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Case report

Acquired vulvar lymphangioma circumscriptum after cervical cancer treatment: Case report

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ABSTRACT

Vulvar lymphangioma circumscriptum (LC) is a rare entity which may present as a painful, warty lesion. In contrast to the congenital form, which occurs in children, the acquired form arises in older adults and may be associated with infection, Crohn's disease, or prior pelvic/regional surgery. We present a case of acquired LC of the vulva in a 55-year-old woman who presented with a 3–4 year history of vulvar pain following chemotherapy, radiation, and brachytherapy for cervical cancer. Vulvar shave biopsies followed by excision revealed a thickened dermis with epidermal hyperkeratosis, parakeratosis, elongated rete ridges and dilated lymphatic channels containing eosinophilic material and scattered thrombi.

The differential diagnosis for this unusual lesion includes more common conditions such as condyloma acuminatum, fungating squamous cell carcinoma and molluscum contagiosum. It is important to recognize the clinical presentation as well as the distinct histological appearance of this rare benign entity.

1. Introduction

Lymphangioma circumscriptum (LC) is an uncommon benign condition that has been described in the chest, mouth, axilla, tongue, and rarely in the vulva (Sinha et al., 2015; Esquivias Gomez et al., 2001; Vlastos et al., 2003). LC usually presents as a warty appearing lesion which may or may not weep lymphatic fluid, and is often painful. The clinical differential diagnosis includes malignant tumor, condyloma acuminatum, molluscum contagiosum, and genital herpes (Sinha et al., 2015; Kokcu et al., 2010). The lesion has been classified as either 1) congenital which presents in childhood, usually before the age of 5 years, due to improper development of lymphatic channels with no connection to the main lymphatic drainage (Roy et al., 2006); or 2) acquired which presents in adulthood as a consequence of surgery, infections, or radiation resulting in a disruption of a previously normal lymphatic system (Sinha et al., 2015; Kokcu et al., 2010; Stewart et al., 2009; Vignes et al., 2010; Buckley and Barnes, 1996).

2. Case report

2.1. Clinical presentation

The patient is a 55-year-old female with a past medical history of cervical cancer status-post chemotherapy, radiation, and brachytherapy 16 years prior to presentation, who presented with a three to four-year history of vulvar pain. She was first evaluated at a dermatology clinic for fungating bilateral vulvar lesions, concerning clinically for condyloma acuminate or fungating squamous cell carcinoma where shave biopsies were performed.

2.2. Surgery and follow up

The patient was referred to a gynecology–oncology physician for further management. She underwent a wide local excision with pudendal flap reconstruction. Follow-up of the patient for 5 months revealed no local recurrence or post-operative complications.

2.3. Histopathology of vulvar biopsies

Microscopic examination of the vulvar biopsies revealed a thickened dermis, epidermal hyperkeratosis, parakeratosis and elongated rete ridges with no epithelial dysplastic features or increased mitotic activity. Dilated lymphatic channels containing eosinophilic proteinaceous material and scattered thrombi in the superficial dermis were seen. Patchy areas of chronic inflammation and vascular proliferation within

Abbreviations: LC, lymphangioma circumscriptum.

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the dermis were also noted. The histopathologic findings were characteristic of acquired vulvar LC.

2.4. Pathology of the vulvar excision

Grossly, the vulvectomy specimen measured 10 cm in maximum dimension. The lesion consisted of clusters of firm, tan-white, lobulated, botryoid nodules bilaterally in the labia majora that measured 7 cm in maximum dimension. There were several satellite nodules in the medial superior vulva up to 0.7 cm in greatest size. The surrounding skin was tan and hair bearing (Fig. 1). Serial sectioning revealed the nodules had white cut surfaces. Upon sectioning, some nodules were smooth, shiny, and homogeneous; while others were gritty and dull. The gritty nodules had central areas of darkening. Identical histomorphological features to those described in the vulvar biopsies were present in the excision (Fig. 2). The lesion was completely excised (negative surgical margins).

3. Discussion

Acquired LC is a rare entity which results from lymphatic damage; and can arise in the vulvar region after radiation therapy for cervical cancer, as seen in this case. Other causes include previous surgery, infection (tuberculosis, filariasis, recurrent cellulitis), and Crohn's disease (Kokcu et al., 2010; Stewart et al., 2009; Vignes et al., 2010). Architectural disruption causes dilatation and sequestration of previously normal lymphatic channels (Stewart et al., 2009). Common presenting symptoms include pain, oozing of lymphatic fluid, sexual dysfunction, and infection. Patients may also have periodic labial swelling, pruritus,

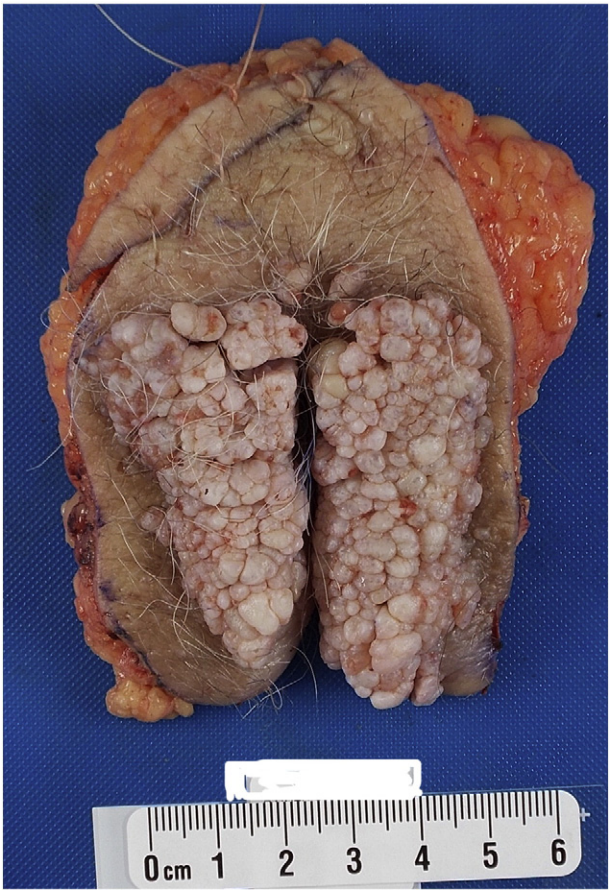

Fig. 1. Gross photograph of vulvar lymphangioma circumscriptum. Vulvectomy specimen showing extensive bilateral involvement of the labia majora by a lesion consists of clusters of tan-white, lobulated, botryoid nodules.

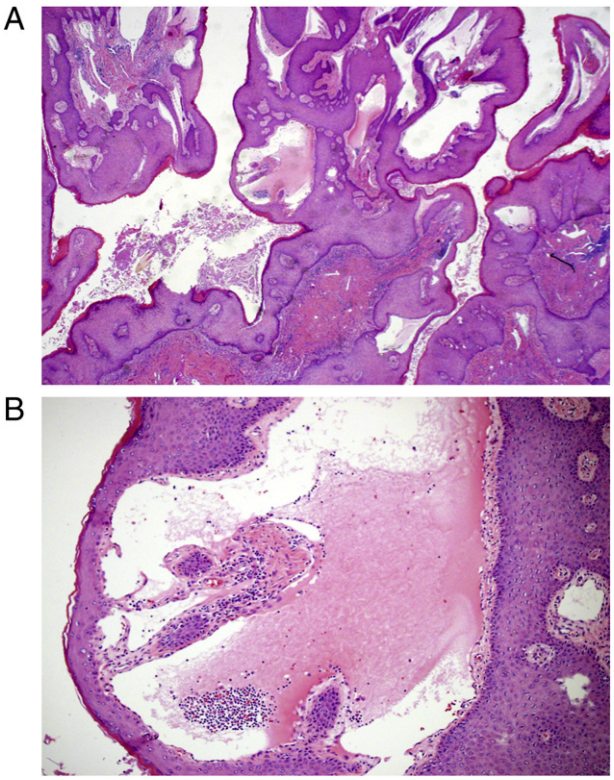

Fig. 2. Morphology of vulvar lymphangioma circumscriptum. Low power (A) and high power (B) captions showing villiform thickened dermis with epidermal hyperkeratosis, parakeratosis and elongated rete ridges with no dysplastic epithelial features. Dilated lymphatic channels containing proteinaceous material and scattered microthrombi in the superficial dermis are seen.

and recurrent cellulitis (Sinha et al., 2015; Kokcu et al., 2010; Vignes et al., 2010). Grossly, the lesions can be wart-like or papulovesicular; and may be confused clinically for fungating squamous cell carcinoma, molluscum contagiosum, condyloma acuminatum, or other viral/fungal infections(Sinha et al., 2015; Kokcu et al., 2010; Stewart et al., 2009; Mu et al., 1999).

Histologically, these lesions are characterized by superficial dilated lymphatic channels lined by flat endothelial cells with no atypical features characteristic of atypical vascular lesions. The lumens usually contain proteinaceous eosinophilic material. The overlying epidermis is usually acanthotic, hyperkeratotic, with irregular elongation of rete pegs that may be overlooked or underappreciated (Sinha et al., 2015; Stewart et al., 2009; Whimster, 1976). The endothelial cells of these dilated channels are usually positive for CD31 and D2-40 by immunohistochemistry. No human papilloma virus changes such as koilocytosis are present; and mitotic activity is confined to the basal layer of the epidermis. Additionally, no significant nuclear atypia is seen in these lesions. A mild to moderate inflammatory infiltrate may be seen (Stewart et al., 2009). The differential diagnosis includes non-specific vascular dilation, conventional lymphangioma, atypical vascular proliferations, differentiated type vulvar intraepithelial neoplasia, and bullous skin diseases (Patel et al., 2009).

The treatment of choice is complete excision of the affected lymphatic channels to avoid rapid recurrence. Other proposed therapies include vaporization with CO_2 laser, cryotherapy, and superficial radiotherapy (Sinha et al., 2015; Vignes et al., 2010; Lai et al., 2001; Huilgol et al., 2002; Sasaki et al., 2014). Due to the rarity of the vulvar cases, there is no reported recurrence rate after vulvectomy. However, the recurrence rate reported after surgery for all cases of cutaneous LC is 23.1% during follow-up periods ranging from 6 to 81 months. The recurrence rate is directly proportional to the original lesion size with

low recurrence rate of lesions measuring 7 cm or less compared to high recurrence rate for lesions greater than 7 cm (Browse et al., 1986).

In conclusion, we are presenting a rare benign lesion of the vulva, LC. It is important to recognize the clinical presentation as well as the distinct histological feature of this benign entity not to overlook it or to overcall it as atypical vascular proliferation or differentiated type vulvar intraepithelial neoplasia to assure proper treatment.

Disclosure

The authors of this paper have no relevant financial relationships with commercial interests to disclose.

Conflict of interests

The authors declare that there is no conflict of interests regarding the publication of this paper.

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