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Lymphangioma circumscriptum of the vulva: a rare case report

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Summary

Lymphangioma circumscriptum (LC) is a rarely seen benign disease that involves deep dermal and subcutaneous lymphatic channels. It is seen as a group of lesions in the shape of thin-walled translucent saccules resembling frog eggs. Although the most frequently seen form of cutaneous lymphangioma is seen in the infantile period, it may occur at any age. The most common locations of these lesions are the proximal parts of extremities that contain a rich lymphatic network such as axilla, shoulder, inguinal region, and perineum. Vulvar involvement is quite rare and usually develops secondary to a radical hysterectomy, pelvic lymphadenectomy, radiotherapy, and Crohn's disease. Here, an extraordinary case of LS with an obscure cause is presented.

Key words: Lymphangioma; Lymphangioma circumscriptum; Vulvar lesion.

Introduction

Vulvar lymphangioma circumscriptum (LC) is a very rarely seen benign tumor. Lymphangioma is a hamartomatous formation composed of dilated lymph channels surrounded by lymphatic endothelium. The most frequently seen type is LC [1]. Histopathological evaluation reveals wide spaces containing coagulated lymph covered with a simple endothelial layer at the upper part of the dermis of the skin [2]. The mechanism for the development of this condition is considered to be vessel obstruction due to injury in the lymphatic vessels as a result of surgery, chronic inflammation, or radiotherapy [3]. Although the most frequently seen form of cutaneous lymphangioma is the congenital form, as seen in the infantile period of life, it can be seen at any age including menopausal period due to the pathogenic mechanisms stated above [4]. Here, an extraordinary case of LC in a young patient with an obscure etiology, is reported.

Case Report

A 23-year-old female patient presented with complaints of edema and wetness in her genital region. No history of prior gynecologic surgery, malignancy, lymphadenectomy, radiotherapy, and Crohn's disease, to explain the etiology of the condition, was present in the patient, who stated that a lesion and her complaints had been present for two years. Physical examination disclosed a general edematous condition of the vulva in addition to a lesion in a vesicular structure with a watery leak from multiple points measuring two- to four-mm in diameter (Figure 1). Routine laboratory tests on the patient were within the normal ranges. Histopathological examination of the incisional biopsy of the lesion revealed epidermal orthokeratosis and acanthosis in addition to an image of numerous dilated lymphatic channels. The diagnosis of LS was thus confirmed. The patient underwent bilateral labiectomy and total excision of the lesion (Figure 2). Histopathological evaluation of the whole specimen also confirmed the diagnosis, vulvar LC (Figure 3). The patient was discharged on the postoperative second day to be followed-up on an outpatient basis. No additional lesions were detected at the examination performed a month after the operation (Figure 4).

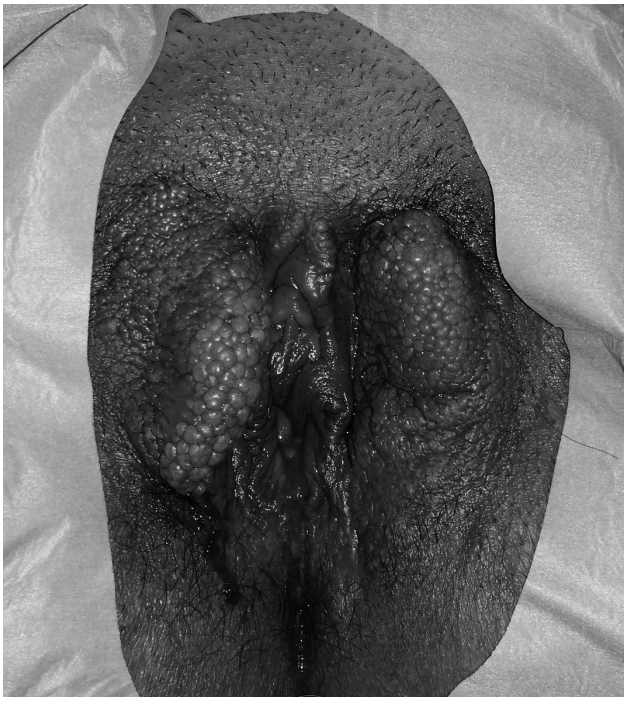

Figure 1. — Cluster of translucent thin-walled vesicles at the vulva with the appearance of frogspawn.

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Figure 2. — Appearance of the vulva after bilateral labiectomy and the specimen.

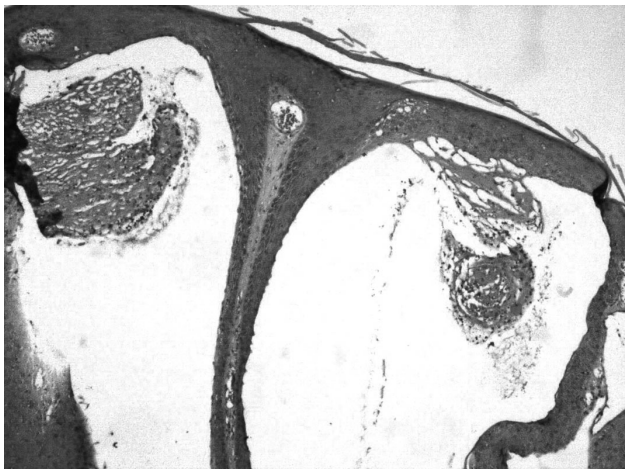

Figure 3. — Histopathology of the lesion under the microscope. Dilated lymphatic vessels are seen (H&E ×40).

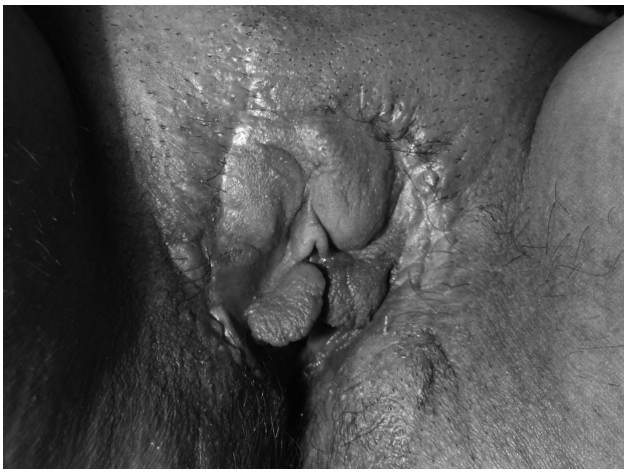

Figure 4. — Appearance of vulva one month after the operation.

Discussion

Lymphangioma is a benign tumor of the lymphatic system. LC, on the other hand, is the most frequently seen form of the lymphangiomas and is observed as dilated lymphatic vessels in the upper dermis and epidermis [5]. Typical appearance is in the form of papulovesicular translucent vesicles that may be pink, red, or black in color. LC has a close histopathological resemblance to cystic hygroma. Cystic hygroma is a congenital lymphatic malformation seen in the intrauterine or infantile period of life. It highly resembles LC histopathologically, in addition to a low number of lymphatic channels in the form of larger spaces [6]. The most frequently seen symptom is the painless growth of clusters of papillo-vesicular lesion in the vulva. The etiology of LC is unknown. Vulvar involvement is quite rare and develops secondary to various conditions such as radical hysterectomy, pelvic lymphadenectomy, radiotherapy, and Crohn's disease.

The most clear-cut treatment of LV is surgical resection. Variable treatment methods have been applied by various groups in the literature, one of which included carbon dioxide laser vaporization in wide lesions at localizations that are not amenable to surgical resection [7]. Surgical resection is easy in pedunculated types; however a wider surgical resection may be necessary in non-pedunculated lesions to completely excise the lesion anatomically. It may be necessary to excise the whole labium major after defining the margins of the lesion. In the case presented here, bilateral labiectomy was needed to excise the whole lesion. Treatment was accomplished with a complete resection without leaving any lesion at the margins of the resection.

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