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Review Article

Lipedema: Therapy Evaluation and Insurance

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Abstract

In this mini-review pathology, diagnosis, signs and symptoms, as well as treatment of lipedema are discussed. As the cause of lipedema is unknown for nearly 80 years, therapy is supportive and aimed at prevention of progression of the disease. Symptoms, signs and phenotypes of these patients are well defined. Guidelines for treatment are clear. When supportive therapy is inadequate surgical tumescent liposuction is the treatment of first choice. Surprisingly, the unproven compression and manual lymphatic drainage therapies of lipedma patients are covered by insurance, while the rational tumescent liposuction is not covered. Quality of life, mobility, lipedema pain, altered gait all improved by tumescent liposuction and disease progression is slowed. Insurance coverage of this procedure will help lipedema patients greatly. Cost-benefit analyses should be made. Research in pharmacotherapy of lipedema that makes sense should be stimulated.

Introduction

Lipedema represents a common medical condition with many misconceptions. It is also known as "adiposis dolorosa" or painful fat and is related to the more extreme tissue disorder, Dercum's disease. Little is understood of lipedema, since the first description in 1940 by Allen and Hines [1,2] The condition occurs almost entirely in females and has an incidence of 11%, or 1 in 9 females. The disease often develops after puberty or other periods of hormonal changes such as pregnancy or menopause. Many people with lipedema are obese or overweight, however, normal weight individuals are also affected. People with lipedema frequently have a family history of relatives with similar enlarged legs. A genetic cause is suspected but not confirmed [3-5].

Far too often, the women who suffer from lipedema are told that their leg growth and swelling is a result of their inability to control their diet or their sedentary lifestyles. As a result of a fixation on diets, exercise and growing body habitus, coupled with "fat shaming", women with lipedema frequently suffer from significant psychosocial distress, including anxiety. depression, eating disorders, and isolation [6]. In this mini-review pathology, symptoms and signs, as well as treatment will be discussed.

Pathology

From a histological perspective, the initial swelling in lipedema is a result of both hypertrophy and hyperplasia [7]. In addition to enlarged adipocytes, there is a thickening of the interstitium with the presence of increased interstitial fluid, secondary to elevated hydrostatic pressure [8-10]. Although interstitial fluid is increased, at least in early stages, the lymphatic system seems to be functioning normally [11]. So,the edema from lipedema at this stage is likely secondary to overwhelming the lymphatic pump, rather than a dysfunction within the lymphatics themselves. However, as lipedema progresses, the lymphatic channels begin to stretch and dilate with the development of many "microaneurysms" that have a tendency to leak. Together with the increased interstitial fluid this results in the development of late-stage lymphedema [12-15].

In addition to adipocyte hypertrophy, interstitial thickening, and lymphatic changes, the subdermal vascular plexus also undergoes conformational changes consistent with a microangiopathy. This microangiopathy results in capillary fragility and leakage, which corresponds to the easy bruising and teleangiectasias seen in lipedema patients [16,17] There also appears to be an influx of inflammatory cells within the interstitium, which may contribute to the signaling mechanisms involved with adipocyte hypertrophy [6].

Diagnosis, Signs and Symptoms

Lipedema generally affects women and often starts during puberty. There are only 2 male cases reported [18,19]. Substantial variability in disease severity and progression is noted. Some women develop minor lipedema, which stabilizes and does not progress over time. Other women show a gradual progression of the lipedema sometimes exacerbated by pregnancy or stressful situations, such as surgeries [20].

The most common complaints are sensations of heaviness and discomfort in the legs, with moderate to severe sensitivity to digital pressure. The swelling and pain worsens during warm weather and exercise and is not alleviated by limb elevation. Weight loss measures generally influences the obesity component but exhibit a minimal effect on the abnormal fat distribution. A typical sign of lipedema is a sharp separation between normal and abnormal tissues at the ankle, the so called cuff sign of lipedema. These patients can wear sneakers and tennis shoes.

The differential diagnosis of lipedema includes obesity, lipohypertrohy, and lymphedema. In cases of more advanced edema chronic venous insufficiency, orthostatic edema and edema due to cardiac failure and internal diseases are included. It is difficult to distinguish among mild forms of lipedema, "normal" variation of fat, and lipohypertrophy. The edema of lipedema is non-pitting, while that in lymphedema is pitting. In addition, lipedema is bilateral, symmetric, while lymphedema is unilateral and sometimes bilateral and asymmetric. The tissue turgor is soft in lipedema and firm in lymphedema. As mentioned before lipedema is tender to palpation and lymphedema is usually non-tender. Infections are rare in lipedema and common in lymphedema A common misconception of patients with lipedema is that they are actually suffering only from lymphedema. Infact, 10%-17% of women being treated for lymphedema have lipedema as well [2]. Patients with lipedema can pinch the skin on the dorsum of their feets and hands without difficulty ("negative Kaposi-Stemmer sign),unlike patients with lymphedema. Diagnostic criteria have been modified by Herbst [6] - (Table 1). Pictures, videos and media are shown at http://www..lipoedema.co.uk/media/ [21].

Table 1

Diagnostic Criteria of Lipedema.
Almost exclusive occurrence in women.
Bilateral and symmetrical manifestation with minimal involvement of the feet.
Minimal pitting edema.
Negative Kaposi-Stemmer sign.
Pain, tenderness on pressure.
Easy bruising.
Persistent enlargement after elevation of the extremities or weight loss.
Arms are affected 30% of the time.
Hypothermia of the skin.
Swelling worsens with orthostasis in summer.
Unaffected by caloric restriction.
Teleangiectasias.

Lipedema consists of 5 major types, with types 1 to 3 being the most common [2,6] - (table 2). Lipedema is more common but can be considered a disease along the spectrum of rare adipose tissue disorders, which includes familial multiple lipomatosis, Madelung's disease and Dercum's disease [6].

Table 2

Type 1	Pelvis, buttocks and hips.
Type 2	Buttocks to knees, with formation of folds of fats around the inner side of the knee.
Type 3	Buttocks to ankles.
Type 4	Arms.
Type 5	Isolated lower legs.

The gold standard for diagnosing and distinguishing lymphedema from lipedema is lymphoscintigraphy., which can demonstrate impaired lymphatic flow in the affected extremity following radiotracer injection. One caveat is that patients with lipedema can have secondary lymphedema [11,15,22]. Other less invasive alternatives to distinguish lymphedema from lipedema are computed tomography (CT) or magnetic resonance imaging (MRI) - [23,24]. MRI is more helpful than CT due to its higher sensitivity. With lymphedema, MRI can show dermal and subcutaneous tissue thickening, as well as a honeycomb appearance to the tissue due to fluid within the subcutaneous fat [25].

Recently, Halk and Damstra issued the first Dutch guidelines on lipedema using the international classification of functioning disability and health [26]. To ensure early detection they advise the use of a minimum data set of repeated measurements of waist circumference, circumference of involved limbs, body mass index (BMI) and scoring at the level of daily practice and psychosocial stress. Promotion of a healthy lifestyle with individually adjusted weight control measures, graded activity program, edema reduction and other supportive measures are pillars of conservative therapy. Tumescent liposuction is the treatment of choice for patients with inadequaete response to conservative and supportive measures, in these guidelines [26].

Treatment

Given that the cause of lipedema remains unknown, lipedema cannot be cured. Treatment is primarily focused on reducing disability and subjective complaints and prevention of progression of the disease.

Conservative Treatment

Although lipedema patients are not generally obese, more than half of patients are overweight [21]. Diets and changes in nutritional behavior cannot reduce the disproportional lipedema tissue that is already present. However, obesity prevention is important because extra weight gain at affected body areas does not respond well to dieting and exercise. Recommended weight control by an individual diet plan and graded exercise are pillars in the conservative management of lipedema [26].

Decongestive lymphatic therapy includes the combination of compression therapy and manual lymphatic drainage therapy and is widely accepted as standard therapy for reducing the edema of lymphedema. Curiously, it has also been generally accepted as standard therapy for lipedema. However, compression therapy and lymph drainage do not reduce fat tissue. Therefore, compression stockings are only advised when there is an edema component present for reducing heaviness and pain [21,27]. Manual lymphatic drainage therapy is not recommended in the treatment of lipedema, in contrast with lymphedema, as the effectiveness of this therapy is not proven in lipedema [21,26,27]. Nevertheless, several home sequential pneumatic compression devices reached the market (e.g. Flexitouch System, Tactile Medical, Minneapolis, Minn., USA or Lymhapress Optimal, Lympha Press USA, Freehold, N. J. Improvements in edema, lymph drainage and capillary fragility have been reported for these devices but the evidence is weak [6].

The same can be said about a wide range of skin care products offered for lipedema at the internet, coconut oil being quite popular at present. Yoga exercise, mindful therapies and aquatic therapy can offer relief for accompanying depression, anxiety and eating disorders, Proper counselling and treatment of these conditions are important [6]. Currently, there is no pharmacotherapy for lipedema that makes sense [6].

Surgical Therapy

From a surgical perspective the least invasive means of removing the painful fat of lipedema is through the use of suction lipectomy. However, it is important to note that the techniques employed for lipectomy of lipedema fat are different from the techniques used in cosmetic liposuction [28-32].The techniques for lipedema liposuction utilize devices that remove fat in a gentle manner, such as the power assisted vibrating canule or water jet- assisted liposuction [29]. Currently, tumescent liposuction is the procedure of first choice [26,28,32]. This technique not only reduces the volume of lipedema fat but also slows progression of the disease [31].

Rapprich *et al.* and Schmeller *et al.* reported reducing lipedema pain, reducing altered gait and loss of mobility, and improving quality of life in these patients after tumescent liposuction [30-32].These results were confirmed by Dadras *et al.* in a longitudinal study of 25 lipedema patients who received 72 tumescent liposuction procedures. The mean follow-up for the first and second postoperative follow-up moment at 16 and 37 months, respectively. Liposuction improved quality of life and symptoms and complaints caused by lipedema, as assessed by standard questionnaires [33].

One of the greatest difficulties with surgical treatment of lipedema is insurance coverage Frustrations on these insurance problems are articulated at best by Sharie Fetzer, the chair of Lipoedema U.K. [34] Mean costs of a tumescent liposuction procedure are € 4429 (range € 1333-€ 8170) depending on the area treated [35]. Sometimes up to three procedures are needed [33]. The procedure is regarded as safe and effective [28,36].

Conclusion

Lipedema is a frustrating and sometimes debilitating disease for the women whom it concerns. The cause is unknown for nearly 80 years. Misconceptions of the public lead to psychosocial problems and social isolation in these patients. The condition is probably underdiagnosed by physicians and confused by lymphedema, lipohypertrophy or obesity. However, diagnostic criteria and phenotypes are well defined and guidelines for its treatment are clear [27]. Conservative treatment of lymphedema including compressive therapy and manual lymphatic drainage is accepted widely as standard therapy for lymphedema. Surprisingly, these therapies are also applied in lipedema patients,. while they don't reduce fat tissue and are unproven in these circumstances. Moreover, these therapies are covered by insurance, while the rational surgical tumescent liposuction therapy is not covered.

Tumescent liposuction is a safe and effective therapy for lipedema patients but rather expensive. Quality of life, mobility, lipedema pain and altered gait all improved in the performed studies and progression of the disease is slowed [31-34]. For that reason tumescent liposuction should be covered by insurance. Cost-benefit analyses must be made for these surgical procedures. In addition, research in pharmacotherapy that makes sense for lipedema patients should be stimulated.

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