Surgical Treatment of an Extensive Lesion of Lymphangioma Circumscriptum of the Vulva

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Abstract : *A 52 years old postmenopausal female patient was admitted because of inability to have coitus due to a lesion in her vulva. It was found that despite various modes of therapy given to the patient who had had the same complaint for ten years, the lesion had enlarged. The couple also stressed that they had problems in their normal social life because of their impaired sexual life. The clinical appearance suggested the presence of a case of lymphangioma circumscriptum. This diagnosis was confirmed by biopsy studies. Total vulvectomy was performed with the removal of the deep subcutaneous fat tissue. No reccurence was observed over a follow up period of two years. The present study aims to demonstrate that extensive surgical treatment might be successful in cases in terms of preventing reccurence, when this lesion infrequently occurs in the vulva in an extensive fashion; and the patient gains the ability of having normal sexual life. (Thai J Obstet Gynaecol 1995;7:81-84.)*

Key words : Lymphangioma circumscriptum, vulva, surgical treatment.

Lymphangiomas are cases of malformation rather than true neoplasms. They are quite rare and are mostly of congenital nature occuring immediately after birth or in the first year of life[1,2]. These are three types, lymphangioma circumscriptum being the most infrequent one[1,2]. The term lymphangioma circumscriptum was first used by Morris in 1889[3]. The lesion manifests itself in the subepidermal vesicles. There are lymphatic cisterns deep set in the subcutaneous tissue. These cisterns are develop-

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mental anomalies and are not connected to the deep lymphatic system. However, they are connected to the dermal lymphatics along dilated lymphatic vessels in cases of lymphangioma circumscriptum. The vesicles forming under the skin are saccula dilatation of superficial lymphatics[1]. They are translucent, pale and 3-4 mm in diameter. When they burst open a mucoid fluid comes out. This fluid contains proteinaceous material and lymphatic fluid. Infection and increment in the size of the lesion are frequently seen. Lymphangioma circumscriptum has been reported to occur most frequently in the upper trunk, the neck, the axillae and the tongue[1,2]. Various modes of therapy have been performed in cases of lymphangioma. In local cases, cryotherapy and cauterization have proved effective[2]. In local and diffuse cases, radiotherapy has not only proven ineffective, but irradiation has given rise to some side effects. Surgical intervention has been attempted, yet recurrences are frequent[1,2]. The process has been found to recur at sites of primary suture and in the graft, in cases where the defect was grafted[3]. It was Whimster who first suggested that recurrence would be the rule, unless the deep cisterns were totally removed[4].

In 1977, Jordan et al observed a case of lymphangioma circumscriptum of the heep, that the lesion did not recur after total removal of the skin and the subadjacent subcutaneous fat tissue[3].

A case of vulvar lymphangioma circumscriptum, with such extensive spread as to prevent coitus, is unique in the literature. Thus, at the outset, the authors were unable to predict the extent to which they might be successul. Furthermore, occurence of the lesion at such advanced age was an additional feature of the case.

Case Report

A 52 years old postmenopausal female patient was admitted to our clinic complaining of a lesion in the vulva. She stated that due to this lesion, she was so uncomfortable, unable to wear underclothes and unable to have coitus. It was found that this complaint had been present for ten years. Previously when the lesion was small in a local state, it was cauterized and various therapies were given in an effort to prevent infection. Yet, it was found that the lesion had progressed and spreaded to the entire vulva. On clinical examination, (Fig.1) large groups of flat papillomas and infected pseudovesicles (3-5 mm in diameter) were observed in the vulvar skin. The skin was thickened and deformed. Biopsy was obtained following antibiotic therapy for infection. Pathological examination revealed that the epidermis had become thinner at some places with the papillae having become elongated and acanthosis in some areas. There were sections of enlarged lymphatic vessels immediately underneath

the epithelium. These were surrounded by the endothelium and contained proteinaceous fluid and lymphocytes in their lumina. A diagnosis of lymphangioma circumscriptum was established. Total vulvectomy was carried out consisting of the removal of deep subadjacent subcutaneous tissue. No postoperative complications occured. Pathological examination confirmed a clinical diagnosis of lymphangioma circumscriptum elephantiasis. The microscopic appearance of the lesion is shown in (Fig. 2).

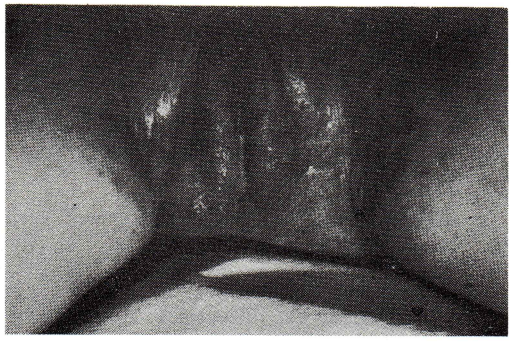

Fig. 1 Macroscopic appearence of the lesion.

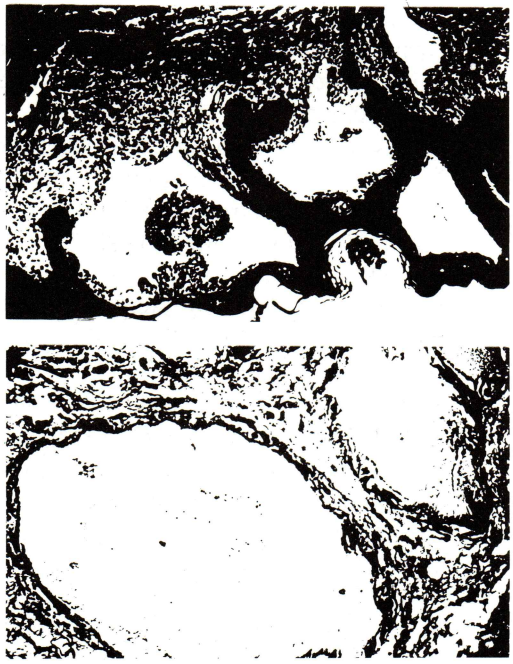

Fig. 2 Microscopic appearence of the lesion.

The patient was followed up for more than two years. No recurrence was seen. The patient felt extremely comfortable upon the elimination of the lesions. She and her husband stated that they had regained the ability to have coitus.

Discussion

Lymphangioma circumscriptum is a rarely encountered disease and is usually considered to be of congenital origin. It is seen at birth and within the first postnatal year. A very small number of cases have been reported in the adult age group[1,2]. In the present case, the quite advanced age of the patient and hindrance of coitus due to pain are striking features. Inability to have coitus constituted a great problem for the family.

Cases of lymphangioma circumscriptum of the vulva in the literature in which various therapy models were applied, resulting in the different sequences. In 1989, Abu Hamad et. al.[5] reported a case which partial vulvectomy was performed with no recurrence in the short follow-up period of six weeks. In 1991, Johnston et. al.[6] reported two cases of lymphangioma circumscriptum of the vulva in which extensive vulvar surgery were per-

formed and recurrences were observed in the follow-up period in both of them. Later in 1992, Mu-rugan et. al.[7] reported a case in which simple vulvectomy, was performed and recurrence was observed after nine months and the lesion was excised again. Our treatment was based on the previous studies byJordan[3] and Whimster[4] who described cases of lymphangioma circumscriptum occuring in other parts of the body. We performed total vulvectomy with the removal of deep subadjacent subcutaneous fat tissue containing the cisterns. Recurrence was not observed over a follow-up period of 2 years. There was no keloid formation. Restoration of the patients comfort and regaining of the possibility of comfortable coitus seem to be favorable results of the present study.

Among the cases of vulvar lymphangioma circumscriptum reported, two cases were described following radiotherapy for squamous cell carcinoma of the cervix[8,9], and another case reported by Sood et al.[10] had a history of surgery and treatment of pulmonary tuberculosis and scars near the left cervical and vaginal region. In the present study, there was no history of surgery or severe extragenital disease.

In conclusion, when simple vulvectomy is indicated in cases of vulvar lymphangioma circumscriptum, total removal of the subadjacent subcutaneous fat tissue, as far as the fascia, is recommended in order to prevent recurrence. We suggest that this therapy model should be the first choice in the treatment of vulvar lymphangioma circumscriptum.

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