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CASE REPORT

Acquired Vulvar Lymphangioma Following CO_2 Laser Treatment for Sebaceous Gland Nevus in a 10-Year-Old Girl: A Case Report

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Abstract: Acquired vulvar lymphangioma (AVL) is a rare condition characterized by abnormal lymphatic vessels in the vulva. We describe a 10-year-old female patient who presented with vulvar swelling for 23 days. She has no history of sexual assault and irradiation. Two months ago, the patient underwent CO_2 laser surgery for a sebaceous gland nevus on the left labia majora, resulting in delayed healing and residual scar tissue. Twenty-three days ago, swelling appeared in the external genitalia, accompanied by mild pain that worsened after activity and lessened when lying down. Treatment with anti-infection and anti-allergy medications at the previous hospital did not show significant improvement. Histopathological examination and genetic testing confirmed Our diagnosis of acquired vulvar lymphangioma with lymphedema.

Keywords: acquired vulvar lymphangiectasia, lymphangioma, lymphedema

Introduction

Acquired vulvar lymphangioma (AVL), also referred to as acquired lymphatic anomaly or lymphangiectasia, and previously known as lymphangioma circumscriptum, is a rare condition characterized by an abnormality in the lymphatic vessels of the vulva. AVL is associated with obstructed or impaired pelvic lymph drainage, which may result from chronic inflammatory or neoplastic conditions, as well as previous surgical or radiation therapy leading to lymphatic disruption. Various factors have been implicated as potential causes, including lymphadenectomy and radiotherapy for pelvic carcinoma. Additionally, AVL has been documented in inflammatory conditions such as Crohn's disease and infectious conditions like tuberculosis.[1,2] There are few reports in the literature regarding scars resulting from CO_2 laser treatment for vulvar masses that lead to AVL of the vulva, which is often misdiagnosed as vascular edema. In this report, we present a case of acquired vulvar lymphangiectasia with lymphedema following CO_2 laser treatment for sebaceous gland nevus.

Case Report

A 10-year-old girl presented with vulvar edema for 23 days, two months after undergoing laser surgery on the vulva. On history, the patient denied sexual assault or irradiation. Two months prior, the patient underwent CO_2 laser surgery for a sebaceous gland nevus on the left labia majora at another hospital without undergoing pathology, which based on clinical photos and symptom descriptions, resulting in delayed healing and the formation of residual scar tissue. The onset of swelling occurred twenty-three days ago in the external genitalia, accompanied by mild pain exacerbated by physical activity and alleviated by rest. No lymphadenopathy was detected upon palpation of the inguinal region. Diagnosed with angioedema previously, the patient showed suboptimal response to antihistamines and antimicrobial

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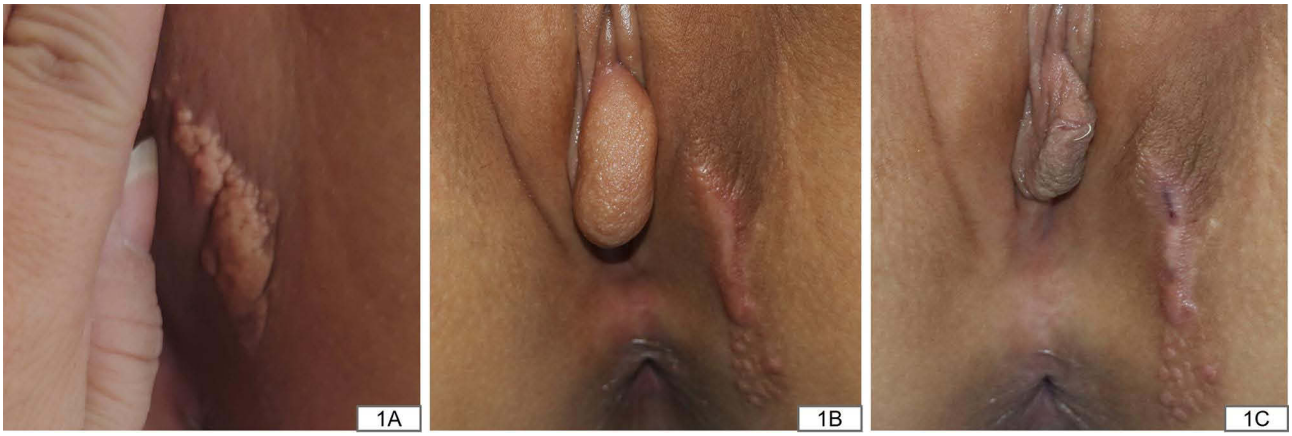

Figure 1 Preoperative CO_2 laser image shows a yellowish proliferative mass measuring approximately 0.5 × 3cm on the left labia majora, exhibiting a rough surface (**A**). Postoperatively, after over a month, a cystic mass measuring about 2 × 3 × 2cm in size is observed on the labia minora. This mass appears skin-colored, without surface ulceration or exudation, and is soft in consistency. The mass was soft in consistency with no palpable fluctuation, tenderness, or abnormal skin temperature. A longitudinal scar approximately 0.5 × 3cm in size is visible on the left labia majora (**B**). Significant reduction in swelling of the labia minora is noted upon waking (**C**).

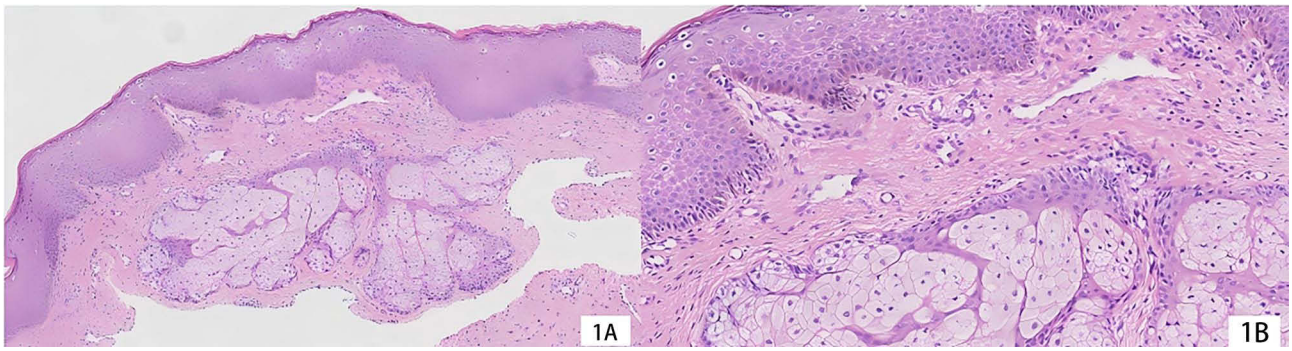

Figure 2 Mild epidermal hyperplasia with downward extension of the epidermis surrounding and proliferating lymphatic vessels. Scattered lymphocytic infiltration around the vessels, along with collagen proliferation. Dilated capillary veins in the upper dermis with elongation of the rete. (Hematoxylin &Eosin, 2A: ×100, 2B: ×200).

treatment. External genital examination (Figure 1) revealed a cystic mass measuring approximately 2x3x2 cm, skin-colored, with translucent or verrucous papules on the surface measuring 2 to 3 mm in diameter. The mass was soft in consistency with no palpable fluctuation, tenderness, or abnormal skin temperature. Furthermore, there was a longitudinally distributed pink, hard scar lump on the left labia majora, with a rough raised surface measuring approximately 0.5×3 cm in size. The edema of the vulva is prominent during activity or standing and alleviates upon rest or lying down.

During the refined pathology examination (4mm punch biopsy), milky white fluid was observed intraoperatively. Pathological findings revealed mild epidermal hyperplasia, downward extension of the epidermis surrounding and expanding lymphatic vessels, scattered lymphocyte infiltration surrounding them, and collagen proliferation (Figure 2). Genetic testing indicated abnormalities in the TIE1 gene, ERG gene, and CELSR1 gene. Based on symptoms, signs, and auxiliary examinations, the diagnosis is acquired lymphangiectasia of the external genitalia with lymphedema. Transfer to another hospital for treatment is recommended. After receiving wearing elastic pants and manual massage at an external facility, the patient's symptoms improved.

Discussion

Various classifications for lymphangioma have been described, such as superficial and deep cavernous lymphangioma, congenital and acquired, diffuse and discrete lymphangioma. Congenital lymphangioma results from a hamartomatous malformation of lymphatic vessels in the deep dermal and subcutaneous layers of the skin. Congenital lymphangiomas,

although present from birth, may go unnoticed for many years. Acquired lymphangioma, on the other hand, results from an acquired obstruction of lymph vessels following surgery or radiotherapy. They can manifest as asymptomatic, erythematous, flat, indurated, or atrophic plaques, or as a swelling.[3]

Symptoms of lymphangioma, particularly in the vulva, may include pain, itching (pruritus), discomfort, painful sexual intercourse (dyspareunia), and a burning sensation. In cases where the lymphangioma is more superficial, there may be drainage of fluid and a foul odor (malodor). These symptoms can vary in severity and may impact the quality of life of the individual affected by the condition. It is important for individuals experiencing these symptoms to seek medical evaluation and treatment.[3] The most definitive histological feature of AVL is the presence of dilated lymphatic spaces within the dermis. This characteristic finding enables pathologists to accurately diagnose AVL upon microscopic examination of tissue samples.[4]

Secondary lymphedema is a complex condition characterized by impaired lymphatic drainage function, often resulting from oncological surgeries, radiation therapy, vascular malformations, or surgical removal of local lymph nodes. The pelvic lymphatic system is rich, and the iliac lymphatic vessel group is composed of lateral chain, medial chain and intermediate chain, among which the external chain is from the inguinal lymph node, perineum, anal canal, lower vagina, uterine floor, scrotum, penis and clitoris. Therefore, once the posterior lymphatic tissue is damaged or blocked due to various factors, this can lead to dilatation or deformity of the anterior lymphatic vessels.[5] In such cases, disruptions to the lymphatic vasculature lead to the accumulation of fluid, solutes, and cellular debris in the tissues, causing progressive swelling, functional limitations, and physical discomfort for the patient.[6]

TIE1 is a cell surface protein expressed in endothelial cells. Involved in angiogenesis and lymphangiogenesis, including morphogenesis of lymphatic valves, essential for the functional integrity of the lymphatic system. Variants in TIE1 could contribute to the onset of lymphedema.[7] Maltese P E[8] suggests that variants in CELSR1 appear to be associated with autosomal dominant lymphedema with incomplete penetrance and variable expressivity. The expression of ERG is vital for maintaining the integrity of the endothelial barrier, which is essential for controlling the movement of molecules and cells between the bloodstream and surrounding tissues. It also plays a role in regulating endothelial cell permeability, affecting the passage of substances through the endothelial layer.[9]

In the case described, the patient does not have a history of tuberculosis or radiation therapy, but shows genetic abnormalities associated with lymphedema, indicating a potential inherent predisposition to this condition. Due to being an adopted child, the patient's familial history cannot be traced, and there have been no similar occurrences noted after birth. Two months prior to the presentation of symptoms, the patient underwent CO_2 laser surgery for sebaceous nevi, which resulted in keloid formation postoperatively. The subsequent vulvar swelling led to the diagnosis of secondary lymphatic dilatation with lymphedema following the CO_2 laser surgery for sebaceous nevi.

Two primary treatment modalities for acquired vulvar lymphangioma are excisional surgery and carbon dioxide laser treatment. Additional treatment options include cryotherapy, electrocoagulation, radiofrequency ablation and sclerosing agent injection. Excisional surgery aims to remove the abnormal subcutaneous lymphatic vessels and cisterns, improving the aesthetic appearance of the swollen vulva. Conservative treatments may provide symptomatic relief by managing weeping and swelling. Methods such as compression and exercise, typically used for limb lymphedema, are less appropriate for the genital area. Techniques like labial reduction or excision of edematous tissue have shown benefits in managing acquired vulvar lymphangioma.[10–13] The patient in this case opted for wearing elastic pants and manual massage at an external facility to minimize the impact on her quality of life.

Acquired vulvar lymphatic dilation is often caused by lymphatic obstruction following surgery or radiation therapy. This case highlights the complexity of lymphedema etiology, where a combination of genetic predisposition and environmental factors, such as CO_2 laser surgical interventions and postoperative complications, can contribute to the development of secondary lymphedema. Additionally, this condition is easily misdiagnosed as vascular edema, which warrants increased awareness among clinicians.

Ethics Statement

The patient gave written informed consent for publication of clinical information and photographs. No ethical committee approval was required because the data were analyzed in a retrospective manner.

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Disclosure

The authors declare no conflicts of interest in this work.

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