

Lipoedema management: Gaps in our knowledge

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Key words

Classification, lipoedema, obesity, oedema.

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Lipoedema does not qualify as a disease in the international disease classification system (Herbst, 2012). However, it is diagnosed in 10% of people attending specialist lymphoedema clinics (Fife et al, 2010; Schmeller and Meier-Vollrath, 2010).

There is no clear understanding of the cause or natural history of this condition, and, in recent years, increased confusion has been caused by wider use of the term “lipoedema”.

Dilemmas in the diagnosis and management of women with lipoedema raised at the 2012 Australasian Lymphology Association Conference in Cairns, Australia, form the basis of this article.

Background

In 1940, Allen and Hines described a condition they called “lipedema” [US spelling]. They recognised a subgroup of patients – all female – attending the vascular leg clinic at the Mayo Clinic, USA, who had a pattern of symmetrical excess subcutaneous adipose tissue in

Abstract

Lipoedema is a condition characterised by abnormal, symmetrical fat deposits in the legs, resulting in a disproportion between a smaller upper body and a larger lower body. Since its first use, the term “lipoedema” devised by Allen and Hines (1940) has been broadened to incorporate patterns of limb adiposity differing from the original pattern, which described involvement of the entirety of the lower limbs except the feet. Obesity co-exists with lipoedema in 50% of women who attend lymphoedema clinics. In the original description of lipoedema, there were anecdotal accounts of dieting not reducing the enlarged lower limbs. The authors report a case of lipoedema plus obesity, which highlights significant reduction in lower limb girth with weight loss. There are gaps in our knowledge regarding lipoedema management due to a lack of assessment for the impact of variable elements of the lipoedema phenotype, such as obesity, venous disease, lymphatic insufficiency, and skin laxity, which each have an impact on patient outcomes. The case study presented in this article shows that, by addressing generalised obesity, leg size in the person with lipoedema can be reduced.

all the regions of the buttock, upper and lower legs, but not the feet. These women had a tendency to develop oedema in their feet as their condition progressed, a condition called “lympho-lipoedema”. It was noted that these women had painful shins and bruised easily. The women who began dieting were found to have lost weight above the waist, but there was no change in the shape of their legs. The authors also noted that the women often had a high level of psychological distress about the shape of their legs.

In 1940, Moncorps et al made further observations that women with lipoedema had reduced elasticity in their skin, and appeared to have joint laxity manifest with flat feet and knee malalignment.

Several subclassifications of lipoedema have been proposed in recent years (Langendoen et al, 2009; Fife et al, 2010; Schmeller and Meier-Vollrath, 2010; Herbst, 2012). These subclassifications have been identified in areas of clinical practice other than in the vascular leg setting, have some overlapping features, but also significant differences to the

traditional classification of lipoedema.

The European Lymphology Society (ELS) accepts a subclassification system with five types of lipoedema (Langendoen et al, 2009):

- Type I – buttock and hips (saddle bag phenomena).
- Type II – buttock to knees.
- Type III – buttock to ankles.
- Type IV – arms and legs affected.
- Type V – lipo-lymphoedema.

The types are determined by the segments of the lower body affected by abnormal adipose deposits, rather than Allen and Hines’ (1940) description of generalised lower-body involvement of the buttock, thighs, and lower legs (but not feet). Allen and Hines’ understanding of lipoedema corresponds to the ELS type III. The ELS subclassification system allows for a mixture of lipoedema types in a given individual.

Some authors describe lipoedema as abnormal fatty deposits in the “extremities” (Rappich et al, 2011), rather than the earlier description of the “lower body”.

Three stages of lipoedema have been described, based on changes in the texture of the limb (Sandby-Møller et al, 2003; Langendoen et al, 2009; Herbst, 2012). In stage 3, out-pouches or large folds of skin deform the shape of the limb. Stage 3 has features in common with the condition called massive localised lymphoedema (MLL; McCrystal and O’Loughlin, 2007), which is generally described as a complication of morbid obesity. MLL usually occurs in the medial aspect of the thigh or in the abdominal pannus. There is usually a single pouch, rather than involvement of the whole limb, but some authors regard MLL with its deforming out-pouch as *ipso facto* stage 3 lipoedema.

Increased confusion in research, diagnosis, and management of lipoedema has been expressed since the clinical criteria defining lipoedema have been broadened.

Types I and II lipoedema overlap with female-pattern gluteofemoral obesity, which has a well-recognised natural history. This type of obesity is diet-resistant, but fat loss does occur during lactation (Bird, 2006).

Research into underlying genetic causes or other aspects of pathogenesis will be more difficult with a more diverse group of people, thus any studies of the natural history or response to intervention for lipoedema will need to stratify their patient populations by lipoedema type.

Causes of lipoedema

Several factors thought to contribute to lipoedema, including hereditary factors, hormones, adipose cell function, and searches for abnormalities in vasculature have been reported on in the literature (Fife and Carter, 2009; Langendoen et al, 2009; Rapprich et al, 2011; Herbst, 2012). Adipose cell research overlaps with work looking at fibroblasts and other elements of connective tissue structure. Relatively weak connective tissue in the subcutaneous and deeper tissues is key in distinguishing lipoedema from lymphoedema.

Clinical presentation

In the clinical setting, it is useful to consider different elements of the lipoedema phenotype, and to assess comorbidities that may be coincidental, causal, or share a common cause.

Lipoedema elements and potential comorbidities include:

- Localised adipose deposits involving buttock, thigh, and lower leg.
- Generalised obesity.
- Oedema.
- Lax skin.
- Pain and hypersensitivity.
- Psychological distress.
- Chronic disorder.

There are many complex interactions between the components listed above. Individual elements may vary in severity from mild to severe.

Localised adipose deposits involving buttock, thigh, and lower leg

This condition is often observed outside the clinical setting, by women who do not regard their condition as a disease, but as an “unlucky heritage”. They usually have a mild form of lipoedema, which remains stable over decades, and is not complicated by comorbidities.

When they present without symptoms such as hypersensitivity and without oedema, some authors use the term “lipohypertrophy” (Langendoen et al, 2009).

Generalised obesity

More than 50% of women attending a lymphoedema services with lipoedema were also found to have generalised obesity (Fife et al, 2010; Schmeller and Meier-Vollrath, 2010), defined in this study by waist measurement (healthy female, ≤ 80 cm; obese female, ≥ 88 cm), and used in addition to BMI (healthy, $20\text{--}25$ kg/m²; obese, >30 kg/m²).

Obesity is now regarded as a chronic relapsing disorder, and it is possible that past impressions about the natural history of lipoedema have been confused with the natural history of comorbid obesity. The interactions between generalised and regional obesity are not well described.

Relationships between generalised obesity, activity level, pain, and psychological distress are complex.

Oedema

Several pathologies other than systemic disease states (i.e. cardiac, liver, renal, and endocrine disorders) potentially contribute to oedema in women with lipoedema. Venous disease is frequently

seen in association with lipoedema (Wold et al, 1951).

It is now recognised that leg oedema can complicate morbid obesity (Fife and Carter, 2009; Dionne, 2011), due to increased fluid exudate from the adipose tissue. Moderate to severe lipoedema is associated with increased interstitial fluid in the legs and progression to lympho-lipoedema.

It has long been known that the anatomy of the lymphatic system is highly variable between individuals. To date, research looking for lymphatic defects in lipoedema has been inconclusive in early stage lipoedema (Langendoen et al, 2009; Fife et al, 2010). Lymphoedema can induce local fatty changes in affected oedematous tissues (Schneider et al, 2005), but it is not known what significance this has in the processes leading to fatty deposits in lipoedema.

Other complex interactions also lead to oedema development. Generalised obesity is associated with increased venous hypertension leading to increased volumes of fluid leaving the capillary bed. Reduced skin elasticity and low activity levels leading to reduced calf pump function can reduce both venous and lymphatic return, further contributing to oedema (Fife and Carter, 2009).

Lax skin, joints, and supportive structures

Lax skin feels soft and loose, and can progress to folds of overhanging skin at the ankle and pouching around the knees. As a person ages, the lower legs develop the classic “tree trunk” or “stove pipe” shape. Supportive tissues within the limb are also affected, which leads to changes in joint alignment (flat feet or malalignment of knees).

Joint laxity can lead to accelerated joint degeneration, (i.e. arthritis), especially involving the knees. Joint pain can limit activity and contribute to a feedback loop of weight gain accelerating knee arthritis, which further reduces activity due to pain. Tortuosity of vessels (venous and lymphatic) in lipoedema has been attributed to lax supporting tissue (Langendoen et al, 2009).

Factors associated with thinner skin in the wider population include sex, obesity, age, location on the body, smoking, and excess corticosteroid use (Sandby-Møller et al, 2003). Skin is thinner in the proximal

limb than the distal limb, and thinner medially than laterally on the limb.

In women with lipoedema, skin bulges are most common medially around the knee, and the stove-pipe shape in the lower leg can be attributed to a failure of the usual support in the gaiter region. These changes increase with aging.

Arterial and venous disease leads to skin atrophy. By contrast, lymphoedema, is associated with thickening of the skin.

Psychological distress

Several of the elements discussed in this article contribute to psychological distress. Self-esteem can be damaged by a perception that body shape defines the worth of the individual. Obese women may suffer from stigma attached to their condition and are vulnerable to social discrimination (in employment and relationships). Most women with obesity have endured repeated failure in the self-management of their weight. This leads to demoralisation and reluctance to re-engage in self-management programmes where their failure will be seen by others.

Low self-esteem, embarrassment about their appearance, and pain may conspire so that women with lipoedema become socially isolated and have low activity levels. For example, when women with lipoedema see a chair, anxiety about their lower body size and whether they will fit in the chair, can be a social barrier (Karasu, 2012).

Lipoedema management

Self-management, along with the continued support of a team of clinicians, is the ultimate goal in chronic disorders. Commitment to management programmes fluctuates depending on other life circumstances of the individual.

Management of the individual with lipoedema requires assessment of all aspect of the index condition (lipoedema) and comorbidities.

Management elements include:

- Accurate diagnosis.
- Psychological assessment and therapy.
- Advice and education on appropriate activity/exercise.
- Advice and education on weight management if obese.
- Modified lymphoedema management plan (with or without compression therapy).
- Consideration of liposuction if waist measurement normal.

- Assessment and management of comorbidities, especially if impacting on pain and mobility.

Management elements

Accurate diagnosis

This gives women with lipoedema a filter through which to access best therapy and assess information, and to view and review her situation.

Psychological assessment and therapy

Implementing sustained lifestyle changes requires energy and some courage. If nothing changes in terms of the lifestyle choices of women with lipoedema, there will be no change in her condition either. Women who have carried the burden of poor self-image, stigma, and pain can benefit from support and counselling and, when they are ready, can engage with a management programme tailored to their needs. Depression needs to be addressed before they can fully self-manage their lipoedema.

Advice and education on appropriate activity and exercise

Both specific “circulation exercises” and increased general activity have clear benefits. Exercise can lift well-being and improve self-esteem, as well as reducing chronic pain and swelling. Specific achievable step-wise programmes create an opportunity for commitment.

The authors’ clinic provides supervised hydrotherapy and supervised gym sessions, tailored to individual needs. Chronic disease management involves therapeutic compromise on occasions when individuals are not as compliant as may be hoped, but compromise may prove better than disengagement.

Advice and education on weight management if obese

There have been no published trials reporting weight management in lipoedema, but it has been repeatedly stated that low-calorie diets will not lead to reduced lower body size (Fife et al, 2010; Herbst, 2012). Generalised obesity is recognised as a common part of the lipoedema picture; as previously mentioned, it is present in at least half of the women attending lymphoedema clinics who are diagnosed with lipoedema. When morbid obesity ($BMI > 40 \text{ kg/m}^3$)

co-exists with lipoedema, lifestyle and psychological factors have contributed to the adiposity.

Weight management is routinely offered to people with lipoedema, and weight stabilisation is a positive outcome.

The authors’ service, with outpatient and inpatient facilities, allows for lipoedema patients to be referred to a multidisciplinary weight management programme. This programme includes access to dietitians and psychologists, as well as supervised exercise programmes.

Modified lymphoedema management plan

Lipoedema without oedema differs from lymphoedema, but a modified lymphoedema management plan can be tailored to the individual with lipoedema. Lymphoedema massage, for example, can reduce the pain of lipoedema in some individuals.

Compression is used for several different reasons in lipoedema. Compliance rates for wearing compression garments in people with lipoedema are higher when compression reduces aching, or when compression reduces oedema. However, compression garments can be uncomfortable in hot climates and the high cost to the patient due to limited financial subsidies for compression garments in Australia are barriers to compression in people with lipoedema.

Moving from negative to positive aspects, compression can lead to reshaping of the limb over time, in particular, reducing ankle cuffing. The degree of change in limb shape is variable.

Aching experienced in those people without oedema, especially when the skin is loose, is often relieved by low-level compression. Similarly, mild venous oedema can be managed with low levels of compression.

Lipoedema patients can benefit from “wrap” forms of compression. These are a fairly rigid bandaging system with Velcro™ (3M) closures. They do not cut into skin creases, and lipoedema patients can adjust the tightness of the bandaging using the straps for optimum pain relief. These are expensive at initial purchase, but last longer than conventional garments. Again, with regards to the use of compression in lipoedema,

Case study

The authors' presented a case study at the Australasian Lymphology Association Conference in 2012, reporting the case of a 27-year-old woman who had lipoedema and generalised obesity. She had been aware of her large legs since the age of 17 and she was diagnosed with lipoedema at 19. Following her first pregnancy aged 26, she saw a persistent increase in leg size and weight, as well as increased pain in her legs. She also experienced easy bruising on her legs and was experiencing distress due to her condition.

At the age of 27, she re-engaged in lipoedema management and a weight management programme. The patient regarded the involvement of a psychologist as a crucial contributor to her wellbeing. With support, she achieved weight loss and reduced the size of her thighs and lower legs, following an 18-month programme of changed eating behaviour, exercise, and importantly, psychological therapy. She did not use any compression as she found the pain to be intolerable. After 18 months, her leg volume was reduced by 14% (Figure 1).

This case challenged previous assumptions made about weight management and showed improvements to quality of life, as well as reduced leg size from weight loss in lipoedema (Table 1).

Table 1. The patient's measurements at baseline and after 19 months' multidisciplinary lipoedema management

	22.09.2010		17.04.2012		Difference	
Weight (kg)	73.8		63.2		-10.6	
Waist (cm)	N/A		80			
Circumference measures (cm)	Right	Left	Right	Left	Right	Left
60	64.6	63.9	61.0	59.2	-3.6	-4.7
50	53	53.9	48.4	49.7	-4.6	-4.2
40	40.3	40.2	38	38.7	-2.3	-1.5
30	46.8	47	42.6	43.0	-4.2	-4.0
20	41	40	36.9	36.8	-4.1	-3.2
10	27.2	26.8	25.6	24.3	-1.6	-2.5
5 cm (ankle)	23.8	24.6	22.8	23.1	-1.0	-1.5
Mid-foot	19.8	19	19	18.7	-0.8	-0.3
Volume (mL)	8747.5	8728.3	7455.9	7503.0	1291.6	1225.3
Total volume (mL)	17475.8		14958.9		-2516.9 (-14.4%)	



Figure 1. Photographs taken after weight loss occurred.

therapeutic compromise may be better than disengagement.

When lymphoedema complicates lipoedema usual protocols for compression garments are recommended (Lymphoedema Framework, 2006).

Liposuction

Liposuction is not currently available in Australia as part of an integrated lipoedema service, but it is available in the community and it has the potential to reduce the disproportion between the upper and lower body. Most women

undergoing liposuction for lipoedema required several (three to five) surgeries over several months.

Langendon et al (2009) state that liposuction is the treatment of choice for patients "with an acceptable waist measurement", and that weight management after surgery is important in ensuring positive patient outcomes.

Rapprich et al (2011) report the results of liposuction on women with type I, II, and III lipoedema, including women with and without generalised obesity. Surgery resulted in an average limb volume

reduction of 6.9%. Some women required continuing, but less intensive, modified lymphoedema self-management after liposuction, including the continued use of compression garments.

Comorbidities

The assessment and treatment of comorbid conditions is essential. For example, knee replacement surgery or management of heart failure will significantly impact on a woman's capacity to engage in a self-management programme for lipoedema.

Discussion

There are gaps in our knowledge regarding many aspects of lipoedema, and there is a need for longitudinal studies of this condition. The interpretation of any report hinges on an accurate diagnosis of lipoedema, especially as the term has been used outside the classic definition.

The case study presented here is evidence that weight loss can lead to lower-body volume reduction in lipoedema and can be sustained. There are also anecdotal reports of leg volume reduction following bariatric surgery for lipoedema associated with generalised obesity (ObesityHelp, 2010; Bauerfeind Life International, 2011). These cases defy the widely held belief that leg volume is not reduced with weight loss in lipoedema.

There has been pessimism about the prospect of weight loss in lipoedema in the literature (Wold et al, 1951; Schmeller and Meier-Vollrath, 2010). When generalised obesity is a comorbidity of lipoedema, it increases the risks and severity of lymphatic decompensation, joint changes, and poor patient morale. Lower-body adiposity is not a benign condition, but is a significant threat to an individual's independence.

Lipoedema is an uncommon condition with multiple variables in the phenotype and there are few data to guide management. Therefore, it is important to build an evidence base on the management and long-term outcomes of lipoedema.

Conclusion

The outcomes of chronic disease management are improved when there is a supportive relationship between the treating clinicians and the person with lipoedema, and when there are realistic expectations based on evidence. Uncertainty and confusion about the nature of treatment reduces compliance.

Unfortunately, there are many unanswered questions about the causes of lipoedema, its natural history, and the impact of interventions on this condition. The authors have outlined a rational approach to understanding and managing excess obesity in lipoedema, based on existing knowledge and experience.

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