Lymphoedema - a chronic disease, not a side effect

Summary: Lymphoedema (soft tissue swelling resulting from obstruction of the lymphatic drainage system) is a chronic illness that has immense physical and psychological impact on a patient’s life.

Relevance: Management of this illness, while conservative, can be life changing when approached by a co-ordinated multidisciplinary team. (1,2) In the UK, specialist lymphoedema services are often run from hospices.

Take-home messages: Although some patients with lymphoedema will require minimal support, it is vital that patients and healthcare professionals are vigilant for rare but serious complications.
LYMPHEDEMA - a chronic disease, not a side effect
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AETIOLOGY

Primary lymphoedema is usually occurs as a result of congenital physiological abnormalities, one example being Milroy’s disease. Secondary lymphoedema may be iatrogenic (following surgical removal of lymph nodes as part of cancer treatment, most common in developed countries) or post infectious (a key cause in developing countries, particularly filariasis in South East Asia). (3,4) Previously known as elephantitis, filariasis is a parasitic infection of the lymphatic vessels by nematodes (roundworms) belonging to the Filarioidea family. (5) Even though this illness poses a huge global challenge, there are very few lymphoedema specialists outside of Europe. (6)

DIAGNOSIS

Lymphoedema is usually diagnosed clinically. Patients complain of uncomfortable swelling and skin atrophy in an extremity (including the scrotum, head, and neck) following infection or surgery. (3) Common pitfalls in diagnosis lie in the differential diagnoses such as cellulitis and peripheral vascular disease. In cases of uncertainty, it is important to note that specialist diagnostic tests are available, including tissue tonometry (non-invasive) and lymphoscintigraphy or MR lymphangiography (invasive). (3)

MANAGEMENT

Most lymphoedema is treated conservatively, guided by symptom burden, and led by a lymphoedema specialist nurse. Specialist lymphoedema services, (1) along with psychology and physiotherapy input have proven to have better patient outcomes. (7) In one intensive specialist intervention programme over half of their participants achieved a 50% reduction in lymphoedema. (2) Patient education plays a vital role. Patients benefit from lymphoedema specialists ensuring they understand the condition, including important and serious risks. (8)

Non-surgically, the aim of therapy is to reduce the volume of lymph fluid in the tissue, thereby reducing pain, weight, and the likelihood of local infection. (4) This can be achieved by a decongestive lymphatic therapy (DLT) which is a combination of manual lymph drainage and compression bandaging. DLT in combination with adjuvant skin care and light exercise can take months of intensive treatment to take effect. Newer techniques such as laser and pneumatic decompression are also available. (4)

Pharmacologically, diuretics are not recommended outside of co-morbid renal/cardiac failure. (9) In general antibiotics should only be used during episodes of cellulitis or for long term prophylaxis in recurrent cellulitis e.g. more than twice in 12 months. (10) In the context of post-infective lymphoedema, the World Health Organisation recommends annual preventative chemotherapy in endemic areas to end the cycle of nematode infection. (5) Liposuction, tissue excision and lymphatic bypass are all potential surgical options. (11) However, at present NICE has only deemed liposuction as adequately safe and effective. (12)

COMPLICATIONS

In the chronic care of patients with lymphoedema, reducing risk of complications is a huge component. The theoretical risk of infection should be avoided. Experts advise diligent skincare and avoiding invasive procedures (venepuncture, cannulation) on a lymphoedematous limb where possible (10, 13, 14). Sudden worsening of lymphoedema should be actively investigated. While a concomitant oedema (for example, cardiogenic) may be a cause, venous thromboembolism, cellulitis, or recurrent carcinoma could also be responsible (3, 14). A rare and fatal condition which can occur in chronic lymphoedema is lymphangiosarcoma, a soft tissue neoplasia, known as Stewart Treves syndrome, which requires specialist input from dermatology and Lymphoedema services. (15) Stewart Treves syndrome has a poor prognosis and should be considered in patients with long term lymphoedema who present with skin changes such as nodules or non-healing ulcers. (15)

WHAT YOU NEED TO KNOW

1. Tissue swelling could be lymphoedema in any patient. Consider congenital and infective causes if there is no history of node removal, as globally, lymphoedema is not always a cancer-related diagnosis.

2. Involvement of specialist lymphoedema services has significantly better outcomes and should be an early priority in suspected cases.

3. Patients with lymphoedema are at high risk of local infection. Bear this in mind when carrying out any invasive procedure, right down to venepuncture.

4. In a patient with pre-existing lymphoedema, sudden worsening should be investigated fully: consider venous thromboembolism, cellulitis, recurrent carcinoma, or oedema of another cause. A rare but fatal complication is lymphangiosarcoma.
REFERENCES


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