

The challenges of lipoedema complicated by secondary lymphoedema

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Abstract

In this article, Anne Williams and Isobel MacEwan explore the pathophysiology of lipoedema, the changes to the lymphatic system in lipoedema, and the challenges for individuals who live with lipoedema complicated by secondary lymphoedema and associated problems such as cellulitis.

Introduction

Lipoedema is type of fat disorder that mainly affects women, and is associated with symmetrical proliferation of diet-resistant inflammatory fat tissue, most commonly in the legs, buttocks and arms (Figures 1 and 2) (Forner-Cordero et al 2012; Wounds UK 2017). In some individuals, a secondary lymphoedema can also develop when the lymphatic system becomes overwhelmed and is unable to effectively drain the interstitial fluid (Figure 3). This leads to additional problems such as skin and tissue changes, and a higher risk of cellulitis. This paper will overview current knowledge relating to lipoedema pathophysiology and the possible changes that occur when secondary lymphoedema develops in someone with lipoedema. It will outline the key steps that can be taken to avoid progressive complications such as cellulitis, including skin care, and the use of compression therapy, signposting the reader to additional information on cellulitis management.

Background to lipoedema pathophysiology and presentation

The cause of lipoedema is not clear (Williams and MacEwan 2016; Wounds UK 2017), but may be associated with endocrine, connective tissue, auto-immune and inherited factors (Langendoen et al 2009; Wounds UK 2017). The condition is often mistaken for obesity, and while someone with lipoedema may also have a co-existing 'obesity-related' problem, lipoedema fat is distinctly different, and does not respond to traditional weight loss strategies.

The various pathophysiological changes that may occur in lipoedema are outlined in Table 1. In the lower limbs, these lead to a characteristic leg shape in the early stages of lipoedema including an 'ankle bracelet' without foot swelling (Figure 1), and/or enlarged buttocks, without a similar increase in the waist and upper torso. Lipoedema also occurs in the arms but usually does not affect the hands. People with lipoedema may experience pain, particularly when pressure is applied to the area, and a tendency

to bruising. Enlargement of the limb, pain and bruising, can all make it challenging to obtain an accurate blood pressure reading, or take a blood specimen on the limb affected by lipoedema (Wounds UK 2017). When palpated, the tissues can often feel cool, due to the insulating effect of fat, and the skin is often distinctly soft and pliable with small lipomas sometimes felt beneath the skin.

Table 1: Possible pathophysiological features of lipoedema (adapted from Wounds UK 2017)

- Increased numbers (hyperplasia) of fat cells and/or increased size of fat cells (hypertrophy) in specific areas
- Infiltration of fat tissue by inflammatory products
- Growth of fragile blood capillaries in fat tissue
- Reduced elasticity of skin and connective tissue.

If lipoedema progresses, later stages can be associated with fat lobes or bulges, for example, adjacent to the knee (Figure 3) which can alter the gait, and may exacerbate a hip or knee joint problem. Significantly, changes also occur in the lymphatic drainage system, predisposing people with lipoedema to develop secondary lymphoedema (Wounds UK 2017).

Lymphatic system changes in lipoedema

The lymphatic system is a one-way drainage system, removing interstitial fluid via a network of tiny lymphatic capillaries which drain into the contractile pre-collector and collector lymphatics (Figure 4). The collector lymphatics filter lymph through lymph nodes and back into the blood circulation. The lymphatic system therefore plays an active role in maintaining fluid balance, providing host defence and immune surveillance; the intestinal lymphatics (or lacteals) are also the main route for fat absorption in the body (Mortimer and Rockson 2014).

Evidence is limited but indicates that while the lymphatics may be normal in early stages of lipoedema, several changes may occur to predispose someone with lipoedema to develop lymphoedema. These include the development of lymphatic microaneurysms (Amann-Vesti et al 2001), increased interstitial fluid formation due to

blood capillary fragility, and inadequate lymph drainage in enlarged fat tissues (Wounds UK 2017). As the lymphatic system is now understood to be the main route for drainage of capillary filtrate (Mortimer and Rockson 2014), it follows that any changes to lymph drainage capacity may result in lymphoedema when the lymphatic system fails to keep up with microvascular filtration rates. It may also be surmised that, as the lymphatic system depends on tissue pressures to stimulate movement within the lymphatic vessels, the soft and pliable nature of skin and connective tissues in lipoedema may reduce the contractile efforts of these vessels. More work is required to better understand these mechanisms. However, from a clinical perspective, the accumulation of interstitial fluid and subsequent development of lymphoedema is a relatively common but often misunderstood complication of lipoedema.

Recognising secondary lymphoedema associated with lipoedema

This is sometimes referred to in the literature as lipo-lymphoedema, and may be suspected when a patient presents with one or more of the following:

- Signs of fluid accumulation including indentations in the skin, feelings of heaviness, or an obvious pitting oedema, when finger pressure is gently applied to the tissues for up to 30 seconds.
- Foot swelling and a positive Stemmer's sign. The feet are not normally swollen in lipoedema, but may begin to show signs of swelling, and eventually a positive Stemmer's sign, with an inability to pinch up the skin at the base of the second toe, due to tissue fibrosis (Lymphoedema Framework 2006).
- Changes in the tissues which become harder (due to fibrosis) and warm (usually due to inflammatory processes). This is distinctly different to the soft and cooler nature of 'pure' lipoedema. Tissue fibrosis may become obvious in the fat lobes, or in other dependent areas, and the skin may look like 'orange-peel', or develop small wart-like bulges called papillomas. For some people these change may also be complicated by ulceration and lymphorrhoea (leakage of lymph), hyperkeratosis or lipodermatosclerosis, resulting in extreme changes to leg shape such as narrowing in the gaiter area.
- Risk of cellulitis in the swollen areas (Figures 5a, 5b, 5c) may be associated with underlying primary lymphatic insufficiency (Damstra et al. 2008), lymph

stasis, and generalised inflammatory processes that readily develop into an acute cellulitis or erysipelas (Figures 5a, 5b and 5c illustrate cellulitis in individuals with lipoedema and secondary lymphoedema).

It should also be acknowledged that people with lipoedema may experience oedema related to other conditions such as heart failure, obesity, or surgery such as joint replacement. A comprehensive assessment of the various factors contributing to oedema formation is therefore required, including a review of medications that also influence and exacerbate oedema formation (Mortimer and Rockson 2014). Importantly, diuretics are unlikely to be useful in this group, unless indicated for other conditions such as heart failure.

Cellulitis

There is limited literature related to cellulitis in people with lipoedema. However evidence from peer and online support services provided by organisations such as Talk Lipoedema, indicates that cellulitis in lipoedema may be more common than expected. In a small survey of 50 women with lipoedema, 17 (34%) reported having had cellulitis (Williams and MacEwan 2016). There are anecdotal reports of extreme reactions to insect bites leading to cellulitis in someone with lipoedema, even in early stages of lipoedema (Figure 6); and fatal sepsis resulting from cellulitis in women with lipoedema.

Cellulitis, sometimes referred to as erysipelas, is a non-contagious acute spreading infection of the skin and subcutaneous tissues with a sudden onset, characterised by redness, heat, swelling, pain, tenderness and, usually, fever with a raised body temperature (British Lymphology Society (BLS) and Lymphoedema Support Network (LSN) 2016). It may develop as a result of an obvious skin break such as a cut, bite, or areas of skin inflammation such as tinea pedis (athlete's foot) or dermatitis, although a specific cause is not always obvious. Recurrent cellulitis can be a debilitating problem, often due to inadequate first-line management. However, it is also imperative that cellulitis is correctly diagnosed and differentiated from conditions such as eczema, dermatitis, deep vein thrombosis and acute lipodermatosclerosis (Levell et al 2011), so antibiotic therapy is not used inappropriately.

When treatment with antibiotics is timely and the response to treatment can be monitored, most patients with cellulitis will be managed at home with oral antibiotic

therapy. This may include amoxicillin 500mg 8-hourly or, if allergic to this, erythromycin 500 mg 6-hourly or clarithromycin 500 mg 12-hourly (BLS and LSN 2016). Patients with signs of sepsis, or continuing or deteriorating systemic signs after 48 hours of oral antibiotic treatment, require admission to hospital for intravenous antibiotic therapy. Blood tests such as C-reactive protein (CRP), which is an indicator of inflammation or infection, and white cell count are recommended where appropriate (BLS and LSN 2016). In one study of 635 patients with lower limb cellulitis referred to a cellulitis clinic run by an experienced dermatology nurse, 18 patients required admission for IV antibiotics (Levell et al 2011). Further work is required to better understand the context and extent of cellulitis in people with lipoedema complicated by lymphoedema, as it is likely that cellulitis itself may further compromise the health and function of the lymphatic system.

Preventing complications of secondary lymphoedema related to lipoedema

Early identification of lipoedema and recognition of when oedema may be developing is key to the prevention of complications such as cellulitis and deterioration in skin and tissue condition. Recognising when someone has lipoedema and related problems with lymphoedema provides opportunity to initiate support with self-management and/or refer to appropriate professionals such as a lymphoedema, tissue viability, vascular or dermatology services.

Self-management approaches aim to enhance the health of the lymphatics and adjacent tissues, reducing swelling and accumulation of fluid, leading to improved physical and mental wellbeing. It is important to note that women with lipoedema may experience considerable psychological distress and poor quality of life (Dudek et al. 2016; Wounds UK 2017). This may be associated with years of misdiagnosis, limited response to extreme dieting, the social stigma of increased body size, and the debilitating physical features of lipoedema such as pain and poor mobility (Wounds UK 2017). This distress will influence motivation towards self-care and may compromise the relationship with health professionals. Therefore a sensitive and empathic approach to self-management support is required.

Self-management support

Skin care: regular washing, care with drying the skin, and use of emollients will help to ensure the skin remains intact and inflammatory processes are minimised (Wounds

UK 2017). The person should be encouraged to identify and respond to changes in their skin and tissues, seeking advice where appropriate.

Preventative care: this can include a range of approaches which aim to prevent deterioration in the skin, tissues, underlying microcirculation and lymphatics (Williams 2010). For example, use of insect repellent to avoid stings, or immediate use of an antiseptic on a skin break. Patients may wish to avoid venepuncture or blood pressure recording in a swollen area, and alternatives may be negotiated, for example, recording of blood pressure on the lower arm. However, further research is required to better understand the risk of cellulitis associated with these activities.

Use of compression therapy: compression therapy aims to reduce capillary filtration and enhance lymphatic and venous function, to improve limb shape, skin integrity, and quality of life (Wounds UK 2015). In a previous paper we outlined the compression therapy choices for people with lipoedema (Williams and MacEwan 2016) and further detail is now available (Wounds UK 2017). When secondary lymphoedema has developed as a result of lipoedema, decongestive treatment including bandaging may be required to reduce the fluid component, before a compression stocking is fitted. The use of adjustable compression wraps now provides additional scope for self-management. Correct measurement and fitting of compression garments is imperative, and referral to appropriate services should be considered, with compression donning and doffing aids made available where necessary.

Healthy eating and weight management: a survey showed that 98% of women with lipoedema had tried to lose weight (Fetzer and Fetzer 2016). Although there is limited research evidence available regarding the most effective dietary approach for people with lipoedema (Wounds UK 2017), support and advice is available through various organisations such as Talk Lipoedema and Lipoedema UK.

Physical activity: where physical activity is important in managing weight, and improving mental wellbeing, it will also enhance lymph drainage. Activities can include swimming, seated exercises, Nordic walking, or specific activities such as the Tripudio Movement System, which specifically aims to improve lymph drainage and physical function. It should be acknowledged that people with lipoedema may lack confidence to take part in these activities, although there is evidence that group approaches can be motivating and effective in people with lymphoedema (McGowan et al. 2013).

Other approaches to improve symptoms: there are a variety of other approaches that may be used as part of self-management and by health care professionals to enhance wellbeing, manage symptoms such as pain, and reduce oedema. These include manual lymphatic drainage, self-lymphatic drainage, gentle skin brushing, kinesio-taping, and deep oscillation therapy (Wounds UK 2017). Further research is required to improve understanding of the influence and effectiveness of these approaches on outcomes for women with lipoedema.

Conclusion

Lipoedema is a poorly understood condition that can be complicated by secondary lymphoedema. This can result in many challenges for people with lipoedema and health professionals, particularly related to poor skin integrity, abnormal limb shape that influences mobility, and cellulitis. A partnership approach, providing support with self-management, and referral to appropriate services, is key to reducing the risk of progressive and challenging complications of lipoedema, and improving quality of life. A newly available Best Practice Document for the Management of Lipoedema (Wounds UK 2017) is a key resource for those with lipoedema, health professionals and service development teams.

For further information please see: <http://www.talklipoedema.org/>

References

- Amann-Vesti BR, Franzeck UK, Bollinger A (2001) Microlymphatic aneurysms in patients with lipedema. *Lymphology* 34, 4, 170-75.
- British Lymphology Society (BLS) and Lymphoedema Support Network (LSN) (2016) Consensus document on the management of cellulitis in lymphoedema. Available at: <http://www.thebls.com/the-bls/professional-and-patients/cellulitis-inlymphoedema/>.
- Damstra, R.J., van Steensel, M.A., Boomsma, J.H., Nelemans, P., Veraart, J C. (2008) Erysipelas as a sign of subclinical primary lymphoedema: a prospective quantitative scintigraphic study of 40 patients with unilateral erysipelas of the leg. *British Journal of Dermatology*, 158, 6, 1210-5.
- Dudek JE, Bialaszek W, Ostaszewski P (2016) Quality of life in women with lipoedema: a contextual behavioural approach. *Qual Life Res* 25, 401-8.
- Fetzer A, Fetzer S (2016) Lipoedema UK Big Survey 2014 Research Report. Available at: <http://www.lipoedema.co.uk/lipoedema-uks-big-survey/>

Forner-Cordero I, Szolnoky G, Forner-Cordero, A, et al. (2012) Lipedema: an overview of its clinical manifestations, diagnosis and treatment of the disproportional fatty deposition syndrome – systematic review. *Clinical Obesity*, 2, pp.86-95.

Langendoen SI, Habbema L, Nijsten TEC, & Neuman HAM. (2009) Lipoedema: from clinical presentation to therapy. A review of the literature. *British Journal of Dermatology*, 161, pp.980-6.

Levell, N.J., Windfield, C.G., Garioch, J.J. (2011) Severe lower limb cellulitis is best diagnosed by dermatologists and managed with shared care between primary and secondary care. *British Journal of Dermatology*, 164, 1326-8

Lymphoedema Framework (2006) Best Practice for the Management of Lymphoedema, International consensus. London: MEP Ltd.

McGowan, A., Williams, A., Davidson, F. & Williams, J. (2013) A self-management group programme for people with lymphoedema: experiences from a Third Sector Project. *British Journal of Community Nursing, Chronic Oedema Supplement*, pp. S6-12.

Mortimer PS, Rockson SG (2014) New developments in clinical aspects of lymphatic disease. *Journal of Clinical Investigations* 124, 3, 1-7.

Williams A. (2010) Self-care in lymphoedema. *Journal of Community Nursing*, 24 (4) 28, 30, 32-4.

Williams, A., MacEwan, I. (2016) Accurate diagnosis and self-care support for women with lipoedema. *Practice Nursing*, 27, 7, 325-332.

Wounds UK Best Practice Statement (2015) Compression hosiery (2nd edn). London: Wounds UK. Available to download from: www.wounds-uk.com.

Wounds UK (2017) Best Practice Guidelines: The Management of Lipoedema. London: Wounds UK, 2017. Available to download from: www.wounds-uk.com.



Figure 1: Early stage lipoedema



Figure 2: Lipoedema of the left arm



Figure3: Lipoedema complicated by secondary lymphoedema



Figure 4: The lymphatic system



Figure 5a: Cellulitis and blistering of the leg in someone with lipoedema and secondary lymphoedema



Figure 5b: Cellulitis of the right leg in someone with lipoedema and secondary lymphoedema



Figure 5c: Cellulitis of the left leg in someone with lipoedema and secondary lymphoedema