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CASE REPORT
Reconstructive

Successful Treatment of Rheumatoid Lymphedema with Lymphatic Venous Anastomosis

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Summary: Rheumatoid lymphedema is a rare but severely disabling condition caused by reduced lymphatic drainage. Most treatment methods are conservative and may lead to the exacerbation of lymphedema. Lymphatic venous anastomosis (LVA) is an effective treatment for lymphedema after surgery involving the lymphatic system, such as lymph node dissection for cancer treatment. LVA has not been used to treat rheumatoid lymphedema. We present a case of rheumatoid lymphedema treated with surgical procedures, including LVA. Following LVA, objective and subjective symptom relief was noted, along with decreased swelling and pain in the affected area. The postoperative course was uneventful. LVA for the treatment of rheumatoid lymphedema may provide definitive clinical improvements. *(Plast Reconstr Surg Glob Open 2021;9:e3763; doi: 10.1097/GOX.0000000000003763; Published online 16 August 2021.)*

Rheumatoid arthritis (RA) is a chronic polyarthritis of unknown etiology that affects approximately 1% of the world's population.[1] A small proportion of patients with RA develop peripheral nonpitting edema that is not correlated with other factors, such as anemia, hypoalbuminemia, fluid retention, or venous obstruction.[2] Patients with RA lymphedema could have chronic, diffuse, and painful swelling in their extremities.[2]

Conservative treatment of rheumatoid lymphedema includes steroid injections, tocilizumab, stretch bandages, and manual lymph drainage.[2] However, some patients experience an exacerbation of lymphedema while receiving conservative treatment. Currently, reconstructive lymphatic surgery for the treatment of rheumatoid lymphedema refractory to conservative treatment has not been reported.

Several reconstructive lymphatic techniques have been used for the treatment of lymphedema that developed after surgery that involved the lymphatic system, such as lymph node dissection during cancer treatment.[3] Lymphatic venous anastomosis (LVA) and liposuction are becoming popular and effective alternatives to conservative therapy for lymphedema; however, no reports regarding the use of these procedures for the treatment of rheumatoid lymphedema currently exist. Herein, we report a case of rheumatoid lymphedema resistant to conservative treatment that was successfully treated with LVA. The patient provided informed consent for the publication of this report.

CASE

A 50-year-old woman presented with a 3-year history of progressive symmetrical edema, intractable pain, and bilateral heaviness in her lower extremities. Physical examination upon admission revealed significant bilateral pitting edema in the lower extremities (Fig. 1) with no obvious cause. The lower extremity lymphedema index,[4] which provides an objective assessment of the severity of lymphedema, was 284 on the right lower extremity and 297 on the left lower extremity before the surgery. The edema was nonresponsive to 1-year compression therapy consisting of 30 mm Hg stockings. The patient had a 10-year history of RA and was in remission after treatment with methotrexate (MTX; 8 mg/week), prednisolone (3 mg/day), and nonsteroidal antiinflammatory drugs.

Indocyanine green lymphography[5] revealed signs of dermal backflow in both lower extremities (Fig. 2). The patient was diagnosed with rheumatoid lymphedema.

LVA (Fig. 3) was performed to reduce pain, volume, and limb circumference of the lower extremities. LVA was performed in the right and left lower extremities three and four times, respectively. After 1 year from the first LVA, the patient's pain and bilateral lower extremity volume decreased; however, some edema remained. We considered that the edema had improved because of

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the lymphatic reconstruction; however, there was diffusely accumulated adipose tissue in the lower extremity due to the retained lymphedema. Therefore, the patient underwent a second LVA, this time with liposuction. LVA was performed three times in both lower extremities during the second surgery. Liposuction was performed for both

Fig. 1. Preoperative image of a 50-year-old woman with bilateral rheumatoid lymphedema in the lower extremities.

Fig. 2. Indocyanine green lymphography showing marked dermal backflow in both lower extremities.

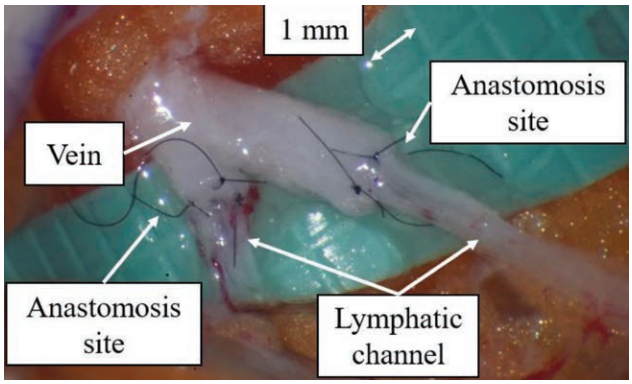

Fig. 3. LVA was performed for the treatment of rheumatoid lymphedema.

lower extremities after LVA, and 500 ml liposuction fluid was removed from each lower extremity.

Eight months after the last surgery, the lymphedema had significantly reduced, and the patient's mobility had improved (Fig. 4). The lower extremity lymphedema index decreased to 245 on the right lower extremity and 240 on the left lower extremity. During this follow-up period, the patient used the compression bandage to prevent the exacerbation of the edema and had no recurrence. There were no changes in the patient's RA symptoms or medications.

DISCUSSION

Edema is caused by an imbalance in microvascular filtration and lymphatic drainage.[5] In patients with RA, filtration edema can be caused by acute synovial inflammation with excessive lymph production.[6] Lymphedema in patients with RA is reportedly due to lymphatic obstruction by fibrin-degradation products that block the lymphatic channels.[2] Lymphatic flow contraction has been reported to result from dysfunctional lymphatic smooth muscle cells due to RA inflammation.[7] Li et al demonstrated that impaired lymphatic drainage from an inflamed synovium is associated with joint flares in murine models of inflammatory-erosive arthritis.[6] Previous studies have indicated an association between lymphatic flow and RA. Indocyanine green lymphography could be used to investigate the correlation between RA flares and lymphatic drainage.[7]

Lymphatic drainage route reconstruction is another option for the treatment of lymphedema. LVA has been performed to reroute the lymphatic system to vein circulation in lymphatic disorders. LVA has been reported to have excellent efficacy for decreasing limb circumference and improving subjective symptoms.[3] LVA has recently been indicated as a treatment option for a wider range of illnesses, such as coexisting Raynaud's phenomenon and lymphedema, morbidly obese patients with lymphedema, and immunosuppressant-induced lymphedema.[8–10] Currently, only conservative methods have been used for the treatment of rheumatoid lymphedema.[2] Surgical treatment might be considered in patients that are refractory to conservative treatment. Due to its fewer adverse effects compared with other surgical treatments, LVA is considered as the first-line surgical treatment for rheumatoid lymphedema.

Fig. 4. Postoperative improvement in the circumference of the patient's lower extremities.

In conclusion, we performed LVA after failure of conservative treatment. Additional LVA with liposuction was required. The surgery was successful, with no adverse

effects observed within the patient's postoperative course. LVA can potentially restore lymphatic flow in patients with rheumatoid lymphedema and might be considered as an alternative treatment option for rheumatoid lymphedema. However, in this case, the follow-up period was short; therefore, prospective studies with large patient populations and longer follow-up periods are required to verify the efficacy of LVA in rheumatoid lymphedema.

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