Unusual association of diseases/symptoms

Vulvar lymphangioma circumscriptum presenting periodic symptoms

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Summary

The authors report a case of acquired lymphangioma circumscriptum (LC), localised in the right labium majus of vulva and characterised by periodic symptoms for 8 years; however, 3 months after the right major labiectomy, the same symptoms developed in both the left labium majus and the right labium minus. To the best of our knowledge, this is the first case of acquired LC with this type of localisation and symptoms without any known aetiological factor.

BACKGROUND

Lymphangiomas are rare benign proliferations of the lymphatic system. There are three types of lymphangiomas: circumscriptum, cavernous and cystic. Lymphangioma circumscriptum (LC) is either primary (usually present at birth or develops in early childhood) or secondary (acquired). Secondary lymphangioma is also termed acquired lymphangioma.¹ Lymphangiomas of the vulva are rare. Thirteen cases of the congenital and 24 cases of the acquired form have been reported in the literature.² We report a case of acquired LC localised only in the right labium majus and characterised by periodic symptoms for 8 years; however, 3 months after the right major labiectomy, the same symptoms developed in the left labium majus and the right labium minus. To the best of our knowledge, this is the first case of acquired LC showing localisation and symptoms in this manner, without any known aetiological factor.

CASE PRESENTATION

A 46-year-old, virgin woman presented to our clinic with complaints of pain and swelling in the right labium majus of her vulva. According to the patient's history, she had pain and swelling in her right labium majus, increasing in intervals of 3–5 days and continuing for another 3–5 days before decreasing and disappearing completely. The symptoms had started 8 years ago. A right mastectomy had been performed 8 months earlier because of breast cancer and four cures of chemotherapy had been given. There was no history of genital ulcer, trauma, tuberculosis, pelvic infection or any digestive symptoms.

INVESTIGATIONS

On examination, there were multiple small raised bleb-like lesions on her right labium majus and lymphoedema limited to the right labium majus. The left labium majus was normal in appearance. Inguinal swelling, lymphadenopathy and varicose vein in the lower extremities were absent. Abdominal and pelvic ultrasound examination did not

show any pathological finding. The results of laboratory tests (routine biochemistry tests, complete blood count, chest x-ray) were within normal limits. The known causes of vulvar LC were ruled out.

A full-thickness dermal biopsy including the subcutaneous tissue was performed on the right labium majus. Histopathological examination revealed multiple dilated lymphatic vessels containing eosinophilic proteinous lymph fluid in superficial and papillary dermis, epidermis elevated on lymphovascular lesion and epidermal hyperkeratosis (figure 1). Clinical and histopathological findings were consistent with LC.

TREATMENT

The right major labiectomy with normal margins was performed. On postoperative control examination 4 weeks later, the patient did not have any symptoms. However, 3 months after labiectomy, the patient presented to our clinic with the same complaints in the left labium majus and the right labium minus. Consequently, simple vulvectomy was performed (figure 2). On pathological examination, LC was diagnosed in the left labium majus and the right labium minus.

OUTCOME AND FOLLOW-UP

Three months after the operation, she had no complaints.

DISCUSSION

LC is a defect of the lymphatics in deep dermis.³ The proposed aetiology of acquired LC is the architectural disruption of previously normal channels, leading to acquired LC by the sequestration and further dilatation of previously normal lymphatics. The predisposing conditions for acquired LC are radical surgery and/or radiation therapy for cervical neoplasia, infections (acute cellulitis, filariasis, sexually transmitted diseases, genital tuberculosis, erysipelas and lymphogranuloma venereum), Crohn's disease, bilateral varicose veins in the lower extremities and rhabdomyosarcoma. Acquired LC of the vulva is induced by

Figure 1 Dilated lymphatic channels containing eosinophilic proteinaceous lymph fluid in superficial and papillary dermis, epidermis elevated on lymphovascular lesion, and epidermal hyperkeratosis, $\times 100$ H&E.

impaired lymph flow.^{1–4} Of the 24 cases reported up to now, tuberculosis was found in 6, radiotherapy due to genital cancers in 8 and all the cases had one of the aetiological factors mentioned above. In the present case, however, aetiological factors were absent.

The most characteristic clinical presentation is of multiple small raised bleb-like lesions ('frog spawn-like') from which watery fluid may ooze, and there may be coexistent lymphoedema extending to the adjacent abdominal or thigh skin. Some cases are complicated by recurrent infections. Reactive epidermal changes occur frequently and in some cases these are sufficiently florid to mask the underlying vascular abnormality. Thus, acquired vulvar lymphangiectasia can mimic human papillomavirus-related condyloma, other viral or fungal infections, or specific dermatoses such as dermatitis herpetiformis.⁵

Vulvar LC can be asymptomatic, pruritic, burning or painful. Clinically, LC is characterised by translucent vesicles, which may be scattered or grouped like frog spawn. Rarely, the lesions can have a firm hyperkeratotic appearance.¹ Clinical and histopathological findings in the present case were consistent with those of LC (figure 1). It is very interesting that the symptoms of this patient were labial pain and periodic labial swelling, decreasing and increasing in intervals of 3–5 days. The fact that the symptoms in this case show a periodic course might well be explained as follows: Most probably, pressure increase, due to lymphatic accumulation in the lymphatic channels whose drainage was blocked, might cause swelling and pain in the vulva. When the pressure in the lymphatic channels reaches a critical level, anastomotic channels between the architecturally

disrupted channels and normal lymphatic channels might open and the lymphatic fluid might be drained. Thus, an asymptomatic period takes place. Complications associated with vulvar LC include cellulitis and psychosexual dysfunction. Follow-up of patients with lymphoedema is recommended because transformation to lymphangiosarcoma may occur in the oedematous limbs. Two cases of squamous cell carcinoma and lymphangiosarcoma arising within LC have been reported. In the differential diagnosis of vulvar LC, herpes zoster, molluscum contagiosum, genital warts, leiomyoma, angiomyofibroblastoma, angioyxoma, cellular angiofibroma and aggressive sarcomas should be considered.^{1–4}

No standard treatment of choice has been described for the management of LC. Depending upon the type and extent of LC, procedures such as abrasive therapy, sclerotherapy, electrocoagulation or surgical excision can be used as treatment options. The most preferred treatment for both the congenital and acquired types is surgical excision. In surgical excision, the deep-feeding lymphatic cisterns should be removed in order to prevent recurrence.⁴ In particular, major labiectomy seems to be more successful than methods such as lymphovenous anastomoses and lymph angioplasties, and laser therapy. A single operation may provide benefit for a long period; also, this approach has the potential to allow patients to rehabilitate to normal life and activity.^{2,6} We, too, at first, performed the right major labiectomy with normal margins in our patient. However, because of the appearance of the same symptoms in the left labium majus and the right labium minus 3 months after the first operation, we carried out simple vulvectomy (figure 2).

Figure 2 Appearance of vulvectomy specimen containing lymphoedema limited to right labium minus and left labium majus (RL: right labium minus, LL: left labium majus).

Learning points

- Four remarkable properties of LC in this case are: it localised only in the single labium majus of vulva for 8 years; symptoms occurred periodically; the same pathologic lesion, with the same symptoms, appeared in the left labium majus and in the right labium minus 3 months after the right labiectomy; and no known aetiological factors were identified.
- These characteristics not reported in literature so far have made this case worth reporting.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Amouri M**, Masmoudi A, Boudaya S, *et al*. Acquired lymphangioma circumscriptum of the vulva. *Dermatol Online J* 2007;**13**:10.
2. **Ghaemmaghami F**, Karimi Zarchi M, Mousavi A. Major labiaectomy as surgical management of vulvar lymphangioma circumscriptum: three cases and a review of the literature. *Arch Gynecol Obstet* 2008;**278**:57–60.
3. **Roy KK**, Agarwal R, Agarwal S, *et al*. Recurrent vulval congenital lymphangioma circumscriptum—a case report and literature review. *Int J Gynecol Cancer* 2006;**16**:930–4.
4. **Dikmen Y**, Cosan Terek M, Akman L, *et al*. Congenital vulvar lymphangioma circumscriptum. *Int J Clin Oncol* 2008;**13**:458–60.
5. **Stewart CJ**, Chan T, Platten M. Acquired lymphangiectasia ('lymphangioma circumscriptum') of the vulva: a report of eight cases. *Pathology* 2009;**41**:448–53.
6. **Ghaemmaghami F**, Karimi Zarchi M, Mousavi A. Surgical management of primary vulvar lymphangioma circumscriptum and postradiation: case series and review of literature. *J Minim Invasive Gynecol* 2008;**15**:205–8.

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