Combined Treatment with Lymphaticovenular Anastomosis and Ethanol Sclerotherapy for Cystic Lymphangioma in a Limb

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Summary: Several treatment options, including sclerotherapy and surgical excision, are available for the management of cystic lymphangioma. Lymphaticovenular anastomosis (LVA) has recently garnered attention in the field of microsurgery as a minimally invasive surgical reconstruction strategy. Combined treatment using surgical excision and LVA for large or persistent cystic lymphatic malformations has been reported but can be very invasive. This case report describes use of a combination of LVA and sclerotherapy to treat cystic lymphatic malformations with satisfactory results. This combination could be a complementary minimally invasive treatment for extensive cystic lymphatic malformations. The patient was an 18-year-old woman with a cystic lymphangioma from the axilla to the subclavicular area beneath pectoralis major and pectoralis minor. The size was a diameter of 12 cm and a maximum depth of 8 cm. The cystic lymphangioma was managed by combined treatment of LVA and ethanol sclerotherapy. The patient had subsequent resolution of the lymphangioma without any symptoms of lymphedema or recurrence. LVA is thought to be a less invasive treatment option when the lymphatics flowing into a cyst are detectable or can be targeted. However, this is not always the case, especially if the cyst is large, persistent, or infected. Combination of LVA with sclerotherapy is a relatively less invasive method with closure of the dead space using sclerotherapy without further exacerbation or occurrence of lymphedema. Therefore, combined treatment using LVA with sclerotherapy can be a complementary minimally invasive treatment option for a large or persistent lymphocele. (Plast Reconstr Surg Glob Open 2022;10:e4348; doi: 10.1097/GOX.0000000000004348; Published online 24 May 2022.)

Cystic lymphangioma originates in the embryonic stage of life and leads to failure of communication and lymph drainage into the venous system.1 There is no consensus on its management, which includes observation, aspiration, ligation, and excision depending on the size of the lesion, its anatomic site, and complications.2 Although sclerotherapy is thought to be less invasive than surgical excision, sclerosants cause fibrosis and obliteration of both a lymphocele and lymphatic vessels,3 thereby impairing lymphatic drainage, which increases the risk of lymphedema.4 Lymphaticovenular anastomosis (LVA) recently garnered attention in the field of microsurgery as a minimally invasive or prophylactic treatment for lymphedema5 and cystic lymphatic malformation,6 although its success rate and indications in terms of the size of the lymphocele for which it is most effective remain unclear.

Combined treatment using LVA with sclerotherapy may be an alternative minimally invasive treatment option. In this report, we describe a cystic lymphangioma in the extremities treated using LVA and ethanol sclerotherapy with satisfactory results.

CASE REPORT

The patient was an 18-year-old woman with a soft mass extending from the axilla to the anterior chest wall which she had noticed 1 month earlier (Fig. 1). Computed tomography revealed a cystic lymphangioma with a diameter of 12 cm and a maximum depth of 8 cm extending from the axilla to the subclavicular area beneath the pectoralis major and pectoralis minor (Fig. 2).

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Written informed consent was obtained from the patient. Combined treatment of LVA with sclerotherapy was planned and performed under general anesthesia. The upper extremity lymphedema index (UEL index) is calculated as follows: (sum of the squares of the circumferences at five areas in lower or upper extremity)/(body mass index). The UEL index was 110. Before surgery, indocyanine green (ICG) lymphography was performed, and 0.2 mL of ICG (Diagnogreen 0.25%; Daiichi Sankyo, Tokyo, Japan) is injected subcutaneously into both upper extremities at the first webspace of the hand and the palmar side of ulnar wrist. ICG lymphography revealed a linear pattern along the ulnar side of the right upper limb. We decided this linear pattern was the main lymphatic route for cystic lymphangioma; however, it was unclear whether this route was the only lymphatic route for the large cystic lymphangioma. Ethanol sclerotherapy was performed first. All the fluids in the cavity were initially evacuated via a small skin incision. The lymphocele is then catheterized using a round-type, 15 Fr, 5-mm diameter Blake silicone drain, (Johnson & Johnson K.K., Tokyo, Japan). Next, absolute ethanol at a volume of 50 mL was instilled and left in situ for 20 minutes. During this time, care was taken to ensure complete exposure of the entire surface area of the lymphoceles to the sclerosant. All the fluids were then discharged and saline was instilled up to one-half of the estimated volume of the cavity. Next, all the fluids were discharged again. The procedure was repeated five times, after which the catheter was left in place for drainage. Two LVAs were created at peripheral sites extending from the cyst of the right upper arm soon after ethanol sclerotherapy. Incisions were made along the marked linear pattern. A 2- to 3-cm skin incision was made, and the collecting lymphatics and subcutaneous veins were dissected and identified under a microscope.

Fig. 1. Cystic lymphangioma extending from the axilla to the subclavicular area in an 18-year-old woman.

Fig. 2. Computed tomography revealed a lymphangioma measuring 12 cm in diameter with a maximum depth of 8 cm extending from the axilla to the subclavicular area beneath pectoralis major and pectoralis minor.

Fig. 3. Combined treatment of LVA with sclerotherapy was planned and performed under general anesthesia. Indocyanine green lymphography revealed a linear pattern along the ulna side of the right upper limb. Two LVAs were established in peripheral sites extending from the cyst in the right upper arm.
The vein and lymphatic vessels were anastomosed in an end-to-end manner using 11-0 or 12-0 nylon suture under a microscope (Fig. 3).

The postoperative course was uneventful. The drainage volume decreased rapidly. The silicone drain was left in place until the daily drainage volume dropped below 50 mL. The drain was removed on postoperative day 4. Prophylactic antibiotics were administered for 1 week. Compression therapy was started immediately after surgery using a cotton bandage. A class 1 compression garment was applied after discharge. Compression therapy was stopped 6 months after surgery. One year later, the UEL index was 108 in the left upper extremity. The cystic lesion resolved and no recurrence was noted during a follow-up period of 18 months (Fig. 4).

DISCUSSION

Cystic lymphangioma is presumed to be congenital or a result of obstruction and retention of the lymph fluid in developing lymphatic vessels.8

Although sclerotherapy is an effective option and there have been no reports on the incidence of lymphedema following sclerosis of a cystic lymphangioma as far as we searched, sclerosants cause fibrosis and obliteration of both a lymphangioma and lymphatic vessel,9 thereby impairing lymphatic drainage. We believe that the ideal treatment in these cases is reconstruction to restore lymphatic flow by anastomosing lymphatic vessels flowing into the cyst to another lymphatic vessel or to a vein to stop the lymph flow within the cyst and maintain effective lymph drainage. LVA is thought to be an excellent option for the treatment of cystic lymphangioma when all the lymphatics flowing into the cyst are detectable. However, they are not always detectable, especially for large cystic lymphangioma. Consequently, we integrated the lymphatic treatment with sclerotherapy. The LVAs are established primarily for diverting lymph flow and preventing recurrence of the primary disease. Nevertheless, there are concerns that treatment of a cystic lymphatic malformation alone leads to worsening of lymphedema or increases the risk of its development. LVA, on the other hand, provides favorable results, not only in prevention but also in treatment of peripheral lymphedema.10 Recently, the focus of lymphedema treatment has shifted to risk reduction and prevention.10 Therefore, it is desirable to perform LVA combined with sclerotherapy in terms of preventing lymphedema and treating cystic lymphatic malformations. It is expected that combined treatment using LVA with sclerotherapy will offer a complementary minimally invasive treatment option for a large or persistent cystic lymphatic malformation. However, further observation, accumulation of cases, and combination of other sclerosants, such as OK-432, are necessary for the evaluation of this method.

CONCLUSION

Combined LVA with sclerotherapy allowed for a relatively less invasive method without further exacerbation or occurrence of lymphedema. Hence, combined treatment using LVA with sclerotherapy can be a complementary minimally invasive treatment option for a cystic lymphangioma.

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