SURGICAL TREATMENT OF MILROY’S DISEASE

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Abstract: Milroy’s disease is a form of non-syndromic primary congenital lymphedema, mainly associated with lower limb lymphedemas but sometimes also upper limb and genital lymphedema can be observed. Clinical presentation is defined by swelling of the extremities that may be congenital or developing soon after birth, characterized by progression in degree. Other possible causes of lymphedema have to be evaluated and excluded. In China Medical University Hospital 9 patients were diagnosed with Milroy’s disease and treated over an 8 years period time. 8 patients presented lower extremities lymphedema, 4 of which bilateral, and 1 patient was affected by unilateral upper extremity lymphedema. Being the case series treated over a long period of time, treatment strategy changed and evolved over time. The first 3 patients presented at an older age with severe lymphedema and were treated with Charles procedure. 3 patients with moderate lymphedema were treated with extensive therapeutic lipectomy. The latest 3 patients treated, which also presented with moderate lymphedema were treated at a mean age 16 y.o. with microvascular LNFT from gastroepiploic region. Postoperatively patients were evaluated by circumferential and tonicity measurements and lymphoscintigraphy. In our case series all patients benefitted from surgical treatment in terms of reduced circumferential measures and reduced infection rate but patient treated with LNFT obtained a better aesthetic result still obtaining a substantial circumferential reduction already at an early six months follow up. In these cases repeated lymphoscintigraphy showed improvement of the lymphatic system in the limbs treated with LNFT. In our practice, early stage LNFT proved to be a reliable technique in moderate cases of congenital lymphedema. This is concordant with literature findings, suggesting that autologous lymph node transplantation induces the neoformation of lymphatic vessels improving
uptake of lymph by the local lymphatic system which appears to be altered in
Milroy's disease.

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