Massive Localized Lymphedema, a Disease Unique to the Morbidly Obese: A Case Study

Caroline Fife, MD

Abstract
Massive localized lymphedema (MLL) is a unique presentation of lymphedema resulting in a large, benign, painless mass that develops in morbidly obese patients, most commonly on the medial thigh. Because nearly 6% of the United States adult population is morbidly obese, MLL is believed to be under-diagnosed. To better guide the clinician in identifying and treating MLL, a case study of a 44-year-old Caucasian woman with type I diabetes who presented to the study wound care clinic with MLL is reported, along with the experience of managing more than 70 patients with MLL. A diagnosis of MLL is usually made based on clinical history and presentation. Routine tissue biopsy is not advisable, and diagnostic tests such as magnetic resonance imaging (MRI) may be impossible due to the morbid obesity of most patients. Complete decongestive physiotherapy (CDP) is recommended. Although surgical removal of the MLL collection may be possible, it is technically difficult and not always advisable due to the risk of perioperative complications, including wound dehiscence. Furthermore, in the author’s experience, recurrence is possible even after surgical removal, particularly if conscientious adherence to compression and weight management do not continue. The advent of advanced pneumatic compression devices designed for the morbidly obese and the possibility of using near-infrared fluorescence imaging to guide treatment may transform the MLL management process. Considering the increasing number of MLL cases, the comorbidities and complexities of treating morbidly obese patients, and associated complications, clinicians caring for the morbidly obese need a heightened awareness of this condition.

Keywords: morbid obesity, lymphedema, intermittent pneumatic compression, manual lymphatic drainage, compression bandaging

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Massive localized lymphedema (MLL) is characterized by a benign overgrowth of lymphoproliferative tissue composed of fibrotic and edematous fibroadipose material, resulting in a large, benign, painless mass that develops in morbidly obese patients. MLL is most commonly located on the medial thigh; however, it can also develop on the posterior and anterior thigh, lower leg, suprapubic/mons pubis region, upper arm, popliteal fossa, scrotum, penis, groin, and vulva.

Due to the large size of these lesions, MLL collections often are mistaken for a malignant sarcoma, earning them the name pseudosarcoma. Although the diagnosis can be made clinically, among biopsy specimens subsequently found to represent MLL, 64% were submitted with sarcoma as the favored clinical diagnosis. Thus, it appears, although not uncommon among the morbidly obese, MLL is still commonly mistaken for a malignant tumor.

MLL is generally considered a rare condition. Rare diseases are those affecting fewer than 1:1,500 individuals. However, the prevalence of morbid obesity is on the rise, with 5.7% of Americans 20 years and older considered to be extremely obese (body mass index ≥ 40 kg/m²). Therefore, it seems likely the prevalence of MLL also is increasing and this condition may no longer be considered rare. Furthermore, although the literature pertaining to MLL consists almost entirely of case reports or case series that describe the clinical experience of 65 patients in total, publications are increasing since Farshid and Weiss first discussed MLL in 1998.

These facts make it increasingly important for clinicians...
involved in the treatment of the morbidly obese to be familiar with the presentation of MLL. The purpose of this case study and overview of the author’s clinical experiences is to help clinicians accurately diagnose MLL and institute optimal protocols of care.

**Pathophysiology and Diagnosis**

MLL is a secondary lymphedema generally thought to be caused by disrupted lymphatic flow and drainage due to fat accumulation from obesity. The pathogenesis of MLL is not well understood, but it may be explained as an obstruction of efferent lymphatic flow (such as by an abdominal pannus) or perhaps lymphatic damage due to prior surgery or infection, as in the case to follow. Morbidly obese patients also have excessive adipose tissue deposits that can obstruct lymphatic flow and lead to MLL. Localized ischemia that develops from the weight of and tension on affected tissue and possibly edema also may be associated with MLL; ischemia may trigger a cascade of biochemical events, including the recruitment of growth factors to the affected area, mimicking the wound healing process and leading to fibrosis and accentuated fat lobules. If left untreated, MLL can progress to an aggressive malignancy, a malignant sarcoma, which has been observed in 13% of cases reported in literature. The documented association of MLL with hypothyroidism deserves attention and suggests that all patients with MLL should undergo thyroid function testing.

Histologically, MLL exhibits vascular ectasia, monocellular cell infiltrates, fibrosis, edema between collagen fibers, and ischemic changes, such as infarction and fat necrosis. The diffuse, ill-defined masses of MLL consist of lobules of mature fat interrupted by expanded connective tissue septa. The lymphatics are profoundly dilated and visible to the naked eye if the MLL collections are surgically removed (see Figure 1).

Table 1 presents a summary of the clinical history, physical findings, and treatments related to MLL. It is important to state that MLL is primarily a clinical diagnosis. Although the tissue exhibits characteristic histological features, biopsy is not mandatory or recommended. MLL collections form slowly over several years, a history inconsistent with aggressive malignancies. Thus, careful history-taking is imperative and often confirmatory. If the MLL has no current area of skin breakdown, performing a biopsy can create a permanently nonhealing ulceration that may drain copious amounts of lymphatic fluid. A biopsy may be recommended if raised or pigmented skin lesions are present. Otherwise, it is recommended that diagnosis be made clinically and treatment started as appropriate for MLL. In the author’s experience, if the patient fails to respond to treatment, further diagnostic studies can be performed later.

Clinicians unfamiliar with the condition may feel compelled to obtain advanced diagnostic studies. However, morbidly obese patients have difficulty obtaining magnetic resonance imaging (MRI) or computerized tomography (CT) images due to their size and weight, and these costly studies will serve only to confirm a diagnosis that can be made with a thorough history and physical examination. A faculty member of a major medical school created a diagnostic challenge for clinicians regarding whether his mysterious massive leg “tumor” required surgical excision. The patient, himself a PhD, made national headlines in part because of his struggle to undergo an MRI, because doing so necessitated the loss of 90 kg. His MLL improved dramatically after conservative treatment with bandaging, manual lymphatic drainage, and pneumatic pumping. Thus, currently available imaging techniques for the lymphatics are unlikely to be helpful in the diagnosis and management of MLL. Lymphoscintigraphy only will confirm the delay in lymphatic clearance obvious to the naked eye. The role of MRI lymphatic imaging is limited for the same reasons stated previously and unlikely to provide further direction for therapy.

Newer lymphatic imaging technology is on the horizon. Near infrared (NIR) fluorescence imaging with indocyanine (IC) green has been used to provide real-time, quantifiable images of the lymphatic system. It has shown promise in assessing the effectiveness of manual lymphatic drainage (MLD) therapy, directing MLD therapy by identifying open lymphatic channels and assessing the effectiveness of pneumatic compression device (PCD) therapy. Given its ability to provide information on both the anatomical and functional status of lymphatics, IC green seems the investigational tool most likely to advance the knowledge base pertaining to MLL. Hopefully, studies using IC green can shed light on why these collections form, as well as the optimal method to treat them.

**Treatment of MLL**

The mainstay of lymphedema treatment is complete decongestive physiotherapy (CDP), which consists of MLD, lymphedema bandaging, exercise, and skin care (see Table 1). MLL presents unique challenges for patients with...
lymphedema. Great skill, creativity, and patience are required to incorporate these massive collections into lymphedema bandages. A variety of materials not often used in clinical care may be required, and few therapists have the training and experience needed.

PCDs can be a valuable addition to the regimen both to reduce MLL volume and maintain reductions long-term. However, due to the size of the MLL collections and their often awkward locations (eg, suprapubic or abdomen), standard PCD compression sleeves are unlikely to fit and may have inadequate compression cycles to effect improvement. A novel PCD system has been designed to treat morbidly obese patients (see Figure 2). It applies therapy to the abdominal, genital, and proximal thigh areas as well as to the lower legs utilizing...

Table 1. Summary of massive localized lymphedema: clinical history, lesion description, diagnostic work-up, and treatment

<table>
<thead>
<tr>
<th>History and observations</th>
<th>Lesion</th>
<th>Diagnostic work-up</th>
<th>Treatment</th>
</tr>
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<tbody>
<tr>
<td>Morbid obesity</td>
<td>Characteristic location is medial thigh</td>
<td>None required with appropriate history</td>
<td>CDP(^d) with MLD(^e) and compression bandaging</td>
</tr>
<tr>
<td>More common in women than in men</td>
<td>Also located on posterior and anterior thigh, lower leg, suprapubic/mons pubis region, upper arm, popliteal fossa, scrotum, penis, groin, vulva</td>
<td>MRI(^i) and CT(^j) unlikely to provide therapeutic guidance and often not possible due to patient weight</td>
<td>PCDs(^f) novel devices designed for the morbidly obese are now available</td>
</tr>
<tr>
<td>General lymphedema (eg, “Stemmer’s sign,” non-pitting edema, fibromas)</td>
<td>Ill-defined, non-tender, often with fibrotic skin changes and “cobblestone” appearance</td>
<td>Biopsy not recommended in most patients due to risk of creating a non-healing wound</td>
<td>Surgical removal only if weight is aggressively controlled and patient compliant with compression</td>
</tr>
<tr>
<td>Occasional history of local trauma or cellulitis in the affected area</td>
<td>Can ulcerate</td>
<td>Recommend that diagnosis be made on clinical grounds with invasive studies like biopsy reserved for patients who fail to respond to conservative care</td>
<td></td>
</tr>
<tr>
<td>Possible history of hypothyroidism</td>
<td>Very large in size and weight</td>
<td>Novel technology, such as IC green,(^c) has some promise</td>
<td></td>
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<td></td>
<td>Longstanding duration</td>
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*MRI: magnetic resonance imaging scan; CT: computerized tomography scan; IC green: indocyanine green dye used for medical diagnostics; CDP: complete decongestive physiotherapy; MLD: manual lymphatic drainage; PCDs: pneumatic compression devices

Figure 1. Surgery for removal of massive localized lymphedema; tissue in foreground is being removed and weighs >50 lb.

Figure 2. Novel pneumatic compression device designed for bariatric patients.
ing a compression profile designed for these larger patients. A recent pilot study of this system has shown promising results.19

Most obese patients with lymphedema have great difficulty wearing compression garments. Limb distortion and redundant skin folds typical among the obese present challenges with regard to garment fit and necessitate expensive custom garments. This is particularly true among patients with MLL. Furthermore, most obese patients, particularly those with MLL, have extreme mobility challenges, further limiting their ability to don garments. Semirigid devices, such as the Circaid® (Circaid Medical Products, Inc, San Diego, CA) or FarrowWrap® (FarrowMed, Bryan, TX), can be useful in the morbidly obese. Unfortunately, the Center for Medicare and Medicaid Services (CMS) does not cover compression garments for patients who do not have venous stasis ulcers, so any type of garment or device for lymphedema management is not covered for Medicare beneficiaries. Other payors might cover this important equipment if clinicians write letters of medical necessity. Despite the challenges encountered in obtaining a compression garment for daily wear, all the benefit of treatment will be lost if some method of routine daily compression is not consistently employed.

Surgical removal of MLL is possible and is the preferred approach for some clinicians, although it is technically difficult. In the author’s clinic, patients with chronic draining ulcers will require surgical removal of MLL because wounds, particularly abscess cavities in the extensive fibrotic tissue, are unlikely to heal with even the most aggressive care (see Figure 3). However, MLL collections have been observed to return and even exceed their prior size among patients who failed to achieve significant weight reduction and who failed to utilize daily compression of the limb after surgical removal of MLL (see Figures 4a-c). In the author’s (albeit) limited experience, removal of MLL collections should only be performed after the patient has undergone significant weight loss or bariatric surgery and after the MLL collection has been reduced in size as much as possible using the techniques of MLD, bandaging, and perhaps a PCD. If the MLL collection has had a significant reduction in edema fluid, the skin will soften, thus facilitating surgical closure.

Case Report

Ms. P, a 44-year-old Caucasian woman with type 1 diabetes, presented to the study clinic with MLL, self-referring due to extreme difficulty walking. Her past history was significant for Hashimoto’s disease diagnosed 20 years prior. Her medications at presentation included levothyroxine and insulin. Ms. P had been overweight most of her adult life. However, leg swelling had been ongoing for approximately 10 years following minor trauma to the posterior left calf with subsequent cellulitis and sepsis, complicated by transient renal failure. After the cellulitis resolved, the left leg edema continued to worsen until the leg was profoundly enlarged, after which right leg lymphedema began. Pendulous enlargements at the back of both thighs began slowly and continued to increase over the years until they began to affect her ability to walk. Despite undergoing gastric banding for her morbid obesity 2 years prior and losing more than 45 kg, the bilateral leg enlargement remained unimproved. Ms. P tried to wear compression stockings, but she was not able to find any that fit.

On clinical examination, Ms. P had lymphedema with el-
ephantiasis and MLL on both legs (the left worse than the right) (see Figure 5a,b). She underwent approximately 16 weeks of CDP, including lymphedema bandaging and manual lymphatic drainage provided 2 to 3 times per week in the clinic by trained lymphedema therapists. In addition, she was prescribed a PCD, which she used faithfully at home at least 1 hour per day. Over the course of therapy, leg volumes, as measured by serial girth measurements, decreased more than 2,000 cc on each leg (see Figure 5c). She was provided custom pantyhose-style compression garments (30–40 mm Hg, flat weave) to help with long-term management. Appeals were made to her insurance carrier, requesting authorization to remove the redundant skin and subcutaneous tissue in hopes of further improving her mobility, simplifying the process of garment fitting and decreasing the likelihood of MLL reformation. Despite numerous requests, her insurance carrier refused to authorize surgical removal of MLL tissue. Although lymphedema is not a curable condition, long-term control of leg edema and MLL collections are possible with rigorous self-care once an aggressive treatment regimen has been concluded.

Discussion

In the 15 years since MLL was first brought to the attention of clinicians, its clinical presentations have become more varied in location and seemingly more common. Half of the patients seen by the author have had lesions on anatomical sites previously considered unusual—e.g., the upper arm (see Figure 6), the suprapubic fat pad (see Figure 7), the anterior thigh (see Figure 8), and the pannus (see Figure 9). This current case represents the sixty-sixth of 73 patients seen in the author’s clinic. This is consistent with the most recently published, comprehensive literature review of all previously reported cases of MLL by Chopra et al, who found that nearly half the lesions of MLL were found in unusual locations. His data, supported by this author’s observations, suggest these presentations are no longer uncommon, and it is increasingly important that clinicians are able to accurately diagnose and treat MLL.

Ms. P’s case exhibits many classic features: an adult female, morbidly obese, with comorbidities such as hypothyroidism, a history of trauma to the leg, long-standing leg swelling, and the gradual development of MLL collections over years. Although only 5% of Chopra’s series had lesions on both legs, it is not uncommon among the patients of the study clinic, as the case study exhibits (as well as the patients represented in Figures 1 and 4, although the contralateral leg of the patient in Figure 1 is not shown). Chopra et al previously reviewed MLL13 and noted in a recent study that the number of cases reported increased from 41 to 65. This current article includes additional patients that have not been previously discussed in literature and increases the total MLL cases reported to 73.

MLL can have dire health consequences with the possibility of developing angiosarcoma and an associated mortality rate of 9%. However, for most patients, social and quality-of-life concerns predominate, including restriction in mobility, social isolation, difficulty finding appropriate clothing, and challenges with personal hygiene.

New technologies, such as PCDs designed specifically for morbidly obese patients19 and novel imaging, such as NIR fluorescence, may improve both diagnosis and treatment.16,17 More studies are needed to understand the pathophysiology of this disorder and to optimize treatment protocols. Prevention of MLL will require control of the national epidemic of obesity and associated risk factors for lymphedema.

Conclusion

Based on the clinical experience reported here, MLL affects a substantial number of morbidly obese patients and is likely no longer “rare,” but perhaps “rarely diagnosed.” The author’s experience emphasizes the need for clinicians to be able to identify and treat this condition. MLL can be diagnosed clinically based on its association with morbid obesity and the development of very large, ill-defined lobular masses that enlarge over months to years in characteristic locations like the medial thigh. It can respond surprisingly well to conservative management with CDP and perhaps with the use of PCDs, particularly newer devices specifically designed to treat the morbidly obese. Unfortunately, clinicians skilled in these treatments are difficult to find. Surgical removal is an option but is
only recommended in conjunction with aggressive weight loss and compression, because MLL collections can recur even after surgical removal. New lymphatic imaging technologies may lead to a better understanding of the pathophysiology of MLL, which may result in better treatments.

References