Surgical Management of Vulvar Lymphangioma Circumscriptum: Two Case Reports

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Objective: To evaluate the efficacy of major labiaectomy as a surgical management of vulvar lymphangioma circumscriptum, we report two cases of this rare clinical entity.
Case Reports: Two female patients, aged 56 and 68 years, presented with persistent edema of the lower limbs, papule-like condyloma of the labia majora, and lymph oozing from these papules of the vulva, which had developed 24 and 10 years, respectively, after radical hysterectomy with adjuvant pelvic radiation therapy for cervical cancer. After major labiaectomy was performed, symptoms in the first case, of extensive resected skin margin, improved clearly, in the second case, vulvar lymphangioma circumscriptum was more severe than in the first case, and a small amount of lymph oozing occurred from residual papules of the labia majora. In both cases, histology revealed lymphangioma circumscriptum of the vulva.
Conclusion: Major labiaectomy is an effective therapy for vulvar lymphangioma circumscriptum. Particularly, in the case which was extensive and deep resected skin margin, symptoms such as papules of the labia majora and lymph oozing from these papules of the vulva associated with lymphangioma seemed to be clearly improved.

Key words: vulvar lymphangioma circumscriptum, acquired, major labiaectomy, cervical cancer, adjuvant radiation therapy

INTRODUCTION

The lymphangioma circumscriptum is a benign disorder of lymphatic origin that can be either congenital or acquired, involving the skin and subcutaneous tissues. Acquired vulvar lymphangioma circumscriptum is a rare disease and known to occur after radical hysterectomy with or without adjuvant radiation therapy for cervical cancer [1–3]. It is caused by damage to previously normal lymphatic channels, due to cancer treatment. Recently, 24 cases of acquired vulvar lymphangioma circumscriptum have been reported in the literature [4].

We report two cases of acquired vulvar lymphangioma circumscriptum with surgical management and present a review of the literature.

CASE REPORTS

Two women, 56 and 68 years old, were treated by the same surgeon. Surgery was performed under general anaesthesia in the lithotomy position. The operative technique consisted of extensive excision of the skin, and the deep layer extended to the level of the Colles' fascia without a skin flap. The wound was closed in two layers. After the surgery, a doctor changed the dressing daily, and antibiotic therapy was done prophylactically.

Case 1

A 56-year-old woman presented to our outpatient clinic with lower-limb lymphoedema, papules of a unilateral labium majus, oozing clear serous fluid in the area, with a tendency to become infected and feverish. The patient noticed a few papules at 24 years after abdominal radical hysterectomy, lymph node dissection, and adjuvant radiation therapy under the diagnosis of a stage 1b squamous cell carcinoma of the cervix.

These papules were associated with a verrucous change (Fig. 1). A histological specimen of the vagina showed a chronic inflammatory change with hypertrophy and multiple dilated thin-walled lymphatic capillaries lined with normal endothelium in the dermis. In the deep dermis, congestive change was also revealed. There was no evidence of recurrence of malignancy.

The patient underwent a unilateral major labiaectomy (Fig. 2–1, 2–2). The lateral margins were cut 1 cm away from the papules and extended down to the level of Colles' fascia. We removed all the papules by performing a unilateral major labiaectomy. After the labium was excised, the wound was closed without a skin flap. No relapse has occurred by the time of follow-up at 20 months after major labiaectomy (Fig. 3). The final pathological diagnosis was lymphangioma circumscriptum of the vulva (Fig. 4).

Case 2

A 68-year-old woman presented in our outpatient clinic with lower-limb lymphoedema, edema and papules of the bilateral labia majora, oozing clear serous fluid and pain in the area, a tendency to be infected.

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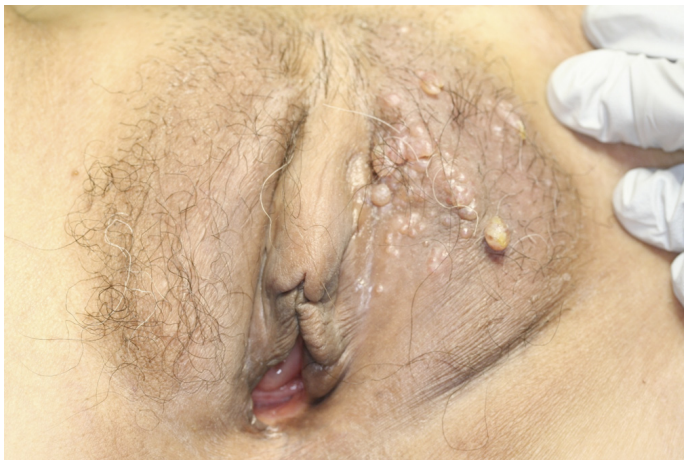

Fig. 1 Unilateral vulvar lymphangioma circumscriptum (case 1)

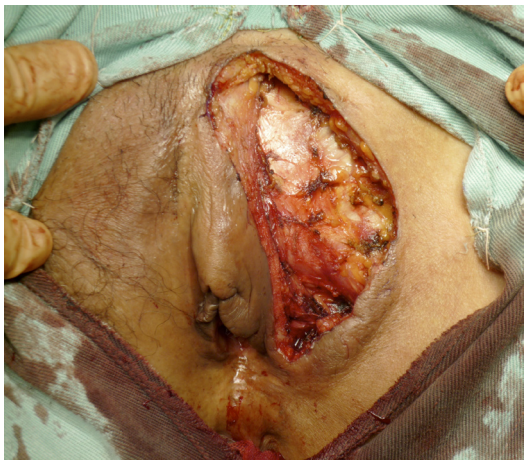

Fig. 2-1 Surgery of unilateral vulvar lymphangioma circumscriptum with major labiaectomy (case 1)

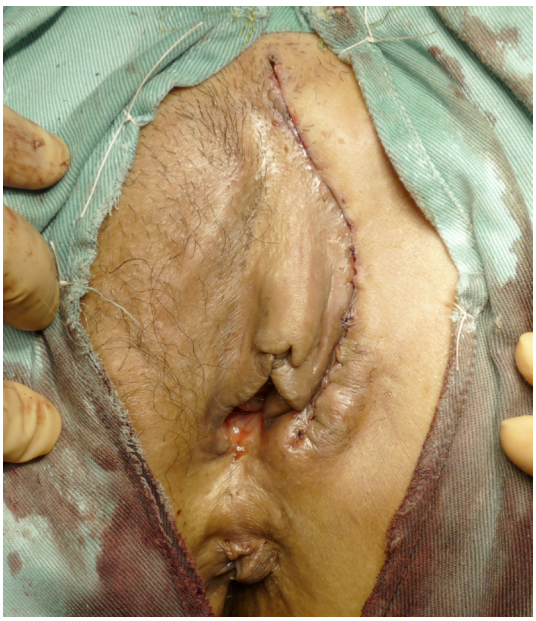

Fig. 2-2 Surgery of unilateral vulvar lymphangioma circumscriptum with major labiaectomy (case 1)

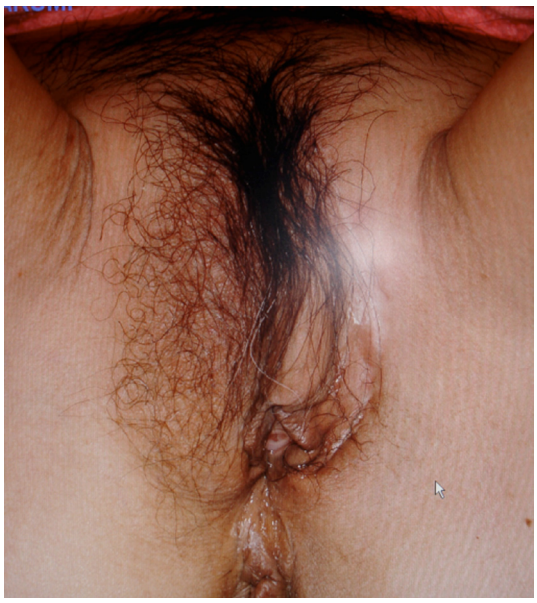

Fig. 3 A follow-up at 20 months after major labiaectomy, no recurrence (case 1)

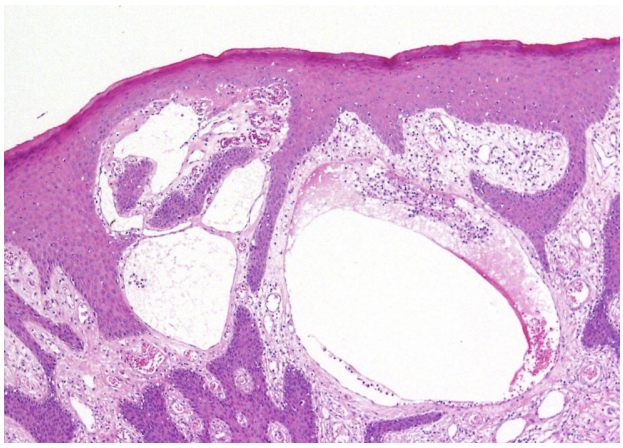

Fig. 4 Microphotograpy showing multiple dilated lymph vessels in the dermis (HE staining, ×20)

She noticed a few papules 10 years after abdominal radical hysterectomy, lymph node dissection, and adjuvant radiation therapy under the diagnosis of a stage 2b squamous cell carcinoma of the cervix.

This cluster of large and small papules in the bilateral labia majora histologically consist of hypertrophic epidermis and multiple dilated lymph vessels. Edematous collagen fibers and fat tissues were observed in the dermis. Immunohistochemically, D2-40, which is a recently available monoclonal antibody that has been used as a lymphatic endothelial marker was positive in endothelial cells. There was no evidence of malignancy.

During major labiaectomy, it was difficult to remove every papule because of more extensive lesions than in Case 1 (Fig. 5). Some residual papules were found (Fig. 6). After major labiaectomy was performed, symptoms have improved by the time of a follow-up at 7 months. However, a small amount of lymph oozing has occurred from residual papules of the vulva (Fig.

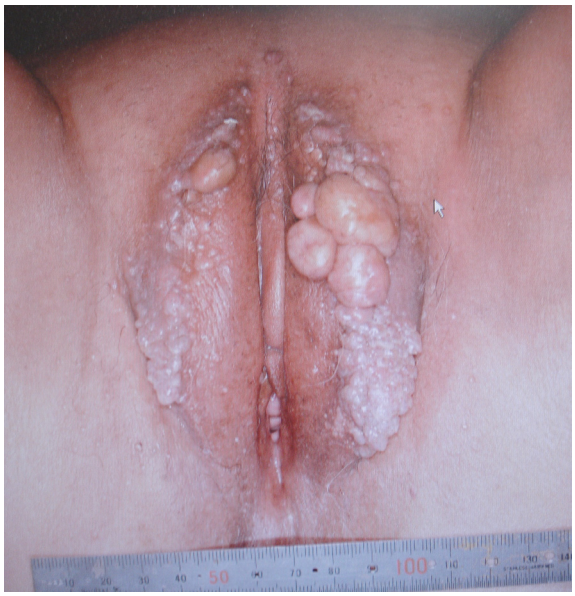

Fig. 5 Bilateral vulvar lymphangioma circumscriptum (case 2)

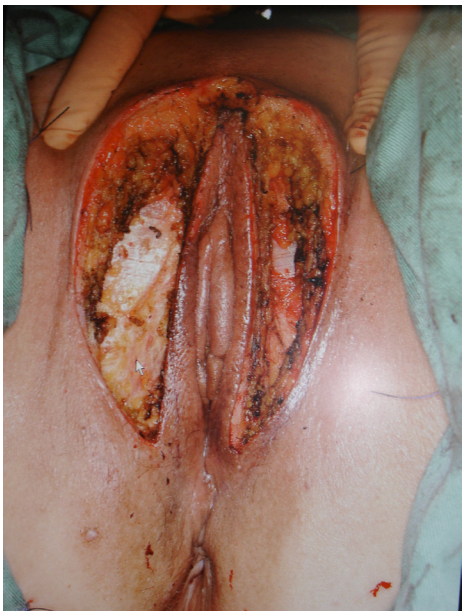

Fig. 6 Surgery of bilateral vulvar lymphangioma circumscriptum with major labiaectomy (case 2)

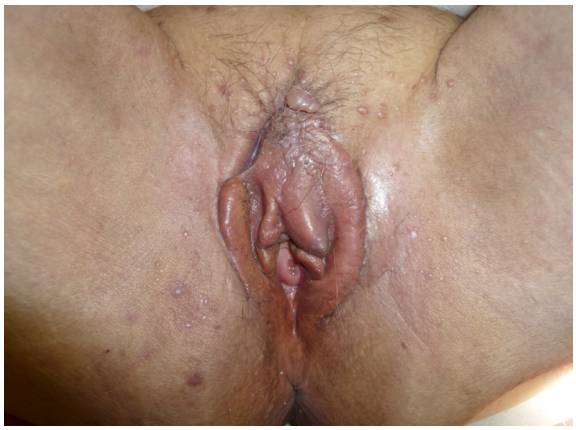

Fig. 7 A follow-up at 7 months after major labiaectomy, recurrence (case 2)

7). Histology revealed the diagnosis of lymphangioma circumscriptum of the vulva.

DISCUSSION

The vulvar lymphangioma circumscriptum is benign but causes frequent secondary infections with distressing symptoms, such as edema and papules of the labia majora and oozing lymph fluid in this area.

There is no standard management for vulvar lymphangioma circumscriptum. The various treatments range from conservative treatment with decongestive physiotherapy, such as manual lymph drainage, exercise, and compression, to abrasive therapy, sclerotherapy, electrocoagulation, laser-therapy with CO_2, and surgical excision depending on the patients' conditions [5, 6]. While lesion recurrence is frequent, out of various treatment modalities, surgical excisions were preferred in 19 of the 37 cases [7, 8]. The recurrence rate after surgical management was 23.1% on a follow-up ranging from 6–81 months. The recurrence rate might be twice as high in lymphangioma circumscriptum without surgical management [5]. Browse *et al.* reported that the recurrence rate after radical excision was high, when the initial lesions were greater than 7 cm diameter as compared with lesions less than 7 cm diameter for which a local excision was performed [9].

We propose that major labiaectomy is an effective and well-tolerated therapy for vulvar lymphangioma circumscriptum. Particularly, in Case 1, a unilateral skin margin extensively was resected, and all the papules were completely removed by performing a unilateral major labiaectomy. As a result, symptoms such as papules of the labia majora and lymph oozing from those papules of the vulva associated with lymphangioma seemed to be clearly improved.

To obtain a successful result in surgical treatment, it is important to completely excise all the papules with a sufficiently deep margin of subcutaneous tissue. In an extensive vulvar lymphangioma circumscriptum, it is impossible to remove the lesion without a skin flap. In fact, a small amount of lymph oozing has occurred from residual papules of the labia majora in Case 2. If radical surgical treatment was considered, a major labiaectomy with skin flap should have been taken into consideration.

Recently, laser therapy with CO_2 has been selected as an effective treatment for this disease. However, the large diameter and deep lesions as in Case 2 may not be treated by CO_2 laser therapy without keloid formation [3]. This treatment is good for small, superficial lesions. Accordingly, we tried it for residual papules in the Case 2. The combination of major labiaectomy without skin flap and laser therapy with CO_2 will be valuable for large diameter and deep lesions and improved quality of life for the patient, because major labiaectomy with skin flap gives considerable stress to the patient despite the benign disease.

We suggest that the indication of major labiaectomy is 1) the large diameter and deep lesions of vulvar lymphangioma circumscriptum, 2) severe symptoms such as more papules and vesicles, a serious lymph oozing, greater edema, pain, fever with infection, 3) therapeutic failure after conservative treatment or CO_2-laser therapy.

In conclusion, major labiaectomy seems to be effective for vulvar lymphangioma circumscriptum and beneficial for the patients' quality of life. Particularly, in extensive and deep vulvar lymphangioma circumscriptum with severe symptoms, major labiaectomy should be performed positively for better results.

Conflict of interest: None.

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