of lymphoedema in malignancy patients is around 15.5%. Out of this, 16% are melanoma, 20% are gynaecological malignancies, 10% are genitourinary malignancies, 4% are head/neck malignancies and the remaining 30% are associated with sarcomas.³ Possible causes of malignancy-associated

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Figure 1 Cobblestone appearance of the bilateral lower limbs up to distal third of tibia, with cellulitis changes. Erysipelas changes were seen above the nodules.

Figure 2 Some of the nodules were ulcerated with foul-smelling serous discharge. Part of the toes are embedded.

lymphoedema are tumour obstructions of lymphatic channels, malignant infiltration of lymphatic vessels, treatment related such as lymphatic dissection or irradiation.

Uncontrolled chronic lymphoedema will lead to ENV. There is no standardised criteria for the diagnosis of ENV. The diagnosis is made based on the patient's history, physical examination and characteristic skin changes.⁴

The classical histology of ENV showed hyperkeratosis of epidermis, loss of dermal papillae, fibrosis of the dermis and subcutaneous tissues and widened lymphatic vessels, which is similar to our case study patient.⁵

The International Society of Lymphology had used the criteria of 'softness/firmness' and volume difference after elevation to classify lymphoedema. Our case study patient was classified as having stage III disease as the skin changes were not reversible with elevation and his disease was complicated with elephantiasis.

The early management of chronic lymphoedema includes a multimodal therapy, which consists of skin care, weight reduction, compression therapy and physiotherapy. These measurements are helpful in all stages of lymphoedema.⁶ There was no strong evidence to indicate that pharmacotherapy benefits patients. One case study in Turkey reported that acitretin therapy was successful in the resolution of ENV.⁷

Mousavi⁸ suggested that surgical treatment should be considered after a trial of 6 months of conservative management. The principle of surgical interventions is to remove the excess tissue to decrease the volume of the extremity.⁸ Surgical procedures such as the removal of subcutaneous fat and fibrous tissue,

lymphatic surgery only showed a modest outcome and not sustained in case series.⁹

The reconstructive surgery was recently introduced with the aim to restore the lymphatic flow by anastomotic surgery or via indirect reconstruction to restore the lymphatic flow. However, it did not show a positive outcome compared with the old ablative surgery. Lee's¹⁰ study explained that the poor outcome was directly linked to 'timing' of surgical intervention as patients were often referred late in their disease, lack of medical expertise and the reconstructive operation was time consuming and technically demanding.¹⁰

Chronic lymphoedema results in multiple infectious complications such as cellulitis, erysipelas and lymphangitis. The protein-rich lymph is an excellent medium for the growth of bacteria.¹¹ The infection fares well with antibiotics, but it frequently recurs. A long-term follow-up is necessary as chronic lymphoedema may turn into malignancy such as lymphangiosarcoma. It also leads to psychological morbidity and a reduced quality of life.

Learning points

- ▶ Lymphoedema is described as the accumulation of tissue fluid in the interstitium secondary to impaired lymphatic drainage.
- ▶ Chronic lymphoedema leads to irreversible skin changes, recurrent infections and also malignancy changes.
- ▶ Interventions are more effective in early stage of disease.

In conclusion, lymphoedema is a chronic disease that is manageable but not curable. It is also difficult to be treated, especially if it has progressed to a late stage such as ENV. It is important to identify this disease early to prevent long-term morbidity.

Contributors NKK, ATSN and WVF: involved in patient's care in ward, data collection and case reporting. ATSN: involved in photo taking and explanation to patient. PM: involved in interpretation of data. PM and WVF: involved in literature review.

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