An overview of lipo-lymphoedema with co-morbid obesity and an example of management

T Vivian
Lymphatic Therapy Centre, Cape Town, South Africa
Correspondence to: info@occtherapy.co.za

Wound Healing Southern Africa 2019;12(2):10-14

According to the national health system in the United Kingdom,[1] lipoedema is a long-term (chronic) condition where there's an abnormal build-up of fat cells in the legs, thighs and buttocks, and sometimes in the arms. The condition usually affects women, although in rare cases it can also affect men. In lipoedema, the thighs, buttocks, lower legs, and at times the arms, become enlarged due to a build-up of abnormal fat cells. The accumulation of fat is bilateral and the feet and hands aren't affected, which creates a "bracelet" effect or "band-like" appearance just above the ankles and wrists. Leg and arm size can vary between individuals with lipoedema, and the condition can gradually get worse over time.

As well as becoming enlarged, affected areas of the body may feel soft, "doughy" and cold, bruise easily, ache or feel painful or tender or have small broken veins under the skin. Someone with lipoedema may eventually get lymphoedema in their tissues. This type of swelling can worsen by the end of the day and may improve overnight, whereas the fatty swelling of lipoedema is constant. In the later stages, lymphoedema becomes more prominent and this is when patients usually seek medical help.

Lymphoedema is still a puzzle in many parts of the world today. According to a study conducted by Blome et al. it took the patients listed in his study on average, "13.5 years from first symptoms to lymph drainage" and "13.7 years until compression therapy". In today's world of modern medicine and with the methods and rates of learning improving day upon day, we as medical practitioners have the opportunity to change the lives of person's living with lymphoedema. This article explores one woman's experience of lymphoedema and the tools she used to manage her life.

"Lymphedema is a chronic and potentially disabling condition which is associated with significant morbidity due to the functional, cosmetic and emotional consequences. It is caused by a dysfunction in the lymphatic system and is a progressive disease." Vojácková et al.[2]

Introduction

Schook et al.[3] suggested "Lymphedema can result from anomalous development (i.e., primary) or injury to lymph nodes or lymphatic vessels (i.e. secondary)." The condition is characterised by swelling of the limbs, face, genitals or less commonly the abdomen. Lymphoedematous tissue slowly enlarges over time because of the accumulation of subcutaneous lymph, which stimulates adipose deposition and fibrosis.[4] Untreated or incorrectly treated lymphoedema reduces quality of life and threatens the patient with complications.[2] Complications of lymphoedema include infection, functional disability, chronic cutaneous changes, and psychosocial morbidity.[4]

The pathogenesis of lymphoedema includes either primary lymphoedema or a secondary impairment of the lymphatic system. Primary lymphoedema is an inherited developmental disorder of lymphatic system and is medically termed lymphangiodysplasia. There are three types of primary lymphoedema namely lymphoedema congenitum, lymphoedema praecox and lymphoedema tardum. These types are determined by the age of onset of the disease.[3] Secondary lymphoedema is an acquired dysfunction where the lymphatic system was functioning normally before the onset of lymphoedema. It could be brought about through infection such as filariasis or other non-inflammatory conditions such as tumours, radiotherapy, lymph node excision surgery or traumatic injury.[3] Secondary lymphoedema is far less common with an incidence of approximately 1.15 per 100 000 persons younger than 20 years old.[4]

Contributing factors

Many factors have been studied with regard to lymphoedema risk such as obesity, weight gain, venous insufficiency and recurrent soft tissue infections. Chronic venous insufficiency often leads to venous ulcers which are further complicated when lymphoedema is present. This leads to delayed healing and can result in infection. Recent projections anticipate a continued rise in the prevalence of

obesity (especially female obesity), which will have an impact on the increased number of patients with lymphoedema.[5]

Signs and symptoms of lymphoedema

Lymphoedema is characterised by a high protein oedema in the tissues. It presents as an enlarged limb girth and generally begins distally and progresses proximally. A positive Stemmers sign is used to assist with diagnosis.

Stages of Lymphoedema: According to International Society of Lymphology, 2007[8]

Stage 0: This stage is also known as the subclinical, or pre-stage of lymphoedema. This term is used to classify those who are at risk of developing lymphoedema but have not yet developed any symptoms of the condition.

Stage 1: In this stage, lymphoedema can be greatly improved with early treatment. The swelling consists of protein-rich fluid. The tissues are soft to the touch, and pressure leaves an indentation that is known as pitting oedema.

Stage 2: This is also known as moderate lymphoedema. Swelling and fibrosis are both present, and the tissue is no longer soft to the touch. Stage 2 lymphoedema can be improved, but it requires intensive treatment.

Stage 3: This stage is also known as severe lymphoedema or elephantitis. The swelling and tissue fibrosis cause the skin to harden and lose its normal elasticity. These changes create folds of tissue that limit mobility and are disfiguring. There may be papillomas and hyperkeratosis.

The management of lymphoedema involves methods to reduce or delay the progression of swelling and to prevent associated wounds which can lead to infection. Methods include: Complete Decongestive Therapy, pneumatic pump therapy, kinesio-taping and surgical techniques. The approach used in this article focuses on Complete Decongestive Therapy (CDT). CDT makes use of external pressure such as short stretch bandages and compression garments, manual lymph drainage (MLD), exercise, meticulous skin and self-care and education to empower the patient.

As prevention of wounds with the associated risk of infection is included as a cornerstone in the treatment of lymphoedema, it is important to look closer at the lymphatic system's role in the development of one of the most common types of wound present with lymphoedema.

Lymphatics contribution to venous ulcers

The rising prevalence of elderly and lifestyle chronic conditions means that we as practitioners are going to be seeing an increasing number of persons with oedema-related conditions. The sequellae of untreated chronic oedema include increased swelling, chronic inflammation, skin changes, superficial ulceration, and lymphorrhoea.[6]

The clinical, aetiological, anatomical, and pathophysiological (CEAP) classification identifies active venous ulcers (C6) as the most advanced stage of chronic venous insufficiency (CVI).[7] Venous ulcers are often recurrent, may persist for years, and can result in repeated infections and cellulitis. In addition, venous ulcers have significant socioeconomic and psychological consequences reducing quality of life and increasing the burden on caregivers and the healthcare system.[9]

The mechanism of ulcer formation involves a series of pathophysiological steps that include (1) obstruction and/or reflux, (2) persistent venous hypertension, and (3) increased capillary filtration and interstitial fluid load. Typically, excess interstitial fluid is removed by the lymphatics, but if the fluid load overwhelms the lymphatic capacity or if the lymphatics are defective, the accumulation of interstitial fluid, macromolecules, and cytokines lead to oedema, breakdown of subcutaneous tissue, and formation of ulcers.[10]

In recent years there has been an emergence of evidence indicating the involvement of the lymphatic system in contributing to the development, healing and recurrence of venous ulcers. Although their contribution is recognised, treatment of the lymphatic system as part of the underlying pathophysiology is poorly reviewed in literature and is currently not being recommended as an adjunct therapy in golden standards of care. Recent advancements in near-infrared fluorescence lymphatic imaging (NIRFLI) technology provide opportunities for non-invasive, real-time assessment of lymphatic contribution in the aetiology and treatment of ulcers.[11]

In a study by Rasmussen et al.,[11] the researchers looked at lymphatic transport in patients with chronic venous insufficiency and venous leg ulcers following sequential pneumatic compression. They made use of intradermal micro doses of indocyanine green (ICG) as a lymphatic contrast agent to image the lymphatics of 12 subjects with active venous leg ulcers (Clinical, Etiologic, Anatomic, and Pathophysiologic [CEAP] C6). The lymphatics were imaged before and after a single session of pneumatic compression to assess impact on lymphatic function. The baseline imaging showed impaired lymphatic function and bilateral dermal backflow in all subjects with chronic venous insufficiency. There was proximal movement of ICG away from the active wound, as indicated by newly recruited functional lymphatic vessels, emptying of distal lymphatic vessels, or proximal movement of extravascular fluid. They did state that subjects with the longest duration of active ulcers had less movement of the lymph. The study confirms lymphatic dysfunction at an early stage in the aetiology of venous ulcer formation and demonstrates the potential therapeutic mechanism of lymphatic therapy in removing excess fluid and its value in hastening venous ulcer healing.

Lymphoedema therapists bring with them knowledge from their primary profession and many are well equipped to provide adjunct care such as exercise prescription or psychological input. Gone are the days when the doctor tells the patient to rest with their feet up. It has long been known that the calf muscle pump contributes to a healthier venous system in the lower limb. In a meta-analysis conducted by Orr K et al., the researchers concluded that exercise directed at improving calf muscle strength and ankle ROM for individuals with or at risk for venous leg ulcers improves calf muscle pump haemodynamics and function.[8] Nurses and occupational therapists often have skills to deal with mental health which, when

considering the psychological stress caused by leg ulcers, can be very useful.

Lymphoedema therapists have a wide variety of tools in their basket such as MLD, exercise prescription, skin care, elastic taping, cupping, compression and pneumatic compression making the blueprint for each patient's treatment unique and effective. With a good foundation in wound care knowledge and how the lymphatic system interacts with wounds, lymphatic therapy should be a well used tool in the team's toolkit.

In an effort to create awareness and put a practical example on paper, the following complicated case aims to bring the condition to life for the reader.

Case X

Background and history: Mrs X is a 55-year-old female who presented with oedema in both of her legs. She was referred for further management from a massage therapist. She has been treated with MLD using the pathways for a healthy and normal functioning body. At the time, Mrs X was well known to the vascular surgeons and no surgical interventions were necessary or recommended. Mrs X displayed poor lifestyle choices such as smoking heavily, low activity level and poor eating habits. She is self-employed and largely deskbound during working hours. Her company is run from home and she has little need to leave the house. Mrs X does most of her own shopping but may call upon the driver of the business to fetch odds and ends that have been forgotten. She has expressed difficulty with walking from the car to the entrance of the shop and requested a disability sticker. She first noticed her enlarged, but not swollen, legs as a teenager, despite being of a normal weight at the time. This could have been the first sign of lipoedema. She has always had a feeling of tired and heavy legs which is consistent with some venous stasis. Approximately five years ago, her ankles began to swell noticeably and she was no longer able to fit into her shoes. She put this down to her weight and never sought medical advice.

The lipo-lymphoedema coupled with the venous insufficiency symptoms and obesity have meant she has difficulty keeping her fitness up and over the years she has become unable to walk long distances, do the shopping or take part in her grandchildrens' school events. She reports that she cannot stand for more than an hour as her legs begin to ache. She has difficulty walking on uneven ground, hopping and making sharp turns due to the size of her legs and her being overweight. Mrs X is very active with regards to her leisure activities. She is a member of a women's group who focus on crafts, inviting guest speakers for presentations and charity events. She is also a member of a gardening club. She is an active and enthusiastic member of both clubs. She enjoys any type of craft and is an avid knitter.

Clinical examination: Other than her obesity and lipo-lymphoedema, the patient presented with good health, which is often seen with lipoedema (an irregular distribution of abnormal fat cells that may cause pain and bruising, often mistaken for obesity). She has no history of heart, liver or kidney abnormality. She has presented with high blood pressure in the past, however this has been regularly checked and controlled with medication. Her pulses were checked and cleared by her doctor and it was noted that she has signs of chronic venous insufficiency as previously mentioned. An ankle-brachial pulse index was taken and reflected a normal reading. Mrs X presented with fungus under her toenails which she was hesitant to treat and dermatitis on the dorsal aspect of her foot, which was later treated and cleared by the doctor. She presented with a positive Stemmers sign.

Assessment: It was concluded that Mrs X presents with stage 2 lipo-lymphoedema, bordering on stage 3 due to hyperkeratosis beginning to form with co-morbid obesity and chronic venous insufficiency. Her skin has lost elasticity and skin folds were beginning to form thus restricting her movements. She has definite fibrosis on the lateral aspect of her lower leg. No pitting oedema was evident. Her skin was dry and flaky. She displayed signs of haemosiderin staining and reported tired and heavy legs at the end of the day. Mrs X weighed 160 kg with a BMI of 66. She displayed poor lifestyle choices but was motivated to make changes.

Therapy: Therapy began with education around the condition and possible strategies to improve the patient's situation. Lifestyle changes such as weight loss and an exercise programme were discussed. Strategies then focused on merging the lymphoedema management programme, lifestyle changes and her current lifestyle.

An intensive phase was implemented. This aimed at using short stretch bandaging system and MLD for four to six weeks. Treatment began on her left leg only. Once this leg had been controlled and managed, the treatment on the right leg commenced. Bandages were kept on for 23 hours of the day, seven days a week. MLD was performed three times per week, each session lasting between 50–60 minutes. The MLD made use of the Vodder technique, which consists of four light stokes comprised of an active phase and resting phase thus encouraging the movement of the lymph. An anastomosis was created between the inguinal and axillary lymph nodes thus facilitating the drainage of the excess lymph via both nodal systems. (It has been noted that there are updated techniques that may have proven more effective such as fluorescence guided MLD, however the at the time of the treatment, the researcher was not trained in the technique.)

By the end of three weeks the patient's ankle had reduced by 10.5 cm and her calf by 7.5 cm.

Mrs X was satisfied with the girth of her leg and decided to get measured for a compression stocking at this stage. Unfortunately, there was a series of complications such as cellulitis, personal difficulties and work deadlines that delayed the fitting of the compression garment. Therapy was eventually completed and measurements sent to an orthotist for a Class 3, below knee, flat knit garment. It was a custom made garment which, at the time was a long lead time. It took four weeks to arrive. In the interim, bandaging continued daily.

When the stocking arrived Mrs X found it to be too short and difficult to don and doff but persevered as she did find some relief when wearing it. Mrs X was also issued a fabricated neoprene wrap for use during the summer months when she is sitting outside by

the pool. The wrap was not providing the optimal compression but did allow her to engage in leisure activities and increased her exercise throughout summer. It was measured at approximately 20 mmHg (10–20 mmHg less than she needs) using a pressure gauge borrowed from a supplier. It was also noted that she was not bandaging at night which left a large portion of the 24-hour day without support. She made use of the wrap at night which aided in her own self-management.

In addition to the MLD and bandaging, an exercise programme was given to Mrs X. She was to perform the exercises everyday for 20 minutes. Mrs X has had a personal trainer come to her home where she performs the exercises. She sees a hypnotherapist to assist with her reducing the number of cigarettes she smokes a day with little success. She has been encouraged to join a weight loss programme.

Mrs X and a caregiver (her sister) were taught self-MLD, self-bandaging and a skin care regimen.

Discussion: Although it was a relief to finally be able to put a name to her condition, Mrs X found it difficult to accept the amount of effort and the change in her lifestyle that is necessary to control and maintain her lymphoedema. She was very compliant during the intensive phase of treatment but when given the responsibility to don and doff her own garments and bandaging, she began to default. This was largely due to the difficulty with the compression stocking. Once she was given a garment that she could don and doff herself, she was more compliant. Once the intensive phase was over, the patient believed she did not have to bandage any further and it took extensive education to change her mind set back to the advantages of intermittent bandaging. It was suggested that she bandage at least three times per week at night.

Overall, the patient found the therapy to be beneficial to her quality of life. She expressed that she is now able to wear certain pairs of shoes and even a pair of pants for special occasions. The most important improvement for her was her increased ability to transfer from sitting to standing as it empowered her to do more with her grandchildren thus enriching their relationship.

The therapist noted an improvement in mobility and found Mrs X re-integrated into her community. She attended more engagements, complained of less pain and was taking on bigger challenges such as going away for weekends. Her skin integrity improved and was much more supple with less oedema. She was able to fit into orthopaedic shoes which increased her confidence as she was proud of her wardrobe. As her legs improved so did her motivation to exercise and she was attending her sessions with her personal trainer more frequently.

Update since the research: Mrs X managed her symptoms successfully for five years, attending therapy approximately once a month for maintenance. She and her sister were able to manage the home-care programme. She began developing venous ulcers which were difficult to manage owing to the amount of lymph in her tissues as well as the cost of frequent dressing changes. She was regularly checked by vascular surgeons. The lymphorrhoea caused complications at work and she weathered many hospitalisations for cellulitis. Mrs X was referred for negative pressure wound therapy but was not a candidate. The most effective treatment was three layer compression systems with a contact layer of an antimicrobial dressing. These worked well and healed many of the ulcers that formed over the years.

Conclusion

Managing lymphoedema on its own can be difficult but when faced with co-morbid obesity, lipoedema, venous insufficiency and venous ulcers one needs to make sure one is using every tool in the toolbox. It also helps to use every member of the team to get perspective over the treatment options available and draw up an achievable treatment plan.

References

1. https://www.nhs.uk/conditions/lipoedema/
2. Vojácková N, Fialová J, Hercogová J. Dermatologic therapy: management of lymphedema, Vol. 25, -2012, -352-357.
3. Schook CC, Mulliken JB, Fishman SJ, Grant FD, Zurakowski D, Greene AK. Plastic and Reconstructive Journal: Primary Lymphedema: Clinical Features and Management in 138 Pediatric s, Volume 127, No 6 pages 126-128
4. Brorson H. Adipose tissue in lymphedema: The ignorance of adipose tissue in lymphedema. Lymphology 2004;37:175-177.
5. Cheville AL. Current and future trends in lymphedema management: implications for women's health. Phys Med Rehabilitation Clinic N Am 2007: 18: 539-553.
6. Todd M, Lay-Flurrie K, Drake J. Managing ulceration and lymphorrhea in chronic oedema. British Journal of Community Nursing Vol 22 No 5 Chronic Oedema.
7. Eklöf B, Rutherford RB, Bergan JJ, Carpentier PH, Gloviczki P, Kistner RL, et al. Revision of the CEAP classification for chronic venous disorders: Consensus statement. J Vasc Surg 2004;40:1248-52.
8. Orr L, Klement KA, McCrossin L, O'Sullivan Drombolis D, Houghton PE, Spaulding S, Burke S. A systematic review and meta-analysis of exercise intervention for the treatment of calf muscle pump impairment in individuals with chronic venous insufficiency. Ostomy Wound Manage. 2017;63(8):30-43. doi: 10.25270/owm.2017.08.3043.
9. Selvaraj D, Kota A, Premkumar P, Stephen E, Agarwal S. Socio-demography and clinical profile of venous ulcer. Wound Med 2017;19:1-4.
10. Rasmussen JC, Aldrich MB, Tan IC, Darne C, Zhu B, O'Donnell TF Jr., et al. Lymphatic transport in patients with chronic venous insufficiency and venous leg ulcers following sequential pneumatic compression. J Vasc Surg Venous Lymphat Disord 2016;4:9-17.
11. J Vasc Surg Venous Lymphat Disord. 2016 Jan;4(1):9-17. doi: 10.1016/j.jvsv.2015.06.001. Epub 2015 Jul 16.