Primary lymphoedema

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DESCRIPTION

A 32-year-old woman presented with a 20-year history of slowly progressive leg oedema with recurrent cellulitis. She was referred from her primary care physician. Her mother had the same condition. Physical examination revealed non-pitting bilateral leg oedema (figure 1). Stemmer's sign was positive. Lymphoscintigraphy showed non-visualisation of inguinal nodes 30 min postexercise and dermal backflow in the lower limbs (figure 2). Compression garments were used but failed. She underwent bilateral lower limb supermicrosurgical lymphaticovenular anastomosis (two times anastomosis and four times incisions) and her oedema markedly improved. Four years after the operation, she was well without relapse.

Primary lymphoedema is due to a congenital and/or inherited condition associated with pathological development of the lymphatic vessels. Lymphoedema praecox is the most common form of primary lymphoedema and generally occurs during puberty or pregnancy with onset prior to age 35 years.¹ It is found in about 1 in 100 000 male and 1 in 400 000 female, and 10% of cases are familial with an autosomal dominant inheritance pattern.¹ The most common presentation is oedema of the foot or ankle.² Lymphoedema is an important risk factor for cellulitis. Testing for Stemmer's sign consists of trying to lift the skin over the second toe. If the skin cannot be lifted, Stemmer's sign is positive. A positive Stemmer's sign is indicative of lymphoedema because the high protein and fat contents of lymph fluid induce fibrosis.³ Differential diagnosis of lymphoedema is lipoedema, which occurs from hip to ankle, usually sparing the feet.³ Lymphoscintigraphy and

Figure 2 Preoperative lymphoscintigraphy. Lymphoscintigraphy revealed no migration of radiopharmaceutical agent in the lower limbs.

lymphangiography are useful imaging modality to rule out lymphoedema as a cause of leg swelling. Ultrasonography can be used to evaluate the lymphatic and venous systems, such as deep vein thrombosis and arteriovenous fistula. CT and MRI are accessible to detect causes of obstructive secondary lymphoedema. Supermicrosurgical lymphaticovenular anastomosis is assumed to have a valuable place in the treatment of lymphoedema.

Learning points

- ▶ Stemmer's sign is indicative of lymphoedema.
- ▶ Differential diagnosis of lymphoedema is lipoedema, usually sparing the feet.

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Figure 1 Non-pitting bilateral leg oedema.

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