Primary lymphoedema in a 45-year-old woman

Commentary by **JOHN P. HARRIS** AM, MS, FRCS, FACS, FRACS, DDU(Vascular)

Secondary lymphoedema was ruled out in this woman with swelling of her legs, and she has no venous obstruction or reflux.

Case scenario

Miriam, a 45-year-old shop assistant, presented with mild symmetrical pitting oedema of both legs of relatively abrupt onset. She said that the swelling would subside overnight but redevelop during the day, causing her legs to ache and feel heavy.

She was extensively investigated but no pathology was found. In particular, she was neither anaemic nor hypertensive, she was euthyroid, she had no deep vein thromboses or venous valvular incompetence, and she had no abdominal or pelvic lesion on CT scan. She was taking no medication and she otherwise felt well. She was overweight (BMI, 30 kg/m^2) but there had been little change in her weight over the preceding five years.

It was finally assumed that she had developed impaired lymphatic drainage. What is likely to have caused this?

Commentary

Sudden onset of bilateral pitting oedema may be due to central causes such as congestive cardiac failure, renal disease and low protein states, or to local causes, the most concerning being acute deep venous thrombosis. These have presumably been excluded by her extensive investigations. In the absence of venous obstruction or reflux, the presentation is therefore consistent with lymphoedema. Lymphoedema is usually secondary, with the aetiology obvious from the history and physical examination. Although filariasis is the most common cause worldwide, this infestation is rare in Australia. A more important secondary cause of lymphoedema in the Australian setting is as a complication of radical lymph node clearance performed as part of a surgical approach to malignancy, particularly if followed by radiotherapy.

In Miriam's case, her lower extremity swelling is likely to be due to primary lymphoedema because there is no obvious aetiological factor present and venous obstruction and severe reflux have been excluded.

Primary lymphoedema is due to hypoplasia of the lymphatics and can be classified according to the age of initial presentation. A congenital form, detected in the first year of life, may be familial (such as Milroy's disease, with a family history consistent with an autosomal dominant inheritance) or sporadic. Lymphoedema praecox occurs between the ages of 1 and 35 years, and is the most common form of primary lymphoedema. Lymphoedema tarda occurs after the age of 35 years, and hence would apply to Miriam.

Lymphoscintigraphy

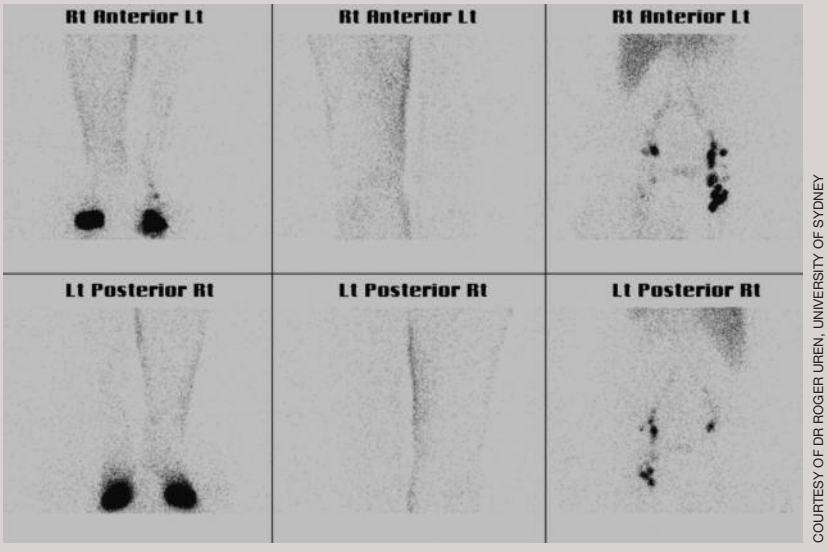

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Figure. A lymphoscintigram of a patient with congenital lymphoedema (delayed two-hour images – images on left, lower leg; centre, knee and lower thigh; right, upper thigh and groin). The images show dermal backflow in the affected leg and fewer lymph nodes in the right groin. (Note that this patient is not the patient in the case scenario.)

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The initial swelling may be precipitated by a mild systemic illness or any event that may add to lymphatic load. This swelling may then be aggravated if there are repetitive episodes of lower extremity cellulitis, further damaging residual lymphatics. Lymphoedema may also follow lower extremity operations such as varicose veins surgery or femoropopliteal bypass.

Lymphoedema is characteristically 'non-pitting' (pressure applied to the skin does not result in a persistent indentation) because of the high protein content of the interstitial fluid, although pitting can be present in the earlier stage. The swelling often extends to the dorsum of the foot, which is a region relatively spared with venous causes of extremity swelling.

The most specific test to confirm the diagnosis is lymphoscintigraphy, in which a radiotracer (99m-technetium [Tc] antimony sulfide colloid) is injected into a web space and then followed up the leg as it is cleared by the lymphatics. With lymphoedema, the draining is delayed. This imaging may show other features of diagnostic use, such as hypoplasia of the lymph glands or dermal backflow – as illustrated by the lymphoscintigram in the figure, which shows features typical of congenital lymphoedema.

Miriam's investigations have excluded systemic causes of oedema such as heart failure, renal failure and hypo-osmotic states. Retroperitoneal lymphadenopathy may present as lymphoedema, but no abdominal or pelvic lesion was seen on her CT scan. If her left limb is predominantly affected, it is important to examine the left common iliac vein because occasionally this can be compressed by the right common iliac artery.

Chronic lymphoedema can be helped by weight reduction and supervised massage programs, compression stockings and early antibiotic cover at the first sign of any infection to minimise further damage to the lymphatics. Debulking surgery is sometimes appropriate in severe cases but such operations are associated with considerable wound morbidity. MT

Further reading

1. Tiwari A, Chend KS, Button M, Myint F, Hamilton G. Differential diagnosis, investigation, and current treatment of lower limb lymphedema. Arch Surg 2003; 138: 152-161.

2. Sieggreen MY, Kline RA. Current concepts in lymphedema management. Adv Skin Wound Care 2004; 17: 174-178.

DECLARATION OF INTEREST: None.