138: Lymphoedema

Lymphoedema commonly presents as chronic swelling due to compromised lymph drainage (Figures 138.1 and 138.2). It has a wide range of causes, and remains a distressing long term problem for many individuals recovering from cancer treatment (Cormier et al., 2010; Shaitelman et al., 2015). In the UK, lymphoedema prevalence has been identified as 3.99 per 1,000 population, rising to 1% of those over 65 years (Moffatt & Pinnington, 2012).

This chapter provides an overview of the lymphatic system, causes, diagnosis and psychosocial impacts of lymphoedema, and describes treatment and self-management approaches for people living with lymphoedema.

The lymphatic system

The lymphatic system is a one-way drainage system working alongside the vascular system to remove excess fluid and cell debris from the interstitial tissues throughout the body (Figure 138.3). Lymph drains via a series of initial, pre-collector, collector lymphatics, and lymph node groups, and into the blood circulatory system near the heart (Mortimer & Levick, 2004). The lymphatic system also has a key role in managing infection, removing inflammatory waste products, and transporting proteins and fats.

Development of lymphoedema

Lymphoedema swelling develops when there is an imbalance between capillary filtration and the transport capacity of the lymphatic system, due to various primary or secondary causes. A staging system for lymphoedema (Table 138.1) illustrates the progressive
nature of the condition, associated with inflammation, tissue fibrosis, skin problems and cellulitis risk (International Society of Lymphology (ISL), 2013).

### Table 138.1: Stages of lymphoedema (ISL 2013) with possible signs and symptoms

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<th>Stage</th>
<th>Pathophysiology</th>
<th>Signs and symptoms</th>
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| Stage 0 | Impaired lymph transport without obvious clinical signs                          | • Subjective changes such as heaviness, warmth, intermittent swelling of one or more area (limb, breast, torso, head, genitals)  
|         |                                                                                 | • Cellulitis with no obvious cause                                                 |
| Stage I | Early accumulation of protein-rich fluid leading to inflammatory processes      | • Swelling reduces with elevation/at night                                           
|         |                                                                                 | • Tissues ‘pit’ with gentle pressure; clothing, jewellery or footwear may mark the skin |
| Stage II| Lymph stasis and inflammation lead to fibrosis in later stages; excess fat may start to accumulate | • Persistent swelling; tissues feel thicker and harder (fibrosis) and do not easily ‘pit’ with pressure; positive Stemmer’s sign (inability to pinch up the skin of the second toe; fragile skin may leak (lymphorrhoea)  
|         |                                                                                 | • The skin may become uneven due to pressure in the lymphatics; skin folds develop |
| Stage III| Inflammation and continued fibrosis lead to trophic skin changes.             | • Persistent swelling and fibrosis                                                 
|         |                                                                                 | • Hyperkeratosis (scaliness), papillomatosis (cobblestone appearance) and cellulitis are common. |

### Causes of lymphoedema

*Primary lymphoedema*
Primary lymphoedema is due to congenital and functional changes in the lymphatic system, including absent, dilated or reduced numbers of lymphatics, or lymph node fibrosis. Classifications of primary lymphoedema are increasingly based on genetic phenotyping (Connell et al., 2013). Milroy disease presents at birth or early childhood as bilateral leg swelling, and is associated with VEGFR3 mutations. Syndromic lymphoedemas are also associated with conditions such as Klippel-Trenaunay or Turner syndrome.

**Secondary lymphoedema**
This acquired lymphoedema and has many cancer and non-cancer-related causes:

**Cancer-related lymphoedema**
Lymphoedema affects around 20 per cent of women after breast cancer treatment, associated with axillary lymph node clearance, radiotherapy, and positive lymph nodes (DiSipio et al., 2013; Tsai et al., 2009). Other risk factors include: high body weight at diagnosis; taxane-based chemotherapy; and post-surgery events such as taking blood from the affected arm (Kilbreath et al., 2016). Sentinel node biopsy reduces the incidence of lymphoedema to around 6% (DiSipio et al., 2013).

Lymphoedema is also associated with gynaecological and genitourinary cancers (Finnane et al., 2011; Noble-Jones et al., 2014; Yost et al., 2014), and head and neck cancer (McGarvey et al., 2013). Incidence of lymphoedema in invasive melanoma is reported as 31 per cent after axillary node dissection, and 40% after inguinal node dissection (Cromwell et al., 2015). Advanced, metastatic cancer also leads to lymphoedema due to metastatic lymphadenopathy, immobility, venous thrombosis and hypoalbuminaemia (International Lymphoedema Framework, 2010).

**Non-cancer-related secondary lymphoedema**
Lymphoedema occurs secondary to: trauma; venous disease and ulceration; obesity; lipoedema; immobility in neurological conditions (Lymphoedema Framework 2006). The normal transport capacity of the lymphatic system is overwhelmed by excess capillary filtrate (ISL, 2013), leading to progressive lymphoedema.
Lymphoedema diagnosis

Diagnosing lymphoedema can be challenging. Vascular and lymphatic system pathologies are not easily differentiated without specific diagnostic expertise and effective management of co-morbid conditions. Individuals should be referred to appropriate lymphoedema services, and may require specialist care from lymphology, vascular or dermatology centres.

Recognising lymphoedema

Clinically, lymphoedema is usually characterised by swelling, increased limb volume, changes in limb shape, and deterioration of skin and tissues, with a risk of cellulitis (Table 1), and associated changes in physical function or gait. However, lymphoedema may also affect the torso, breast, head or genital area, sometimes without limb swelling. Swelling may be minimal in early stages or intermittent, becoming evident after certain activities or in hot weather.

Psychosocial effects of lymphoedema

Literature reviews have identified psychological and social impacts of lymphoedema, including poor self-identity, poor body image, emotional disturbance, psychological distress, financial burden, social isolation, public insensitivity, poor work support, and perceived diminished sexuality (Fu et al. 2013; Upton and Solowiek, 2011). A variety of lymphoedema quality of life tools have been utilised and validated in the literature (Cromwell et al. 2015; Devoogdt et al. 2011; Franks et al. 2006; Keeley et al. 2010; Klernäs et al. 2015). Despite this, evidence is mainly limited to observational and qualitative studies, often with inconsistent reporting of symptoms and quality of life outcomes following treatment (Leung et al. 2015). One study of 189 individuals with lower limb lymphoedema utilised the MOS SF-36 questionnaire at baseline and at 24 weeks, reporting poor health in relation to role physical and role emotional, with improved scores correlating with reduced limb volume after treatment (Franks et al. 2006).

Studies of people with cancer-related lymphoedema provide insight into specific risk groups. For example, an online survey of 166 women with lymphoedema after breast
cancer (Alcorso et al. 2015) reported a relationship between body image disturbance and depression and anxiety, particularly in older women. A survey of 277 individuals with lymphoedema following surgery for malignant melanoma, reported lower (poorer) FACT-M scores in those with lower extremity lymphoedema compared to those with upper limb swelling (Cromwell et al., 2015). Other consistent themes associated with lymphoedema include: feelings of uncertainty, guilt and shame, loss of control, and disappointment with treatment outcome (Karlsson et al., 2015; Williams et al. 2004; Williams 2011). Specialist psychological care may be indicated for some individuals with lymphoedema.

Management of lymphoedema

Lymphoedema management usually involves two phases, based on a partnership approach between the person and professional:

- Decongestive treatment (Complex Decongestive Treatment or intensive treatment) with regular (sometime daily) sessions over a 2-3 week period (Lymphoedema Framework 2006)
- Self-management by the person to maintain and optimise the condition (Ridner et al., 2012).

Decongestive treatment

Undertaken in those with later stage lymphoedema, poorly shaped limb, torso oedema, or skin problems such as papillomatosis or ulceration. Treatment may include manual lymphatic drainage massage (Ezzo et al., 2015) combined with compression therapy such as multi-layer bandaging, skin care and exercise, to reduce swelling, reshape and decongest the limb, and improve symptoms.

Self-management

Meticulous skin and preventative care is essential to ensure the skin remains intact, reduce the risk of skin damage, and minimise the risk of infection. Many individuals will be fitted with a lymphoedema compression garment to reduce accumulation of tissue fluid. Wearing this daily draws attention to the condition, and can be an additional source of anxiety (Williams 2011). Exercise and movement are used to enhance lymph
drainage; exercise prescription must be individualised, and may include: chair exercises; swimming; group exercise; or progressive resistance training at a gym where appropriate. Additional approaches include self-massage, compression pumps, kinesio-taping, vibration and deep oscillation machines, although research evidence is limited.

Poor motivation, inadequate information, psychosocial and functional difficulties will influence successful self-management (Armer et al., 2013; Fu, 2010; Williams 2011). Opportunities for individual and group self-management support (McGowan et al., 2013), regular treatment and effective psychological care from professional, NHS services, and third sector community resources are essential but often inequitable. Stress management approaches and psychological support including Cognitive Behavioural Therapy and mindfulness may be a key part of self-management, although these are not commonly discussed in the lymphoedema literature.

Other treatment approaches

Surgery such as liposuction and microsurgical reconstruction is suitable for a small number of people with lymphoedema (Cormier et al., 2012). Diuretics are not used for lymphoedema, unless indicated for concomitant health problems such as cardiac disease.

Conclusion

Lymphoedema is a long term condition with many causes and various possible psychosocial sequelae. Effective self-management and physical treatment of the condition are essential to reduce the risk of progressive symptoms and complications such as cellulitis. Ongoing psychosocial support enables people with lymphoedema to live well with lymphoedema, and specialist psychological care may be indicated for some individuals.

References


Figures

Figure 138.1: Diagram of the lymphatic system
Figure 138.2: A woman with breast cancer-related lymphoedema

Figure 150.3: A man with bilateral leg swelling