Lymphatic system

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INTRODUCTION

The lymphatic system was first described by Erasistratus in Alexandria more than 2000 years ago. William Hunter, in the late eighteenth century, was the first to describe the function of the lymphatic system.

Anatomy and physiology of the lymphatic system

Functions

1. Removes water, electrolytes, low-molecular-weight moieties (polypeptides, cytokines, and growth factors) and macromolecules (fibrinogen, albumen, globulins, coagulation and fibrinolytic factors) from the interstitial space and returns them to the circulation.
2. Permits the circulation of lymphocytes and other immune cells.
3. Intestinal lymph (chyle) transports cholesterol, long-chain fatty acids, triglycerides and the fat-soluble vitamins (A, D, E and K) directly to the circulation, bypassing the liver.

Development and macroanatomy

Lymph from the lower limbs and abdomen drain via the cisterna chyli, lying between the aorta and azygos vein, into the thoracic duct. This duct is a major lymph channel which passes cephalad on the left of the bodies of the thoracic vertebrae to enter the left side of the neck, where it drains into the left internal jugular vein at its confluence with the left subclavian vein.

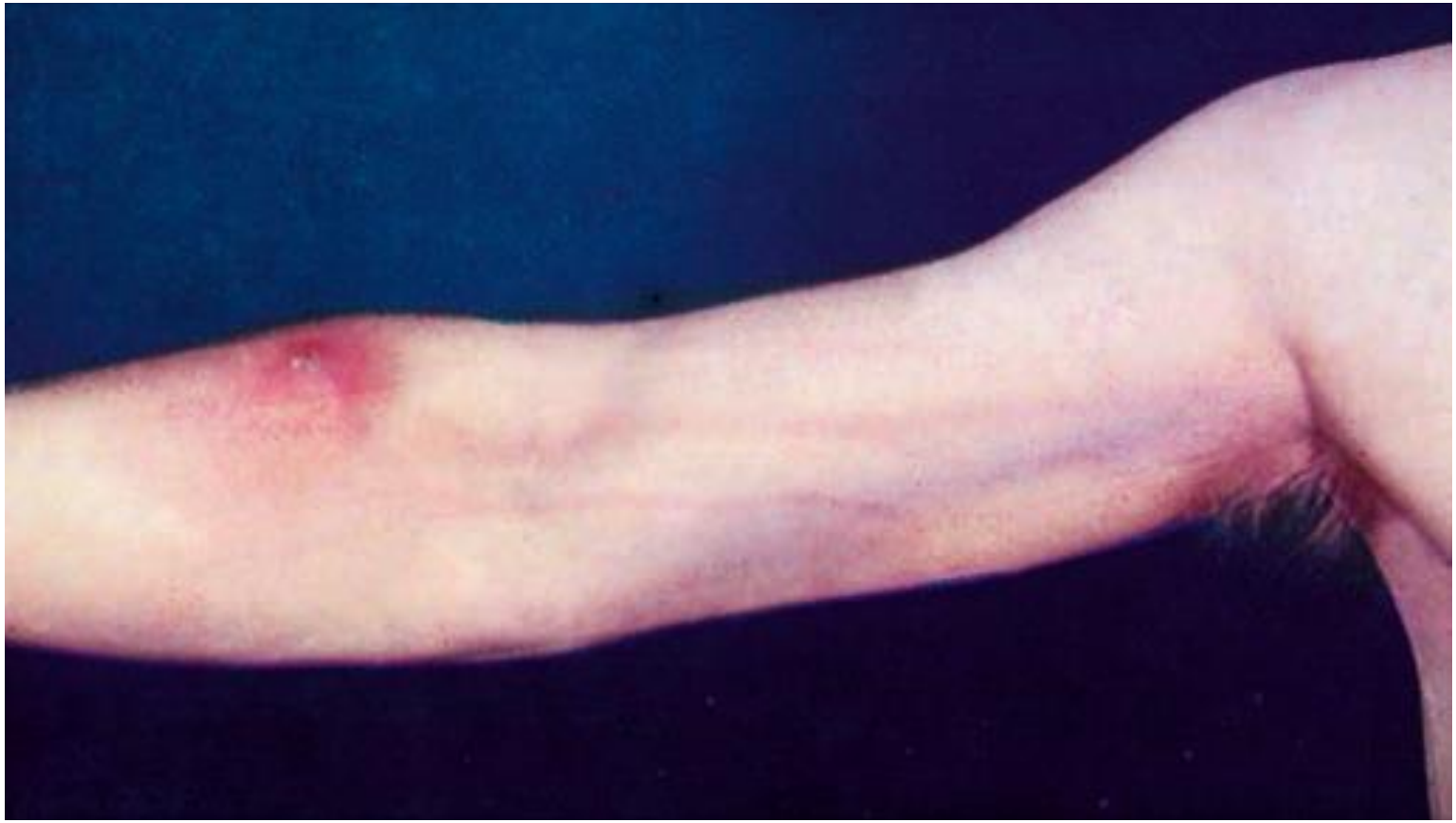
Lymph from the head and right arm drains via a separate lymphatic trunk, the right lymphatic duct, into the right internal jugular vein. Lymph nodes develop as condensations along the course of these lymphatic highways.

Acute inflammation of the lymphatics

Acute lymphangitis occurs when a deep or superficial infection, often due to Streptococcus pyogenes or Staphylococcus aureus, spreads to the draining lymphatics and lymph nodes (lymphadenitis) where an abscess may form. Eventually this may progress to bacteraemia or septicaemia. *The normal signs of infection are present and a red streak is seen in the skin along the line of the inflamed lymphatic.*

Treatment

The inflamed part should be rested, elevated to reduce swelling and the patient treated with intravenous antibiotics. Failure to improve within 48 hours suggests inappropriate antibiotic therapy, the presence of pus either in the lymph nodes or at the site of primary infection. The lymphatic damage caused by acute lymphangitis may lead to recurrent attacks of infection and lymphoedema.



Acute lymphangitis of the arm

Lymphoedema

Lymphoedema may be defined as abnormal limb swelling caused by the accumulation of increased amounts of high protein ISF secondary to defective lymphatic drainage in the presence of (near) normal net capillary filtration.

Pathophysiology

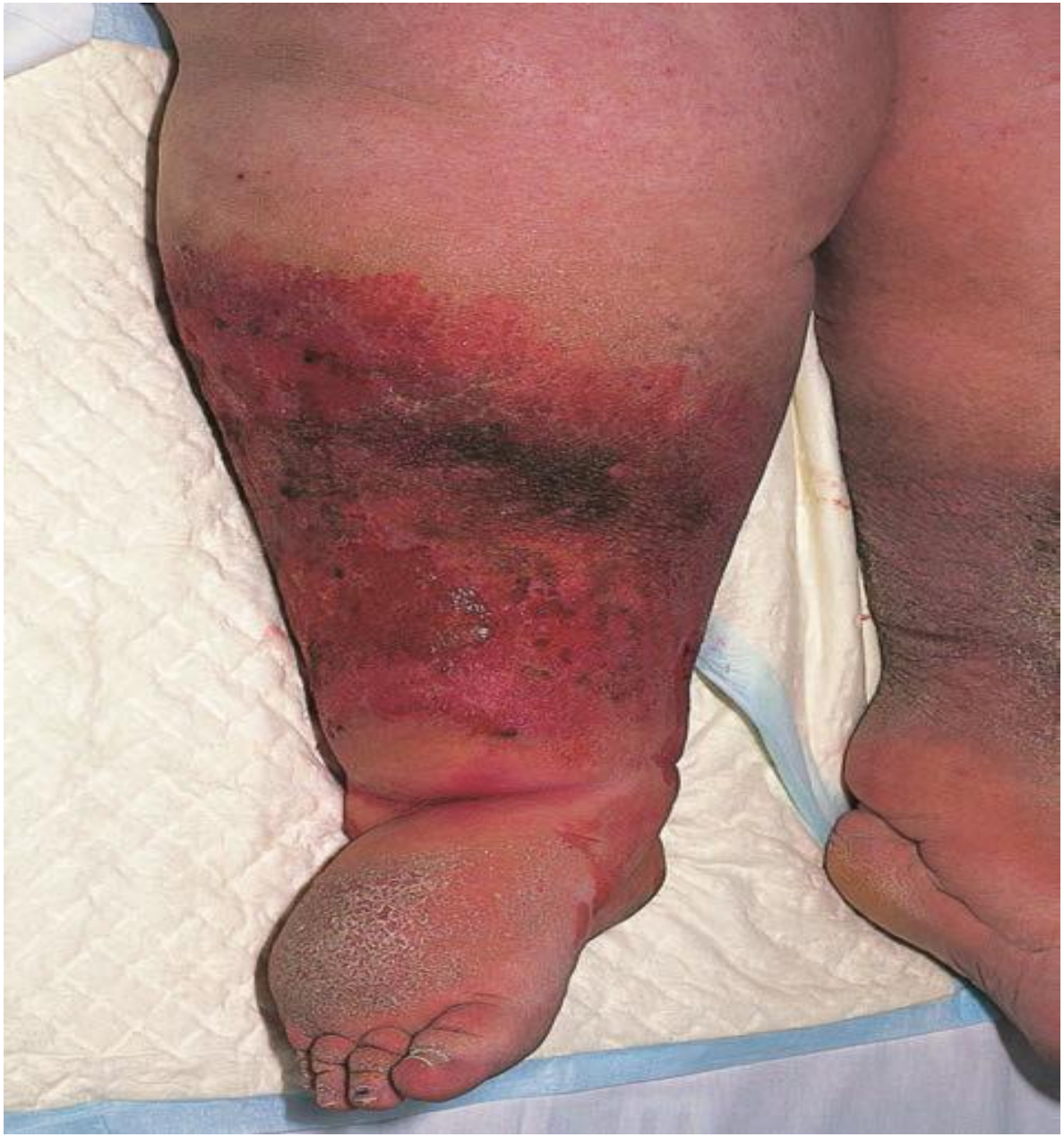
Lymphoedema is the end result of insufficient lymphatic outflow due to aplasia, hypoplasia, primary decreased lymphatic contractility or inflammatory obliteration. Lymphostasis leads to the accumulation of fluid, proteins, growth factors and other active peptide moieties, glycosaminoglycans and particulate matter, including bacteria. The end result is protein-rich oedema fluid, increased deposition of ground substance, subdermal fibrosis, and dermal thickening and proliferation.

Classification

Two main types of lymphoedema are recognised:

1 Primary lymphoedema, in which the cause is unknown (or at least uncertain and unproven); it is thought to be caused by 'congenital lymphatic dysplasia'.

2 Secondary or acquired lymphoedema, in which there is a clear underlying cause.



The lower leg of a patient with typical lymphoedema

Aetiology and classification of lymphoedema

Primary lymphoedema

- Congenital
- Praecox
- Tarda

Secondary lymphoedema

- Parasitic as filariasis
- Fungal as tinea pedis
- Exposure to foreign body materials as silica particles
- Primary lymphatic malignancy
- Metastatic spread to lymph nodes
- Radiotherapy to LN
- Surgical excision of LN
- Trauma particularly degloving injuries
- Superficial thrombophlebitis
- DVT

Risk factors for lymphoedema

Upper limb/trunk

- Axillary LN dissection
- Scar formation, fibrosis and radio dermatitis following radiotherapy
- Drain/wound complications or infection
- Seroma formation
- Advanced cancer
- Obesity
- Congenital predisposition
- Trauma
- Chronic skin disorders and inflammations
- Hypertension
- Taxane chemotherapy
- Insertion of pacemaker
- Arteiovenous shunt
- Air travel
- Living in or visiting an area for endemic lymphatic filariasis

Lower limb

- Inguinal LN dissection
- Radiotherapy
- Recurrent soft tissue infection at the same site
- Obesity
- Varicose vein stripping
- Congenital predisposition
- Advanced cancer
- Orthopedic procedures
- Thrombophlebitis and chronic venous insufficiency
- Any unresolved asymmetric oedema
- Chronic skin disorders and inflammations
- Concurrent illness as phlebitis, hyperthyroidism,renal or cardiac disease
- Immobilization
- Living in or visiting an area for endemic lymphatic filariasis

Primary Lymphoedema

Pathophysiology

Non familial primary lymphoedema

It is likely that most non familial primary lymphoedema is due to chronic injury over many years due to seemingly trivial (but repeated) bacterial and/or fungal infections, insect bites, barefoot walking, deep venous thrombosis or episodes of superficial thrombophlehitis.

Primary lymphoedema is much more common in the legs than the arms. This may be due to gravity and the fact that the lymphatic system of the leg is less well developed than that of the arm or the increased susceptibility of the lower extremity to trauma and/or infection.

Familial primary lymphoedema

In familial cases it is assumed that there must be some genetic susceptibility of the lymphatic system to such injury. ***This may be:***

- A structural problem such as aplasia or hypoplasia
- A functional problem such as defective lymphatic contractility;
- An immune deficiency.

However, at the present time, the exact mechanisms causing familial primary lymphoedema remain uncertain.

Lymphoedema congenita

Congenital lymphoedema (onset at or within a year of birth) is more common in males, more likely to be bilateral and to involve the whole leg, and accounts for less than 5 per cent of primary lymphoedema. Milroy's disease describes familial lymphoedema that is present at birth or is noticed shortly thereafter.

Lymphoedema praecox

Lymphoedema praecox (onset from 1 to 35 years of age) is three times more common in females than males, has a peak incidence shortly after menarche, is three times more likely to be unilateral than bilateral, usually only extends to the knee and accounts for about 20 per cent of primary lymphoedema. The familial form is called Meiges disease and account about one third of all cases

Lymphoedema tarda

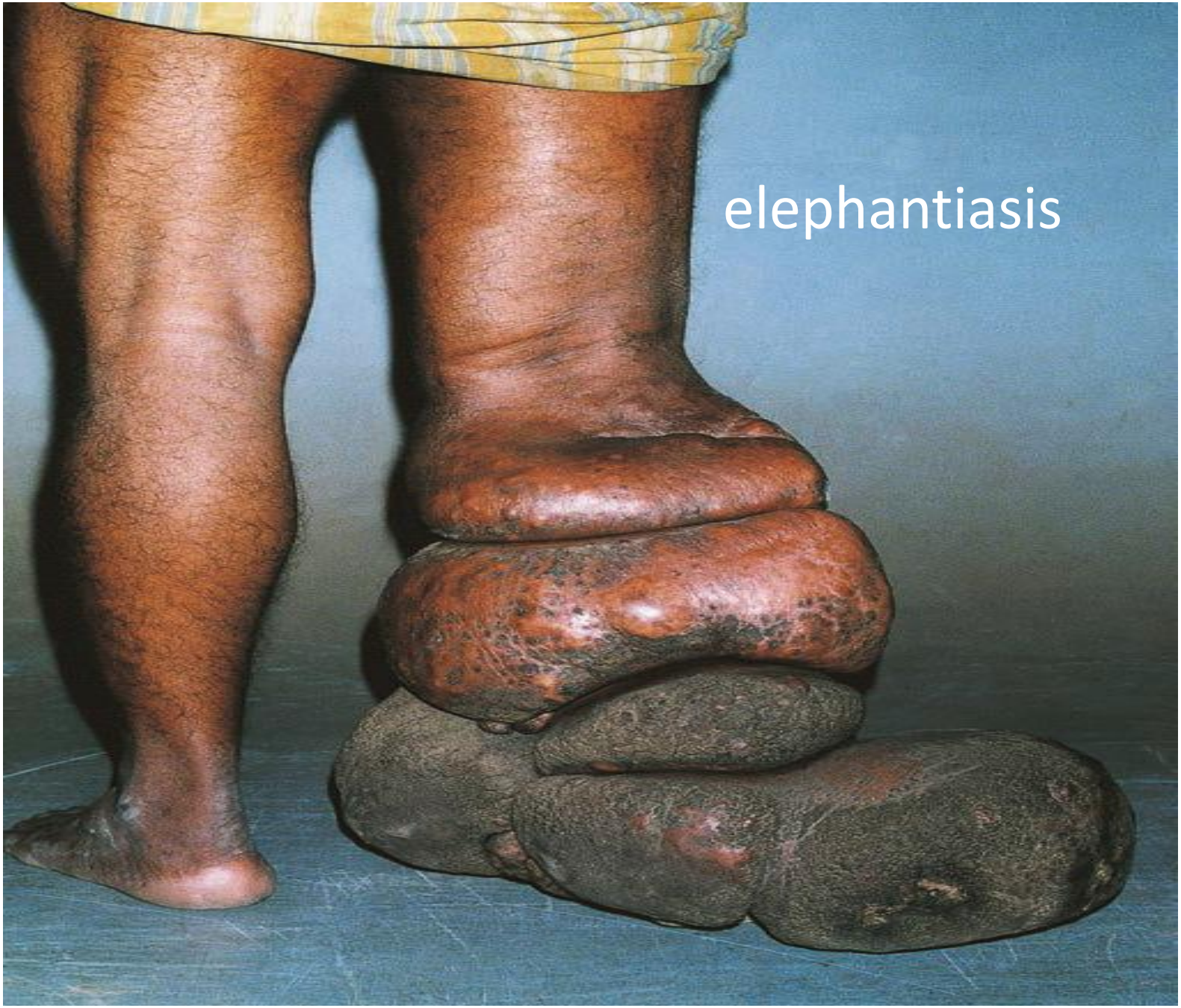
Lymphoedema tarda develops after the age of 35 years but, in practice, is a disease of middle age. It is often associated with obesity and, histologically, lymph nodes are replaced with fatty and fibrous tissue. The cause is unknown. Lymphoedema developing for the first time in later life should prompt a thorough search for underlying malignancy, particularly of the pelvic organs, prostate and external genitalia; such malignancy may be found in up to 10 per cent of patients.

SecondaryLymphoedema

This is the most common form of lymphoedema. There are several well recognised causes.

Filariasis

This is the commonest cause of lymphoedema worldwide, affecting up to 100 million individuals. It is particularly prevalent in Africa, India and South America where 5—10 per cent of the population may be affected. *The viviparous nematode Wucheria bancrofti*, whose only host is humans, is responsible for 90 per cent of cases and is spread by the mosquito. The disease is associated with poor sanitation. The parasite enters lymphatics from the blood and lodges in lymph nodes where it causes fibrosis and obstruction. Proximal lymphatics become grossly dilated with adult parasites. The degree of oedema is often massive, in which case it is termed 'elephantiasis'.


elephantiasis

Diagnosis

1) Immature parasites (microfilariae) enter the blood at night and can be identified on a blood smear, a centrifuged specimen of urine or in lymph itself.

2) A complement fixation test is also available and is positive in present or past infection.

3) Eosinophilia is usually present.

Treatment

Diethylcarbamazine destroys the parasites but does not reverse the lymphatic changes. Once the infection has been cleared treatment is as for primary lymphoedema.

Investigation of lymphoedema

Routine' tests

A full blood count, plasma urea and electrolytes, creatinine, liver function tests, chest radiograph.

Contrast lymphangiography

It is now generally reserved for preoperative evaluation of patients with megalymphatics who are being considered for bypass or fistula ligation.

Isotope lymphoscintigraphy

This has largely replaced contrast lymphangiography and is used in most centres as the primary diagnostic technique.

Computerised tomography

The main role of CT is to exclude pelvic or abdominal mass lesions.

Magnetic resonance Imaging

Magnetic resonance imaging can provide clear images of lymphatic channels and lymph nodes, and can also distinguish venous and lymphatic disease as the cause of a swollen limb.

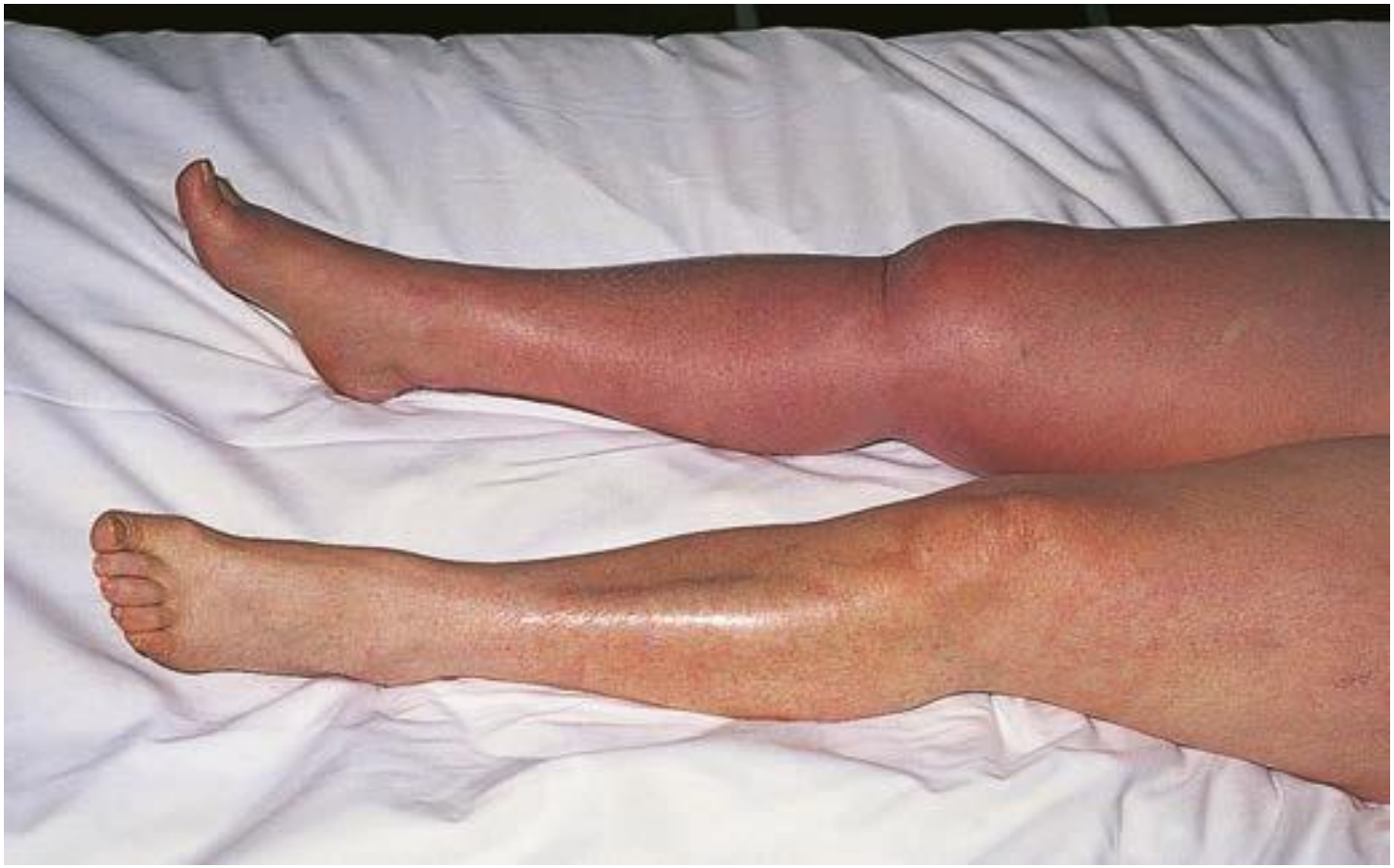
Pathological examination

In cases where malignancy is suspected, samples of lymph nodes may be obtained by fine needle aspiration, needle core biopsy or surgical excision.

Management of lymphoodema

Physical methods

The patient should elevate the foot above the level of the hip when sitting, elevate the foot of the bed when sleeping and avoid prolonged standing. Below-knee stockings are usually sufficient.



Drugs

Diuretics are of no value in pure lymphoedema. Their use is associated with side effects including electrolyte disturbance. Antibiotics should be prescribed promptly for cellulitis.Antibiotics should be continued for at least 7 days or until all signs and symptoms have abated. Fungal infection (tinea pedis) must be treated aggressively; topical clotrimazole 1 per cent or miconazole 2 per cent used regularly is sufficient in most cases, but in refractory situations systemic griseofulvin 250—1000 mg daily may be required. The feet must be dried after washing and the skin kept clean and supple with water-based emollients to prevent entry of bacteria.

Surgery

Only a small minority of patients with lymphoedema benefits from surgery. Operations fall into two categories: ***bypass procedures and reduction procedures.***

Bypass procedures

Limb reduction procedures

Lymphangiomas

Lymphangioma circumscriptum

This involves superficial proliferation of capillary sized lymphatic vessels which comprise fluid-filled vesicles on the surface and larger cisternae in the subcutaneous tissues and even adjacent muscles. The affected skin and a generous amount of subcutaneous tissue containing the vessels may be excised if they cause symptoms.

Cystic hygroma

Cystic hygroma is an abnormal lymph-filled, often multilocular, space which usually presents in childhood as a soft, brilliantly transluminable swelling in the base of the neck. They develop from primitive lymph cisterns. It behaves like a benign tumour and grows gradually in size, leading to cosmetic problems and compression of surrounding structures. Recurrence is common after simple aspiration and injection of sclerosant. Excision is technically challenging due to the large number of vital structures in the vicinity.


Thank you