

RESEARCH ARTICLE

Intra-abdominal chylovenous bypass treats retroperitoneal lymphangiomas

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Abstract

Background: Retroperitoneal lymphangiomas (RL) is a rare form of primary lymphedema featuring aberrant retroperitoneal lymphatic proliferation. It causes recurrent cellulitis, repeated interventions, and poor life quality. This study aimed to investigate proper diagnostic criteria and surgical outcomes for RL with extremity lymphedema.

Methods: Between 2012 and 2018, 44 primary lower-extremity lymphedema cases received lymphoscintigraphy, magnetic resonance imaging, and single-photon electron computed tomography to detect RL. RL patients underwent vascularized lymph node transfers (VLNT) for extremity lymphedema and intra-abdominal side-to-end chylovenous bypasses (CVB) for chylous ascites. Complications, CVB patency, and quality of life were evaluated postoperatively.

Results: Six RL patients (mean age of 30.3 years) had chylous ascites with five had lower-extremity lymphedema. All CVBs remained patent, though one required re-anastomosis, giving a 100% patency rate. Four unilateral and one bilateral extremity lymphedema underwent six VLNTs with 100% flap survival. Patients reported improved quality of life ($P = 0.023$), decreased cellulitis incidence ($P = 0.041$), and improved mean lymphedema circumference ($P = 0.043$). All patients resumed a normal diet and activity.

Conclusions: Evaluating primary lower-extremity lymphedema patients with MRI and SPECT could reveal a 13.6% prevalence of RL and guide treatment of refractory extremity lymphedema. Intra-abdominal CVB with VLNT effectively treated RL with chylous ascites and extremity lymphedema.

KEYWORDS

chylovenous bypass, extremity lymphedema, retroperitoneal lymphangiomas, vascularized lymph node transfer

1 | INTRODUCTION

Lymphangiomas is a rare condition of unknown incidence with diffuse involvement of all tissue types through hyperproliferation of lymphatic vasculature, causing multiple lymphangiomas.^{1,2} This condition affects more children than adults.^{2,3} Lymphangiomas

is classified as either generalized lymphangiomas or Gorham Stout syndrome, with the former primarily involving soft tissue and viscera and secondary osseous involvement, while the latter features lymphovascular proliferation within bones and occasional soft tissue/viscera involvement.^{3,4} Major complications of lymphangiomas include chylothorax, chylous pericardial effusions, and chylous