

Lipoedema: presentation and management

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Lipoedema is a distinct clinical condition characterized by bilateral, symmetrical enlargement of the buttocks and lower limbs owing to excess deposition of subcutaneous fat (Figure 1) and was first described by Allen and Hines in 1940. Moncorps et al (1940) described a similar condition in females consisting of local fat deposits around the hips, wrists and ankles, erythrocyanosis crurum puellarum, perniosis follicularis and disturbances in the menstrual cycle. This condition was given the term *Typus Rusticanus* because the women had a rosy rural look. Many synonymous terms have been used in the literature to describe lipoedema and include lipodosis, lipohypertrophy, painful fat syndrome and morbid obesity of the legs. It commonly goes unrecognized or misdiagnosed as lymphoedema, chronic venous insufficiency, cellulite or morbid obesity. Tiwari et al (2006) found in their study that only 46.2% of vascular surgeons saw or recognized lipoedema.

Foldi and Foldi (2003) have found that in their experience, the incidence of lipoedema is increased following head injury or resection of pituitary adenoma. Onset is commonly thought to be associated with puberty but is just as likely to develop at any age between puberty and menopause (Allen and Hines, 1940).

Although present in lipoedema, cellulite is quite different (Harwood et al, 1996). Cellulite is found almost exclusively in women and histological investigation has failed to differentiate it from normal adipose tissue (Terranova et al, 2006). The cellulitic appearance is caused by the fibrous bands within the subcutaneous tissue running perpendicular to the surface of the skin. When adipose

tissue is increased in the subcutaneous region the fibrous bands are tethered with the fat lobules protruding beyond the bands giving the lobular appearance. In males the fibrous bands are situated in a more 'crisscross' manner which even in extreme adipose deposition does not cause protruding of the fat lobules (Terranova et al, 2006).

Pathophysiology

Each fat cell in the subcutaneous tissue has its own supply of blood. There is adrenergic nerve supply and there is also thought to be sensory nerve supply. There are no lymph capillaries between the fat cells, instead reticular fibres constitute a prelymphatic transport system.

Histology of lipoedematous tissue has shown much less robust capillaries supplying the fat cells. These fragile vessels also have an increased permeability to plasma proteins. There is little resistance to the movement of fluid into the tissue spaces from the blood vessels owing to the lack of supporting structure of the fat cells (Allen and Hines, 1940). The net result is the development of oedema and easily bruised skin. The oedema expands the prelymphatic structures which severely reduces the flow of fluid towards the initial lymph vessels. Oedema formation is also caused by the absence of the veno-arteriolar reflux. This mechanism causes vasoconstriction under orthostatic conditions in healthy people which prevents oedema. The elasticity of the skin is greatly impaired in lipoedema which causes greater compliance, i.e. there is no external resistance to muscle activity (Jagtman et al, 1984). Normal compliance is essential for the pump action of the calf muscle which has a major influence on venous and lymphatic drainage of the legs.

Tenderness associated with disproportionate pressure is thought to be caused by a fault in the autonomous nervous system. This tenderness is thought to cause neurogenic inflammation which contributes to the microangiopathy.

Results of radiological investigations

Various authors have investigated the morphological and functional changes in the pre-lymphatic structures and the lymphatic vessels in patients with lipoedema. A variety of imaging methods have been used. Fluorescence microlymphography (FM) has shown that multiple microlymphatic aneurysms of lymphatic capillaries are a consistent finding in the affected skin regions of lipoedema patients (Amann-Vesti et al, 2001), and lymphatic

ABSTRACT

Lipoedema is a distinct clinical condition characterized by bilateral, symmetrical enlargement of the buttocks and lower limbs owing to excess deposition of subcutaneous fat. It is found almost exclusively in women. The common features associated with this condition are 'column-shaped' legs with sparing of the feet, bruising, sensitivity to pressure, and orthostatic oedema. The progression to lipo-lymphoedema or morbid obesity is possible. Conservative measures used in the management of lymphoedema can prevent progression/limit the orthostatic oedema. Surgical procedures may also play a part in the management of lipoedema.

KEY WORDS

♦Lipoedema ♦Lymphoedema ♦Compression therapy ♦Liposuction

Figure 1. Typical column shaped legs in lipoedema



microangiopathy with destruction of microvessels develops in lipoedema (Bollinger, 1993).

Abnormal lymphoscintigraphic patterns with a reduction of the lymphatic flow have been demonstrated similar to the changes found in lymphoedema patients (Bilancini et al, 1995). These authors also noted a frequent asymmetry in the lymphoscintigraphic findings that is in contrast to the symmetry of the clinical profile. However, other studies have shown that anatomical and functional status of the epifascial and subfascial lymphatic compartments show no significant alteration in lymphatic transport in patients with lipoedema (Brautigam et al, 1998; Van Geest et al, 2003).

Magnetic resonance imaging (MRI) and ultrasonography studies have shown significantly increased thickness in skin and homogeneously-enlarged subcutaneous tissue in lipoedema (Vaughan, 1990; Duester, Hagspiel, Zuber et al, 1992; Dimakakos, Stefanopoulos, Antoniadis et al, 1997).

Examining both lymphatic and venous function in

10 lipoedema patients, Harwood and colleagues (1996) found minor abnormalities of venous function in only two patients, one had moderately impaired lymphatic function and seven had minimal level of impairment. However, the degree of impairment was not as severe as found in true lymphoedema.

Presentation of lipoedema

Patients presenting with lipoedema are often mistakenly diagnosed as having lymphoedema. Allen and Hines (1940) detailed the characteristics of lipoedema that differentiate it from lymphoedema (Table 1).

Sex

Lipoedema is found almost exclusively in women and an epidemiology study carried out in 2001 demonstrated that it is present in 11% of the female population (Foldi and Foldi, 2003). There are very few reported cases of males being affected. Of 119 patients studied, Wold et al (1949) report only one male with symptoms similar to the women with lipoedema. A more recent case study was reported of a man presenting with 'painful fat syndrome' (Chen et al, 2004). It has been postulated that lipoedema in men is likely to be a result of reduced levels of male sex hormones and/or alterations in liver function (Foldi and Foldi, 2003).

Obesity

Despite being two completely distinct entities, lipoedema and morbid obesity often co-exist. However, the lipoedematous fat deposition in the lower body is not reduced by dieting or exercise and any attempts at reducing weight results in weight loss from the upper body only. Paradoxically, any weight gain tends to affect the lipoedematous regions first (Browse et al, 2003).

Region involved

The excess adipose tissue is demonstrated bilaterally and symmetrically from the ankles to the buttocks giving column-shaped legs and steatopygia. This type of body build, especially the steatopygia, is common in women of the Khoi tribes in Africa or 'Hottentots' (Figure 2) and is considered an aspect of beauty. There is a lack of ankle definition caused by the presence of pads of fatty tissue at the malleoli which stop abruptly at the feet. The feet are always spared which Wold et al (1949) suggest may be owing to the compressive effects of long-term use of footwear. Occasionally the arms are also affected from the shoulder to the wrist with sparing of the hands.

Pain on pressure

Many patients report moderate to severe tenderness disproportionate to the pressure applied. Wold and colleagues (1949) found that 40% of their subjects complained of pain or ache in their lower legs especially when they were on their feet for long periods of time. The majority of patients reviewed by Rudkin and Miller (1994) also reported

Figure 2. Steatopygia is common in the women of the Khoi tribes in Africa



discomfort in the plantar region of the foot resembling neuropathic pain. This sensation was also aggravated by long periods of standing or wearing compression hosiery.

Progression

Lipoedema either develops initially in the hips and progresses to the ankles or vice versa (Foldi and Foldi, 2003). At the outset, the skin appears normal and the lipoedema body shape is relatively mild. As the condition progresses the skin appears similar to cellulite and the distortion in body shape increases. In the later stages the legs and hips can swell to elephantastic proportions (Figure 3). The dependent aspects of these folds

of fatty tissue can develop lymphoedematous skin changes. Often there is also development of obesity of the upper body which causes a wide range of co-morbidities (SIGN, 1996).

History of cellulitis

Cellulitis is very unusual in patients with lipoedema. However, in patients who have gross lipoedematous limbs accompanied by morbid obesity and reduced mobility, the development of lipo-lymphoedema is very likely. In this case cellulitis is not uncommon (Figure 4).

Nature of swelling/oedema

The skin and tissues in lipoedema remain soft and spongy, resembling cellulite. Pitting is absent or minimal if orthostatic oedema is present. Sufferers often experience fluctuating fluid retention related to warm weather, static positioning of the limbs, or the menstrual cycle (Browse et al, 2003).

Effect of elevation

Elevation has no impact on the size of the limbs except where there is minimal orthostatic oedema.

Family history

A positive family history is not uncommon in lipoedema patients. Sixteen percent of the cases in Wold et al's study (1949) reported a family history of similarly large legs. In the nine cases examined by Rudkin and Miller (1994) almost 50% reported a family history. It is therefore possible to postulate that genetic factors are among the causes of lipoedema.

Psychological impact

The psychological morbidity associated with having lipoedema can be immense. In modern society 'size zero' women represent the 'ideal' and there is often huge negative publicity when famous people are seen with flaws, e.g. cellulite or weight gain. An example of this is the article 'Time to ditch the shorts, Mischa! Miss Barton sports some unflattering spring wear as she relaxes with her dogs in the park' published on the *Daily Mail* website (Littlejohn, 2010) in March of this year.

Having lipoedema can represent the opposite of the popular female magazines' generated 'norm'. The emotional impact of having lipoedema can range from mild upset to noticeable anxiety and depression. Wold et al (1949) found that 29% of their lipoedema patients had developed neurosis. Some patients have said that their lives have been ruined because of the shape of their legs, and some have admitted to being 'mirror peepers' – constantly looking in mirrors with the hope of seeing that their legs are not really as bad as they think. Sufferers will go to great lengths to hide their legs either in the type of clothing worn, by avoiding activities that would expose their legs such as swimming, or standing behind furniture. Eating disorders are common in lipoedema; either anorexia to try and lose weight or overeating as a source of comfort (Foldi and Foldi, 2003). A variety of terms have been used either

Table 1. Differential diagnosis		
Characteristic	Lipoedema	Lymphoedema
Sex	Women only	Both sexes
Obesity	Present	Present or absent
Region involved	Always both limbs	Usually one limb
Pain on pressure	Usually present	Usually absent
Progression	All parts of limb are involved simultaneously	From distal to proximal portion of limb
History of cellulitis	Absent	Occasionally present
Nature of swelling	Usually soft	Usually firm
Pitting oedema	Usually minimal	Usually marked
Effect of elevation	Persistent enlargement	Reduction to normal size in early stages
Family history	Frequently obtained	Almost always not obtained



Figure 3. Lipo-lymphoedema and morbid obesity

by patients or to patients, to describe the shape of their lower bodies and include mushroom hips, muffin hips or pumpkin hips. These names can cause great distress.

Management of lipoedema

The treatment of lipoedema is generally deemed unsatisfactory (Allen and Hines, 1940). However, there are some differences of opinion regarding management of lipoedema in the literature. Because of their skill and knowledge in the management of lymphoedema, lymphoedema practitioners are the most appropriate professionals to assess and manage patients with lipoedema.

It is possible to prevent the orthostatic oedema with the use of compression hosiery but Allen and Hines (1940) found that the benefit was greatly outweighed by the discomfort of wearing the garments. Foldi and Foldi (2003) recommend avoiding compression in the first week of decongestive lymphatic therapy (DLT) and use manual lymphatic drainage (MLD) initially until the discomfort is reduced. DLT has been shown to reduce the capillary fragility in lipoedema and thus reduce the haematoma formation (Szolnoky et al, 2008b). Compression therapy is believed to be useful in preventing the progression of lipoedema to lipo-lymphoedema, and if lipo-lymphoedema has developed then compression is essential. DLT has been shown to achieve significant volume reductions in leg oedema in lipoedema patients (Szolnoky, et al, 2008a).

Beninson and Edelglass (1984) report that most patients significantly reduce the abnormal fat deposition by wearing

properly measured and fitted Jobst garments combined with dietary weight management. However, other authors have asserted that adipose tissue cannot be compressed (Foldi and Foldi, 2003) and therefore compression will not reduce the fatty component of lipoedema.

If conservative management of lipoedema is unsuccessful some authors believe surgical intervention may be an option (Allen and Hines, 1940; Warren et al, 2007). Liposuction is the most common procedure used. Tumescant liposuction involves injecting large volumes (6–10 litres) of fluid (sodium chloride, adrenalin and local anaesthetic agents) into the suprafascial space to provide a firm consistency of the area to be aspirated (Stutz and Krahl, 2008). This method is thought to prevent severe tissue trauma (Sattler, et al, 1999). The procedure is delayed by approximately 30–90 minutes to allow infiltration of the fluid into the adipose cells. The outcomes of tumescant liposuction on 28 patients with lipoedema were reviewed (Schmeller and Meier-Vollrath, 2006). All had successful normalization of body proportions with complete disappearance or striking improvement in pain, sensitivity to pressure and bruising. Complications consisted of temporary mild post op swelling only.

Rudkin and Miller (1994) reviewed seven patients who had undergone surgery for lipoedema. Three underwent skin and subcutaneous excision which significantly improved the size and shape of the limbs. One patient however developed persistent post-operative foot swelling with exacerbation of plantar discomfort. Liposuction in conjunction with limited excision of skin and subcutaneous tissue was performed on four patients. Again, size and shape of limbs were improved but no post-operative foot swelling developed. Seroma formation was the only complication reported by Chen et al (2004) in the liposuction of their patient.

Water-jet assisted liposuction (WAL) was carried out on 30 patients with lipoedema (Stutz and Krahl, 2008). This procedure uses a fan-shaped water jet directed at the subcutaneous space to separate the adipose cells from the tissue. At the same time the injected water plus the detached cells is aspirated via a specified vacuum pressure.

Figure 4. Cellulitis



The cannula is directed only along the axis of the lymph collectors. The volume of fluid used (1–1.5 litres) is less than the tumescent method and because there is no need for an infiltration time, the procedure time is reduced. Details of outcomes or post-op complications were not reported in this study but through analysis of the aspirate the authors determined that to a large extent, damage to the lymph vessels could be avoided using this method.

Foldi and Idiazabel (2000) reviewed the clinical outcomes of 263 patients who had lymphoedema, lipo-lymphoedema or lipoedema and had undergone surgery for the management of varicose veins. In each group the results were poor, i.e. swelling was increased or unchanged in more than 90%, and symptoms such as heaviness, fatigue and cramps were improved in less than 10%. Surgery for varicose veins in these groups of patients should only be carried out if there is an absolute surgical need, e.g. ascending phlebitis or bleeding (Pereira et al, 2005). The most appropriate form of treatment for patients with lipoedema is conservative measures only and liposuction is always contraindicated (Foldi and Foldi, 2003).

Weight management

It is generally acknowledged that weight reduction measures will not reduce the fatty deposit caused by lipoedema. However, it is prudent to stress the importance of avoiding any unnecessary weight gain to prevent the development of lipo-lymphoedema and upper body obesity. This in turn will prevent obesity-associated health issues. It may be difficult to motivate some patients to follow a healthy eating lifestyle when psychologically they are affected by the shape of their bodies. Support and sensitivity is important when discussing these issues.

Psychological support

Psychological support should never be underestimated for lipoedema patients. Macdonald et al (2003) commented that emotional support and reassurance that lipoedema is not the patient's fault, is the most important service provided by practitioners. A sensitive and patient-led approach to presenting the patient with the news that there may need to be adjustments to lifestyle and acceptance of the condition may encourage the patient to do so (Allen and Hines, 1940).

Conclusion

Lipoedema is a recognized clinical condition that is distinct from lymphoedema, obesity and cellulite. The physical and psychological sequelae require a sensitive and

knowledgeable approach to assessment and management. Lymphoedema practitioners have the skills and knowledge to provide lipoedema patients with a holistic plan of management.

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KEY POINTS

- Lipoedema is a distressing condition affecting approximately 11% of women.
- Features include excessive fat distribution in the legs, pain and orthostatic oedema.
- Lymphoedema practitioners can provide appropriate diagnosis, management and psychological support for this patient group.

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