Single Center Experiences in Chronic Lymphedema: What We Learned Through Clinical Analysis

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Background

Chronic lymphedema is a clinical manifestation of a progressive disease involving the entire soft tissue layer, due to malfunctioning of the lymphatic system. Lymphedema (LE) is the excessive accumulation of fluid and proteins within the interstitial tissue caused by inadequate transport of lymphatic fluid. Simple initial lymphatic fluid retention progresses to chronic inflammatory changes, affecting the entire soft tissue and resulting in fibrosis. Lymphedema, finally, becomes a systemic problem with recurrent local/systemic sepsis that can be life-threatening.

Although LE can develop anywhere in the body where the lymphatic function is impaired, it more commonly is seen in the extremities as a unique vascular problem. The etiology of lymphedema is generally divided into two groups: primary, caused by congenital defect of the lymphatic transport system (referred to as primary lymphedema), and secondary, caused by surgery, radiation therapy, trauma, infection, and/or cancer (referred to as secondary lymphedema).1-3

Objectives

We reviewed the medical records of 1,218 patients registered at the Lymphedema Clinic, SamSung Medical Center in Seoul, Korea, December 1994 through December 2003. A retrospective analysis included sociodemographic data, etiology of their LE, which limbs were affected, the clinical stage of LE at the time of diagnosis, onset, duration from onset of swelling to proper diagnosis, factors that may have triggered the LE, previous treatment received, and subjective symptoms related to the LE.

Etiology & Sociodemographic Data

A total of 1,218 patients consisted of 283 primary lymphedema (23.2%) and 935 secondary lymphedema. Mean age = 32.4±18 years (primary lymphedema) and 53.2±11 years (secondary lymphedema). Sex (M & F) ratio = 1:1.7 (primary) and 1:20 (secondary). Out of a total of 283 primary lymphedema patients, 45 identified as congenital lymphedema, developed/confirmed at birth (15.9%); 170 lymphedema praecox (60.1%), developed through age 35; and 68 lymphedema tarda (24.0%) developed after the age of 35.

Among a total of 935 secondary lymphedema patients, 887 developed LE following lymphadenectomy and/or radiation therapy for cancer management (94.9%). Only 48 developed following a non-cancer-related problem: trauma, infection or liposuction (5.1%) (Figure 1).

Interpretation: The majority of patients in our Clinic presented with secondary lymphedema (76.8%). There were striking differences in age and sex distribution between the primary and secondary lymphedema patients: more younger patients (32.4) and a lower sex ratio (1:1.7) among those with primary lymphedema, and more older (53.2) and female (1:20) patients among those with secondary lymphedema.

Affected Site (Limb)

Primary lymphedema affected 248 lower limbs (87.6%) and 27 upper limbs (9.5%), mostly unilateral without significant difference on the predilection of left or right. Eight patients had multiple involvement to face, upper and/or lower limbs (2.8%).

Secondary lymphedema affected 559 lower limbs (59.8%) and 376 upper limbs (40.2%), mostly following uterine cancer or breast cancer therapy.

FIGURE 1 ~ Types of Lymphedema
TOTAL PATIENTS = 1218
Lymphangitis Development and Management

Four hundred, ninety-nine (41%) of 1,218 LE patients experienced erysipelas/cellulitis, and 300 out of 499 (60.1%) were treated with antibiotics, analgesics, and/or empirical control of symptoms, either from the hospital/clinic or pharmacy, while another 199 (39.9%) received no treatment at all. However, the majority of the 499 patients experienced rapid deterioration of swelling, etc., following an episode of lymphangitis/cellulitis.

Interpretation: Once lymph stasis develops as a consequence of LE, this fluid is the perfect culture media for the bacteria (streptococcus and staphylococcus), and the risk of infection (erysipelas/cellulitis/lymphangitis) increases. Infection can result in thrombosis and obliteration of the lymphatics, further aggravating local edema. Therefore, a suspected episode of erysipelas/cellulitis/lymphangitis in a lymphedematous limb should be medically evaluated and treated aggressively with appropriate antibiotics.

Preliminary Education and/or Information on Possible Lymphedema Development

Among a total of 887 patients who received cancer treatment, only 166 (18.7%) were given the necessary information regarding their risk of developing lymphedema following lymph node dissection and/or radiation. The rest were never properly informed nor educated about this risk, and were not aware that they had LE when their swelling first developed.

Interpretation: Due to poor information/education on the risk of lymphedema development following cancer management, the majority of patients did not understand the cause of their swelling, nor how to treat it properly.

Subjective Symptoms and Quality of Life

The impact of the degree of pre-treatment edema on the subjective symptoms of 952 patients was done using the

Clinical Staging

All 1,218 patients had their LE staged based on the modified International Society of Lymphology (ISL) staging system as one of the parameters for clinical evaluation. 975 individuals had clinical Stage II to III LE (80%), 182 Stage I LE (15%) and 61 Stage IV (5%).

Interpretation: Most patients came to our Clinic after the disease had already progressed to Stage II or III LE (80%), in addition to the end/final stage, Stage IV (5%). Only 15% arrived at the Clinic with only early stage I LE (15%) (Figure 2).

Clinical Onset and Duration Before Diagnosis

Cancer-related secondary LE developed 35.6 ± 57.4 months following cancer treatment, and 658 developed within 3 years (74%) with a range of one month to 30 years post-treatment.

For individuals diagnosed with primary LE, the average delay from actual onset of swelling to proper diagnosis after the initial symptoms developed was 10 ± 11.1 years (1mo.–56 yrs.), and 4.5 ± 5.4 years (1mo.–33.7 yrs.) for secondary LE.

Interpretation: LE can develop any time after cancer treatment—up to 30 years, in our experience—which is compatible with previous reports by Brennen et al.2, and Kocak & Overgaard10. The delay of proper diagnosis was much longer in the case of primary LE (10 yrs.), compared to secondary LE (4.5 yrs.) (p>0.05).

Trigger Factor

Although 796 patients (65.4%) could not recall any specific incident that precipitated the development of their lymphedema, the majority (239 patients-19.6%) recalled performing especially intense/hard work in a short period of time just prior to the onset of swelling, with trauma and/or infection reported by the minority (15%).

Previous Treatment

Despite the presence of limb swelling, 511 (42%) failed to seek any treatment, and only 395 (32.4%) tried simple limb elevation² to reduce early swelling. Almost none of the patients had systematic therapy based on complex decongestive therapy (CDT), which included elastic stockings, low-stretch bandages and/or pneumatic compression therapy. A limited number of patients (360), however, utilized compression stockings or simple bandage application combined with pneumatic compression therapy randomly and/or intermittently. 552 (45.3%) used hot baths, sauna, hot compresses, and/or ice pack application, and 402 (33%) tried acupuncture, vacuum therapy, and/or moxibustion to reduce swelling. The majority of these patients (80%) experienced further deterioration/aggravation of the swelling following these various therapies.

Interpretation: The majority of primary lymphedema cases belonged to the praecox type (60.1%), the majority affecting the lower limb (87.6%). Absolute majority of secondary lymphedema developed in either in upper (40.2%) or lower (59.8%) limb/s, suggests a close relationship with management of breast or uterine cancer, the leading cancers among the Korean female population.

Clinical Staging

TOTAL PATIENTS = 1218
Visual Analog Scale (VAS). We studied the impact of pain, heaviness, decreased sensation, functional disability, and psychological difficulties caused by the cosmetic problem—regardless of upper or lower limb involvement—on quality of life.

We found that the severity of the swelling (p < 0.05) had no significant impact on the subjective reporting of pain, but had a significant impact on decreased sensation, functional disability, and psychological difficulty with body image and accompanying increased difficulty of sociopsychologic adaptability (p < 0.05).

Discussion

Through our limited experience, based on 1,218 lymphedema patients, we learned that most of the patients and their family members were ignorant about the cause of their LE and the importance of risk reduction and treatment due to the ignorance/negligence of their healthcare providers. Therefore, proper understanding of the nature of the disease should be promoted to healthcare personnel, as well as to the patient and family. Basic education in self-care should include: general care of the skin, emphasizing prevention of skin injury; avoidance of abuse/exertion of the affected limb/s; proper recognition of the early symptoms of LE onset, including tight/heavy sensation, numbness, rigid feeling, and ache/pain in the limb at risk; education in the signs and symptoms of cellulitis/lymphangitis, as well as how to respond immediately when an infection is suspected, and what steps they can take to decrease the risk of developing or worsening an existing LE.

The significance of the psychological impact of post-mastectomy lymphedema and its negative impact on quality of life has been previously reported by Carter et al. and Tobin et al. Compared to other post-mastectomy patients who did not develop LE, post-mastectomy patients who developed lymphedema experienced severe difficulties with family relationships and psychological adjustment to their post-mastectomy condition. They were more anxious about disease progression, alterations in body image and self-esteem, and were fearful of the functional disabilities caused by the LE. Therefore, the importance of proper education to reduce the risk of developing these additional psychological impairments caused by the presence of LE following the ablative cancer surgery cannot be overemphasized. Early detection and appropriate management of LE and its complications, combined with proper patient education in LE management, can prevent the progression of the disease. Educating the patient about strategies that can reduce their risk of developing/worsening LE is essential to successfully manage this lifelong condition.

Conclusion

Every healthcare provider should give appropriate attention to reducing the risk of an individual developing chronic lymphedema and should guide their patients in making the necessary modifications in their activities to maintain their normal daily life. The patient and family should be encouraged to continue to be compliant with the daily management of their lymphedema, which is a lifelong condition. Ignorance of the serious nature of LE is the number one enemy, after all.

REFERENCES


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