CASE STUDY:
Managing Genital And Truncal Lymphedema Caused By A Rare Disorder
By Lisa Curtin, PT, CLT-LANA

I. Reason for Presenting Case Study:
This case presentation first introduces Sinus Histocytosis with Massive Lymphadenopathy (SHML), a rare and benign lymph node-based disorder affecting children and young adults. Secondly, it shares the challenges and successes of managing genital and truncal lymphedema in a young man with SHML.

Treatment of genital edema is discussed very briefly in training courses and nearly neglected in reference materials (Textbook of Lymphology, Lymphedema Diagnosis & Treatment). Modification of basic principles and strategies along with creativity and a highly motivated patient resolved an encompassing problem in this unusual case.

II. Lymphedema Diagnosis:
The patient is a 34-year-old male who presented to physical therapy with genital, truncal and right leg lymphedema resulting from SHML. SHML presents in children and young adults as massive painless cervical lymphadenopathy. Many patients also show signs of systemic illness including fever, weight loss and malaise. The diagnosis is made by a biopsy of an enlarged cervical node. SHML can affect any nodal group, but most commonly involves the inguinal, axillary and mediastinal nodes. In this disorder, lymph nodes become greatly enlarged with dilation of the sinuses due to a buildup of lymphocytes. The node capsule can become fibrotic, resulting in the dilation of surrounding lymphatics. Fibrosis also may extend into perinodal soft tissues. Additionally, SHML can affect almost any organ system in the body. Forty percent of documented cases demonstrate SHML with extra nodal involvement. Most commonly involved areas include: bone, skin and soft tissue, nervous system, eye, and upper respiratory tract. Despite documented lymphatic involvement, neither edema nor lymphedema were mentioned as part of the disorder in an extensive literature review. The cause of SHML remains unknown; researchers are investigating immune dysfunction and viral theories.

Prognosis correlates with the number of nodal groups and other systems involved. Most patients with SHML have complete and spontaneous remission. Some have persistent stable lymphadenopathy and a few have died. Medical treatment with corticosteroids has been most effective. Masses may be excised to enhance patient comfort or for cosmetic reasons. Chemotherapy and radiation are used with systemic cases or when extensive lymph node involvement persists.

This patient was diagnosed with SHML at age 14. He had severe involvement of bilateral cervical and right inguinal nodes, and moderate involvement of bilateral cubital, sternal and left inguinal nodes. Extra nodal systems also were involved in this case. He had many skin lesions indicating cutaneous involvement and an orbital mass. The lower respiratory tract was also involved and, at one point, required a tracheotomy to facilitate breathing. His medical treatment included prolonged bed rest, several cycles of antiviral chemotherapy, and daily interferon injections. Additionally, the patient underwent lymph node resections of the right cervical chain, as well as the bilateral cubital and right inguinal nodes.

At age 27, the right inguinal mass enlarged, became ulcerated and resulted in severe cellulitis in the right leg and the diagnosis of secondary lymphedema. The patient had multiple episodes of cellulitis that eventually led him to treatment. He successfully completed a course of Complete Decongestive Therapy (CDT) and began wearing right leg compression stocking. Prophylactic antibiotics were initiated at that time. At age 32, the patient developed fluctuating abdominal edema and had the onset of genital involvement by age 33. He consulted many physicians who prescribed only topical medications. The lymphedema therapists he contacted wouldn’t treat genital edema. Almost a year later, he sought treatment at our clinic to address these issues.

Upon physical evaluation, the patient presented with severe supra-pubic involvement, abdominal trunk edema and mild right leg lymphedema. Inspection revealed a large protrusive papillomatosis (4”x 2” x ¾”) with lymph cysts and fistulas in the supra-pubic area. This area was draining heavily causing severe maceration along the right lateral aspect of the mass. There was considerable congestion and non-pitting edema was also present.
He reported regular supra-pubic infections that required a daily oral antibiotic regimen. He was unable to keep this area clean and dry despite twice daily cleaning with prescriptive products, topical medications and dressing changes. There was no evidence of edema in other genital areas. The patient had a large palpable mass (walnut size) in the right inguinal area with significant fibrosis and scarring. The right leg demonstrated well-controlled lymphedema with minimal fibrosis or skin issues. He also presented with lower abdominal edema, which was symmetrical and spongy in nature. Edema extended to the level of the umbilicus. He reported feeling “congested and full all the time” and was unable to eat without discomfort.

Examination also revealed remaining adenopathy of bilateral cervical and left inguinal node groups consistent with SHML. There was no evidence of edema present in these areas.

III. Medical History:
This patient had no other medical conditions.

IV. Psychosocial Issues:
The patient had recently left a position to start a home computer-based business that allowed him flexibility to manage his symptoms. He lives with his wife who also had a significant medical condition. The patient expressed concerns regarding the impact of his condition on self-esteem and body image, as well as intimacy. Concerns for future medical complications and life changes were unsettling to him. Additionally, patient had little family support locally as he had moved recently.

V. Functional Limitations:
Work performance, activity level, driving, and social activities were considerably impacted due to his inability to sit or stand for more than one hour. The patient required multiple wound and clothing changes daily, and was unable to wear fitted clothing because of fluctuating abdominal edema. Often, he was sidelined by recurrent episodes of cellulitis. In addition, he needed to self-administer four interferon injections each day. The patient’s quality of life was considerably diminished and, as a result, he spent most of his time at home.

VI. Pain:
The patient reported discomfort only in the right groin and abdominal areas during episodes of increased congestion and infections.

VII. Degree of Impairment in ADL (Assisted Daily Living):
The patient was independent in all ADL.

VIII. Pre-Treatment Goals:

IX. Summary of Therapeutic Intervention:
This patient was seen for 17 visits during February and March 2004. Given the severity of his skin condition and high infection rate, CDT was provided daily. Manual Lymph Drainage (MLD) emphasized intensive trunk work and deep abdominal treatments re-directing lymph to the bilateral axillary nodal groups. He was educated in proper skin care and wound management, which was challenging due to the nature of the pelvic area. The patient changed the interferon injection site from the abdomen to the upper arm for lymphatic protection. The home program emphasized positioning for edema control, compression to trunk and pelvic regions, diaphragmatic breathing, exercise for lymph drainage, daily walking and self-MLD.

He wore a right thigh-high compression garment during the day and an alternative garment sleeve at night. Abdominal area was treated with compression cycling shorts and either two 15 cm isobands on the trunk or an abdominal binder (isobands were more comfortable; binder was easily self-applied). The supra-pubic area was treated with a dense foam pad held in place with the compression shorts. Multiple foam pads were trialed and modified to create the most effective shape and design. Within three treatments, most lymphatic fistulas had closed and drainage was minimal. After two more sessions, we noted a 50% reduction in size of the supra-pubic cysts, fistulas and papillomatosis (especially in the central area). At that point, we created a double-layer dense foam pad with aggressive beveling and channels (see diagram) to increase compression, address fibrosis and enhance lymph drainage. The new foam insert also extended inferiorly further along the lateral base of the penis to manage edema and papillomatosis.

During the course of treatment, many different compression products and strategies were tried to address this patient’s supra-pubic edema. In an attempt to address the entire lower quadrant, compression pantyhose were trialed, but failed as it caused significant skin irritation of the massive right inguinal mass. To increase pubic compression, a sacroiliac belt was used. It was ineffective in maintaining compression over the dense foam pad as it slid substantially with position changes. A double truss (hernia product), which was used over the dense foam pad, only effectively addressed the central pubic areas. We experimented with a hip spica style wrap to increase pressure on the lateral pubic aspect. It worked well, but was difficult for the patient to
apply and was not comfortable when sitting. In terms of compression, the combination of compression shorts with additional trunk compression and the double-layer dense foam pad with inferior extensions, and aggressive beveling and channeling gave the best results.

X. Treatment Outcome
At discharge, the patient had a 75% improvement in supra-pubic edema and skin quality. The papillomatosis were completely flattened and continued to resolve throughout the entire area except for areas lateral and inferior to the base of the penis, which remained slightly raised. It was incredibly difficult to apply enough compression there without skin irritation and patient discomfort. The skin breakdown, lymphatic cysts and fistulas had completely resolved. The patient reported a 90% reduction in abdominal symptoms and could now wear normal clothing comfortably throughout the entire day. His sitting and standing tolerances improved dramatically, which allowed him to return to most social and work functions. The patient was independent in his lymphedema self-management that consisted of: daily use of compression shorts with double layer dense foam pad; right thigh-high garment; and use of abdominal compression as needed. The patient performed self-MLD, an exercise program for strengthening and decongestion, and walked regularly. He used a full-legged alternative garment and compression shorts with dense foam pad at night.

All treatment goals were achieved and the patient was extremely satisfied with the outcomes. Additionally, he was infection-free throughout the course of treatment.

XI. Follow-up
The patient was seen three months later for a follow-up visit. He had remained infection-free, although still on an antibiotic regimen. He had complied with all areas of the self-care plan. Trunk and leg reductions were well maintained and the left leg remained free of edema. The supra-pubic area was 98% healed on the right side and 90% on the left side with minimal papillomatosis noted lateral to the penis. No lymphatic cysts or fistulas, skin breakdown, or drainage were present. The patient reported normal functional ability with only occasional discomfort. His psychosocial demeanor improved as well, noted by his new goal of “getting back into shape.” The self-management plan was updated and guidelines to increase activity levels safely were reviewed.

XII. Conclusion
This case study reflects how creativity, a motivated patient, and a consistent plan of care can successfully tackle a difficult problem and dramatically improve quality of life. As a therapist, I needed to work outside my comfort zone, think resourcefully and take on a challenge. By doing so, this experience has been one of the most rewarding of my career. I thank the patient for having the courage to continue to seek medical treatment even as every door was closed to him. I’m glad to have had the opportunity to help such a wonderful person get his life back on track.

XIII. References