



Vascularized Lymph Node Flap Transfer and Lymphovenous Anastomosis for Klippel-Trenaunay Syndrome with Congenital Lymphedema

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Summary: A female patient with Klippel-Trenaunay syndrome, including hypertrophic bone and soft tissue in the forelimbs, bilateral lower limbs lymphedema, port-wine stains, and superficial veins of Servelle, was presented. The diagnosis of lymphedema was confirmed by lymphoscintigraphy and indocyanine green lymphography. The novel treatments consisted of vascularized lymph node transplantation to the left lymphedematous extremity and lymphovenous anastomosis to the right lymphedematous extremity. Significant improvements in subjective and objective clinical outcome were observed early in the postoperative period with continued improvements during the follow-up period. (*Plast Reconstr Surg Glob Open* 2014;XXX:00-00; doi: 10.1097/GOX.000000000000099; Published online Day Month 2014.)

CASE PRESENTATION

A 13-year-old female patient diagnosed with Klippel-Trenaunay syndrome (KTS) presented port-wine stains along the left lower extremity and trunk, anomalous superficial vein along the lateral border of left thigh, or vein of Servelle¹ associated with severe swelling over her bilateral lower limbs with disproportionate growth since early childhood. The superficial and deep venous systems in other parts of both legs were unremarkable with no signs of thrombosis or insufficiency.

Lymphoscintigraphy demonstrated abnormal drainage pattern in bilateral lower extremities with diagnosis of primary lymphedema (Fig. 1).

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OPERATIVE PROCEDURE

Delayed lymphatic flow in both lower extremities was visualized after injecting 0.2 mL of indocyanine green (ICG) before surgery. Tortuous lymphatic channels were identified on the dorsum of right foot, and one lymphaticovenous anastomosis was performed in a side-to-end configuration. For the left lower limb, a right vascularized submental lymph node flap was transferred to the left ankle according to the previously published technique² (Fig. 2). When the anastomoses of the pedicle were completed, 0.01 mL of ICG was injected superficially in a transferred lymph node. The fluorescence was observed with a camcorder (Sony HD Handycam CM05, Sony) (Fig. 2).

Symptomatic improvement in both extremities were reported, with circumferential reduction rates in right lower limb at 15 cm above knee, 15 cm below knee, and 10 cm above ankle were 50%, 53.3%, and 33%, respectively. Likewise, in left lower limb, the circumferential reduction rates were 67% at 15 cm above knee and 61% at 15 cm below knee (Figs. 3 and 4). At a 3-month follow-up, circumferential reduction was maintained in both extremities. According to the "Quality of Life Measure for Limb Lymphedema,"³ the preoperative overall score was

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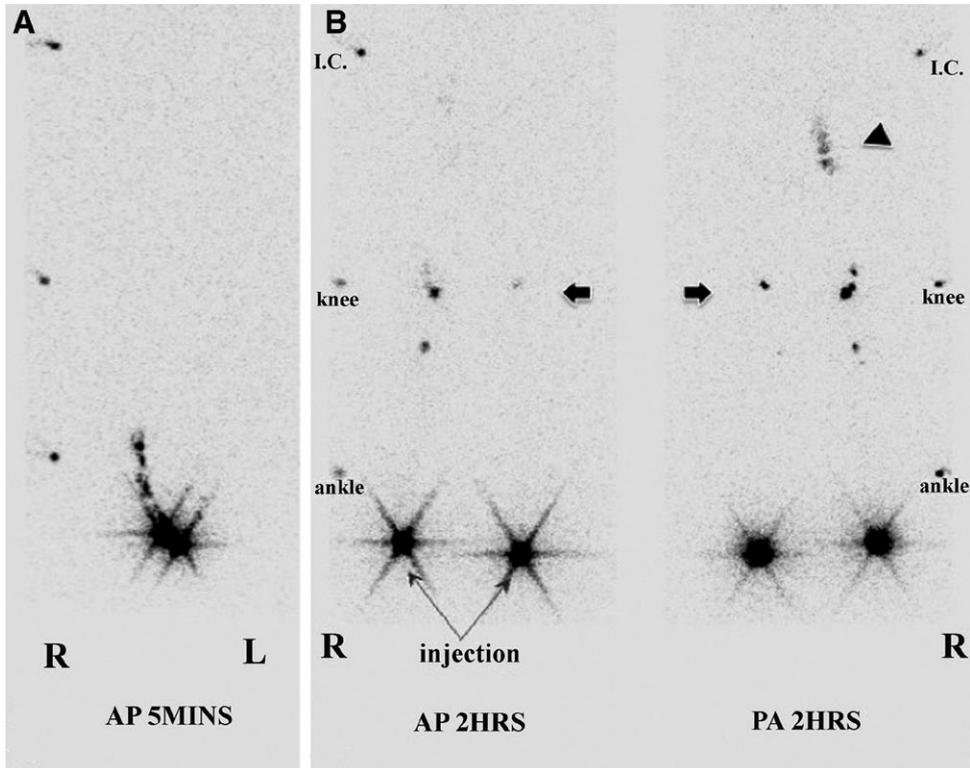


Fig. 1. Lymphoscintigraphy revealed lymphatic tract on right foot but not on left foot at 5 minutes after Technetium-99 injection (A). Note that inguinal nodes appeared on right side but not on left side (arrow head) after 2 hours on the posterior-anterior (PA) view (B). Deep lymphatic trunks and several popliteal nodes were visible on both sides (black arrow) after 2 hours injection (B).

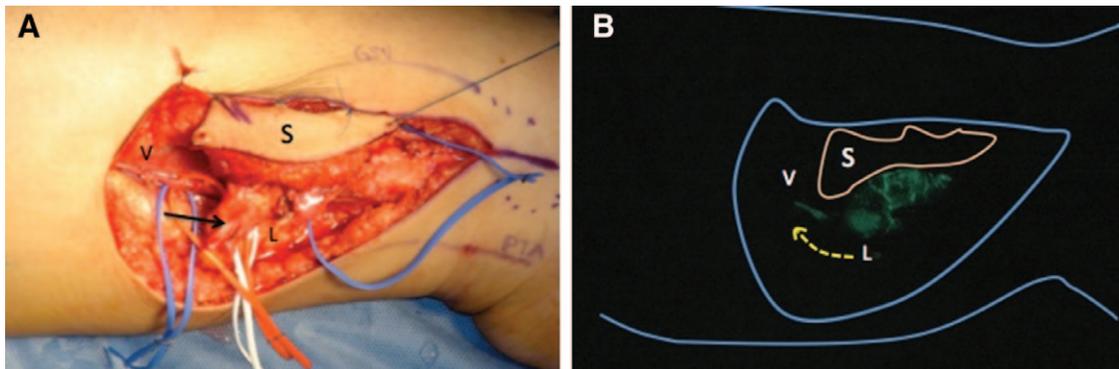


Fig. 2. A, Intraoperative picture after completing the vascular anastomosis of the vascularized submental lymph node flap on the left medial ankle. The arrow indicated the transferred lymph node, in which 0.01 mL of indocyanine green was injected. B, The fluorescent area showed how the lymph traveled from the lymph node to the flap's donor vein and then to the recipient vein that is captured by a near-infrared camera. The dotted line indicated the direction of the lymph. L indicates transferred lymph node; S, skin paddle of the submental flap; V, recipient vein.

43 and at 3-month postoperative was 67, indicating patient's satisfaction in improvement of quality of life after the surgery.

DISCUSSION

KTS is an uncommon congenital vascular anomaly described in 1900 by Maurice Klippel and Paul

Trenaunay.⁴ The classic clinical triad consists of capillary malformation, soft-tissue and bone hypertrophy, and lateral varicosities. All 3 features of KTS are present in approximately 63% of patients, and the remaining 37% have 2 of the 3 diagnostic features.¹ KTS is unilateral in 85% of the patients, bilateral in 12.5%, and crossed-bilateral in 2.5%.¹

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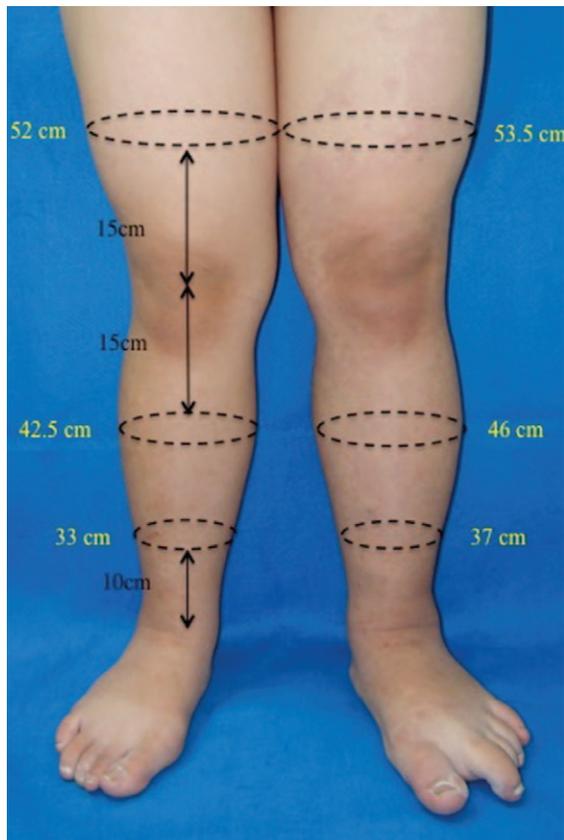


Fig. 3. Preoperative image of the patient. The port-wine stain is remarkable in the left thigh.

Few previous studies have focused on the lymphatic system derangements within this syndrome. The frequency and the nature of the lymphatic system involvement in KTS are poorly addressed due to the lack of a proper diagnostic method or the rare occurrence of this syndrome.⁴ According to Servalles,¹ 3 groups of lymphatic malformations may be associated with KTS: lymphatic malformations resulting from obstruction of the deep veins, common lymphedema, or the presence of chyliferous vessels. Other authors^{5,6} stated an intimate relationship between the venous and lymphatic system in the functional, structural, and developmental period. These previous theories can explain the presence of lymphatic impairment associated with venous malformations in KTS. The development of accurate diagnostic tools, such as lymphoscintigraphy and ICG lymphography,⁷ has allowed for functional analysis of the potential impaired lymphatic system. In the present case, the patient was diagnosed with KTS with the presence of forelimb soft-tissue and bony hypertrophy, port-wine stains in the left lower limb and trunk, and bilateral lower limb lymphedema, confirmed by lymphoscintigraphy and ICG lymphography.

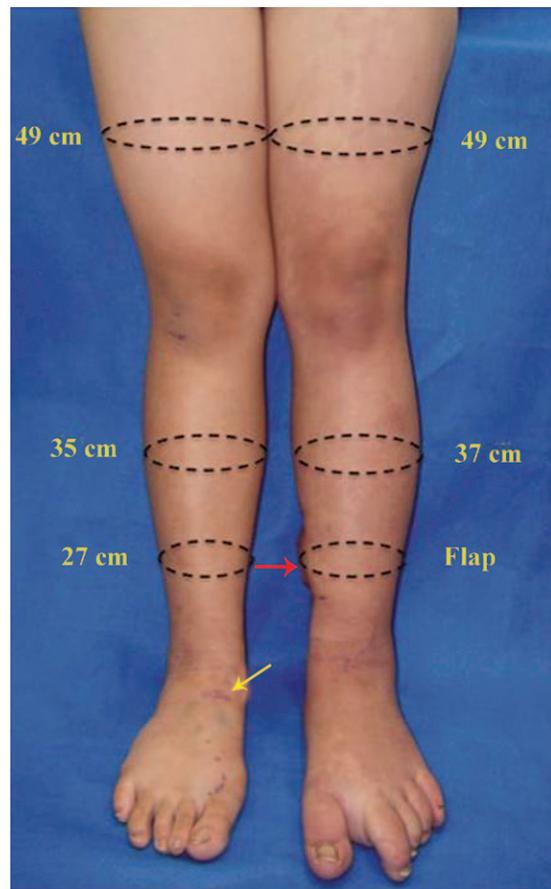


Fig. 4. Image at 3 months postoperatively. The vascularized submental lymph node flap (red arrow) can be partially seen in the medial aspect of the left ankle. A linear scar (yellow arrow) on the right dorsal ankle is the location of lymphaticovenous anastomosis. Measures are given in centimeters.

Treatment of KTS has traditionally been conservative in nature.⁸⁻¹¹ Management of this syndrome is focused primarily on the treatment of the complications that arise from these vascular malformations.¹¹ However, associated lymphedema can be managed as a separate entity. Classically, primary lymphedema is treated conservatively with the use of massage therapy, intermittent pneumatic compression, elastic garments, and compression bandages.⁴ As experience has grown in the microsurgical treatment of secondary lymphedema, improvements in clinical outcomes and understanding of the disease process has significantly presented. During the last 2 decades, techniques have been described that create neolymphatic connections allowing for venous drainage of lymphatic fluid.^{12,13} The 2 most common microsurgical procedures include lymphaticovenous anastomosis and vascularized lymph node transplantation. Lymphaticovenous anastomosis has been shown

to improve symptoms following the development of secondary mild lymphedema.^{14,15} Vascularized lymph node transplantation has shown significant improvements in limb circumference reduction for both moderate and severe secondary lymphedema.^{2,16,17} Few reports have investigated the role of these microsurgical procedures in congenital and primary lymphedema.¹³ Limitations of lymphaticovenous anastomosis in congenital cases have been the uncertainty of patent lymphatic ducts.

The role of vascularized lymph node transplantation in the treatment of congenital lymphedema has not been previously explored. At our institution, we have been using vascularized lymph node flap transplantation with proven successful outcomes² and developed animal models for deeper understanding of the pathophysiology of lymphedema.¹⁸ Intrinsic lymphovenous connections within the transplanted lymph node tissue allow for venous drainage of lymph from the local environment.^{2,16,18} Placement of these lymphatic pumps in nonanatomic locations near areas of lymphedema allows for optimal clearance and rerouting of interstitial fluid into the venous system. This new route of lymphatic drainage can be demonstrated by direct visualization of fluorescence in the recipient vein immediately after injection of ICG into the transferred lymph nodes inside the flap after completion of pedicle anastomosis (Fig. 2).¹⁹ Initially, the lymph is driven by interstitial pressure gradients between the edematous limb and the lymph node flap. Early clearance of lymphatic fluid is followed by catchment effect and neolymphatic generation.^{7,17} These characteristics may make this method a preferable option for primary lymphedema as its mechanism of action is independent from the number and function of viable lymphatic ducts.

As knowledge in using these novel techniques grows, increasing indications will become apparent in lymphedematous conditions. KTS represents a challenging clinical condition with few good potentially curative options. Symptomatic lymphedema related to this congenital syndrome may now have a valuable treatment option providing significant relief to affected patients. In this case, early improvements in both objective and subjective patient outcomes are mirrored by sustained and continued improvement in the postoperative period.

CONCLUSIONS

In summary, vascularized lymph node transplantation and lymphaticovenous anastomosis can yield significant improvement in patients with primary lower extremities lymphedema.

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