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Lipedema: Painful, Swollen Legs Due To Disordered Fat Deposition



by Robert C. Kiser, DO, MSPH

You have almost certainly seen many patients with lipedema. Lipedema is frequently misdiagnosed as lymphedema, obesity or venous insufficiency (and these can occur as comorbidities). It is a common condition in many areas of the world, including United States, but it is not a common diagnosis in this country.

Lipedema (known in Europe as “lipoedema”) is a clinical entity distinct from lymphedema or interstitial edema. Roughly translated the name means “fat swelling.” It was first described and named by Allen and Hines at the Mayo Clinic in 1940 describing a case series of patients.¹

Definition

The hallmark of lipedema is bilaterally symmetric, abnormal fat deposition and adipose hypertrophy, below the waist leading to progressive leg

enlargement. There are rare clinical variants that affect the arms and related conditions that affect other body sites (such as the shoulders and upper back in Madelung’s disease), but this article will concentrate on the form affecting the lower extremities.

Epidemiology

Lipedema is principally a disorder of women, and on the rare occasions when men are affected it is generally associated with a pro-estrogenic or anti-androgenic state. It is estimated that between six million and 12 million American women suffer from lipedema.* Reports suggest that there is frequently family history of large legs among female relatives.*² Onset is after puberty and most commonly the first symptoms occur between puberty and a woman’s twenties. Cases have been described as occurring or becoming clinically noticed after child-bearing, at menopause, and after gynecological surgery.

Clinical Presentation and Staging

Lipedema is categorized into three stages:

Stage I

In Stage I, the legs have a trunk-like or “stove-pipe” appearance, with fatty deposition over the buttocks, thighs

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Not All Superficial Thrombophlebitis is Benign: The Special Case of Proximal Saphenous Vein Thrombophlebitis

by Robert C. Kiser, DO, MSPH & Alejandro Arnez, MD

Historically, superficial vein thrombophlebitis (SVT) has been often considered a benign and self-limited pathology. However, there are certain types of SVT that warrant heightened attention by physicians; certain areas of thrombosis have increased risk of propagation into the deep system or direct embolism to the pulmonary tree. How to diagnose and when to treat higher-risk SVT appropriately is the subject of this article.

Risk of developing deep vein thrombosis (DVT) or pulmonary embolism (PE) from SVT is related to the location, quality, and duration of the thrombus. In particular, thrombophlebitis located in the proximal (above the knee) portion of the great saphenous vein (GSV) is associated with increased risk of extension to the deep system via the common femoral vein, as well as direct embolism to the pulmonary arteries. Small saphenous vein (SSV) thrombophlebitis that encroaches on the popliteal vein also has been suggested to be of particular concern. (Ascher E, 2003)¹ Appropriate early treatment is therefore important to reduce the risk of potentially fatal PE.

Moreover, it is important to accurately diagnose and document the location of SVT, particularly when it may involve the GSV above the knee. The diagnosis should consider two main sources of information: a clinical exam and Doppler ultrasound. First, the clinical exam that shows a painful, firm, palpable cord surrounded by erythema and edema at or near the anatomical location of the GSV. A Doppler ultrasound scan should then be performed, evaluating the affected vessel and the deep system in order to rule out DVT. For patients with recurrent SVT or a current or past unprovoked DVT or strong family history of the same, the clinician should consider a complete thrombophilia work-up. For recurrent and or migratory SVT one also should have a high index

of suspicion for malignancy and consider appropriate testing.

The treatment for SVT has been quite controversial in the past decade mainly because it was long considered a benign disease. This has changed as ultrasound has become more accessible and studies have shown SVT's association with DVT and PE. In light of these developments, treatment should be tailored according to the location of the thrombus and whether it extends into or threatens the deep system. If the thrombus is located in the GSV below the knee, or the distal half of the SSV, conservative measures may be adequate, including compression stockings and non-steroidal anti-inflammatory (NSAIDs), along with warm compresses, to relieve symptoms. For limited areas of superficial thrombophlebitis secondary to a venipuncture, the American College of Chest Physicians (ACCP) suggests that a topical NSAID (such as Voltaren Gel or Flector patches), may be adequate treatment.³ If the proximal segment is involved, aggressive treatment with low-molecular-weight heparin (LMWH) at prophylactic or intermediate dose for greater than four weeks is recommended.³ The involvement or extension into the deep system is, of course, a DVT and should be treated as such.

Cochrane Review has at least 24 different studies that analyzed the treatment of SVT in more than 2,500 patients, and came to the conclusion that the administration of LMWH plus NSAIDs seems to be the best therapeutic option as it was an effective symptomatic treatment and also reduced recurrence by more than 70% as compared to placebo.⁴ Other alternatives for the treatment of SVT include the administration of Vitamin K antagonist (warfarin) overlapped with 5 days of heparin and continued for greater than four weeks. Surgical thrombectomy and proximal ligation is reserved for intense, localized pain not responsive to medical therapy.

As a conclusion we can suggest that, although throughout history SVT of the GSV has been considered a benign

disease, clinical evidence along with Doppler ultrasound proved the association of SVT with DVT and PE and has renewed interest in this disease. Many studies from different groups have been collected by the Cochrane Review, coming to a conclusion that the use of LMWH for four weeks or greater, with or without NSAIDs, seems to be effective. If the deep system is affected or there is imminent encroachment on the deep system, SVT may be treated as a DVT. Less extensive thrombus in low-risk patients, as well as mid and distal thrombus, should be treated with compression stockings until resolution, as well as NSAIDs and ambulation. Warm compresses may alleviate pain by inducing relaxation of the vein.

All physicians who suspect SVT of the GSV should order Doppler ultrasound imaging, and for patients with unprovoked recurrent venous thromboembolism of any type, they should consider the possibility of a hypercoagulable state such as thrombophilia or malignancy.

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and legs but without enlargement of the feet. At the ankle there is an abrupt “step-off” or cuff of fat. Because the feet are spared, no Stemmer’s sign is seen as would be seen in lymphedema. In layman’s terms the patient may appear to be a dress size 8-10 at the waist, and a 16+ below the waist—although concurrent obesity complicates the assessment. The leg swelling is non-pitting (or minimally pitting late in the evening), and is non-responsive to leg elevation or compression. Compression garments are poorly tolerated because lipedema causes pain, primarily described as anterior and lateral leg (shin) pain upon pressure to the area. Palpation, leg crossing, compression stockings, and even wearing normal pants become painful experiences. Frequent bruising and subcutaneous hematoma is often described. Small, firm nodules occur in the SQ and these are also painful.

Stage II

Stage II includes the above and in addition to the smaller firm nodules, larger rubbery nodules appear. Skin develops a peau d’orange (orange peel) appearance. The typical “jodhpur” (“riding pants”) fat distribution pattern becomes more pronounced during stage II.

Stage III

In Stage III, larger lobule and fat pads (such as in the medial thigh) develop and may interfere with gait. Lymphedema may occur; causing vascular skin changes, pitting edema, skin breakdown. Once lymphedema develops, the Stemmer’s sign will be present, the feet will be affected and the condition is then known as “lipolymphedema.”

Pathophysiology

Increased capillary permeability and friability leads to orthostatic edema, and ecchymosis with or without subcutaneous hematoma. On lymphangiography and lymphoscintigraphy, abnormal, non-linear, coiled lymphatic circuits as seen in the in skin, even in early stages of lipedema (before obvious signs of lymphedema are visible). In addition to the disordered deposition of fat, there

also is fat-cell hyperplasia, with foci of aseptic inflammation and fatty necrosis seen.* The precise mechanism of the associated pain has not been elucidated, but may include neuropathic impingement by fatty units, inflammatory changes, or capillary friability with microhemorrhage. The cause of the peculiar fat distribution also is not clear, but what is clear is that it is distinct from truncal obesity. Unlike the adipose tissue of obesity, the fat of lipedema is not responsive to caloric restriction even to the point of starvation, nor is it responsive to exercise. If the patient with lipedema is obese, dietary restriction will reduce the adiposity of the trunk, head, neck and unaffected limbs, although the lipedemic limbs will remain unchanged or minimally changed. This suggests that the lipedemic fat is atypical at a cellular level in a way that has not yet been fully clarified. One possibility would be estrogen mediated up-regulation of alpha-2 adrenergic receptor activity has been suggested.³

Treatment

Lipedema is generally considered a disheartening and demoralizing condition due to the unresponsiveness of the adipose accumulations to caloric manipulations. Although accumulated lipedemic fat is not responsive to caloric measures, diet and exercise is nevertheless an important part of therapy. For the patient with lipedema, fat will be preferentially stored in the legs during times of caloric excess, and preferentially removed from everywhere else during times of caloric deficit. Therefore, to reduce further accumulation of the fatty deposits of the lower extremities, caloric control and exercise are important preventive measures. Because the bulk and weight of the lobules can interfere with gait mechanics, and because the pain over the adipose accumulations can be an impediment to exercise, water-based aerobic exercise seems to help some patients to maintain ambulatory function.* Lipedema is not obesity, although lipedema does raise the risk of obesity. It is important for physicians to understand that their lipedemic patients need to be educated about the disease,

or they are likely to complain bitterly that their diets are not working to decrease their fat legs. They need to be counseled that caloric restriction or exercise will not diminish the fat on their legs, but that it will help prevent further disfiguring accumulations.

“Complex decongestive therapy,” or CDT, a technique generally performed by occupational and physical therapists who have special training, has been reported to be very helpful for lymphedema, lipolymphedema, and may be helpful in lipedema to reduce pain and orthostatic edema. The therapy includes light lymphatic massage to increase lymphatic flow and decrease lymphatic congestion. Eventually the patient can then tolerate short-stretch bandaging and may later be able to eventually tolerate graduated compression stockings to maintain decongestion.*⁴

Newer Treatments, Future Treatments

Liposuction for lipedema was tried when it was first developed. Unfortunately, the early techniques using sharp cannulas

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Khanh Nguyen, D.O. Appointed Senior Medical Officer

We are pleased to announce that Khanh Nguyen, DO, has been appointed Senior Medical Officer of the Center for Vein Restoration.

Dr. Nguyen, based in Greenbelt, Maryland and Alexandria, Virginia, joined CVR in 2009, and coordinates and oversees the efforts of the various clinical and medical staff ensuring the highest quality in medical care and safety.

"I'm thrilled to be taking on this new role," said Dr. Nguyen. "We're in a very exciting field and I'm proud to help the Center for Vein Restoration maintain, grow and promote the highest standards we can."

In addition to his role at CVR, Dr. Nguyen serves on the executive board of Silver Spring Emergency Physicians and has been a managing partner since 2005. He is an assistant



professor of Emergency Medicine at Georgetown University and is board certified in Emergency Medicine. He continues to be on staff at Holy Cross Hospital in Silver Spring and serves on the Blood Utilization Committee for the hospital.

Dr. Nguyen has a special interest in medical processes and electronic medical records and has been a national speaker and consultant for the McKesson Corporation since 2005. He also is recognized by Cambridge Who's Who in Medicine.

Since joining CVR, Dr. Nguyen has taken on a special interest in improving the quality of patient care and improving patient safety. Dr. Nguyen has undergone extensive training in the treatment of venous insufficiency and has done more than 2,000 ablation procedures. In his spare time he enjoys spending time with his wife and four sons.

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and circumferential techniques damaged lymphatics and resulted in some reports of secondary lymphedema. Liposuction was therefore considered contraindicated for some time. With the newer technique of using blunt cannula and tumescent anesthesia with a linear suctioning technique, liposuction is again being used with good results at three and 10 years.⁵ The technique appears to restore not only the normal contour of the legs, but there also is some suggestion that ecchymosis, hyperalgesia and inflammation are reduced.⁶

Wound care and lymphedema research has benefited lipedema patients as well. In a small pilot study, rapid cycling hyperbaric oxygen pressure has been suggested to improve the pain of lipedema in as little as five days.⁷

Lipedema research may benefit from the ongoing work of researchers to find cellular mechanisms of common obesity. Given the resistance of lipedema adipose tissue to shrink with dieting, there are likely cellular-level differences in the fat that might be exploited to modulate normal adiposity, as well as lipedemic adiposity. This author wonders if these cellular-level differences account for the effectiveness of aminophylline cream on thigh but not truncal fat.*⁸ (same as citation 3)

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Grading of Lipedema

Stage I	Skin surface normal, nodular (small) fatty tissue structure
Stage II	Skin surface uneven (peau d'orange), nodular (big) fatty tissue structure
Stage III	Lobular deformation due to increased fatty tissue

W. Schmeller, I. Meier-Vollrath, *Lipoedema*, in Weissleder, Chuchhardt, *Lymphedema. Diagnosis and Therapy*, 2007

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Associate Editor,
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Hospital
Shekeeb Sufian, MD, FACS

This is an exciting time at the Center for Vein Restoration.

We're serving more patients than ever in an expanding list of communities, including our newest center in Kalamazoo/Portage, Michigan. As always, we're excited to help people learn about vein health and to treat vein disease so our patients can look better, feel better and, ultimately, live better.

A priority for us this year is to educate consumers about vein disease. Many patients think that varicose veins and spider veins are simply normal signs of aging, when in fact they are related to venous insufficiency, a previously overlooked and undiagnosed condition.

You'll see a variety of things from us in the coming months related to this education effort, including our new consumer advertising campaign that shows people, through lifestyle situations, how varicose veins and spider veins affect their daily lives. By working with you, and ultimately clinics like ours, our goal is to show people how they can improve their appearance, wellness and performance, through elective procedures, in ways they might not have expected.

We've also launched our new website, www.centerforvein.com, which we hope better serves not only consumers but also you in the clinic. We've added a new "Physician's Corner" that includes archived editions of this newsletter, bios of our team and more. We've also added a "Doctors Referral" page that makes it easier to refer patients to us.

Thank you again for reading the Venous Review. We hope you find these articles educational and interesting.

Regards,

Robert C. Kiser, DO, MSPH

Editor

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