Gynecologic Oncology Reports 12 (2015) 7–8

Contents lists available at ScienceDirect

Gynecologic Oncology Reports

journal homepage: www.elsevier.com/locate/gore

Case Report

Primary mycosis fungoides of the vulva: The first reported case

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ARTICLE INFO

Article history:
Received 13 December 2014
Accepted 28 January 2015
Available online 7 February 2015

Keywords:
Mycosis fungoides
Vulva
Cutaneous T-cell lymphoma
Non-Hodgkin's lymphoma

Introduction

Cutaneous T-cell lymphomas (CTCLs) or B-cell lymphomas of the vulva are rare, aggressive, and often prone to either delayed or misdiagnosis. Other cutaneous diseases, such as lichen sclerosis, sexually transmitted diseases and dermatopathologies (Citarella et al., 2003) can mimic the presentation of these cutaneous lymphomas. Furthermore, most cases of lymphoma of the vulva are secondary manifestations of other primary disease processes (Vang et al., 2000). In the following case, we present a primary mycosis fungoides (MF) of the vulva in a biopsy of redundant vulvar tissue. To our knowledge this is the first reported case of primary MF of the vulva, and underscores the need to include primary lymphoma in the differential diagnosis when patients present with vulvar lesions.

Case report

A 50 year-old premenopausal gravida 0 presented to her gynecologist for evaluation of labial swelling and occasional pruritus. Initial evaluation of the labia demonstrated redundant labia and a random biopsy was collected. Labial biopsy was consistent with MF. Patient was then referred to gynecologic oncologist for further care. Initial exam showed labia to be redundant, five times normal size. The involved area included the right labia majora and portion of labia minora; it had a grayish white hue without ulceration or specific lesion. Her most recent Pap smear showed abnormal squamous cells of unknown significance (ASCUS); otherwise, her gynecological history was unremarkable. There were no palpable groin lymph nodes bilaterally.

Extensive initial diagnostics included CT scans of the chest, abdomen, and pelvis, which were negative. Bilateral bone marrow biopsy results revealed a mild-to-moderate decreased cellularity of the bone marrow without evidence of metastatic carcinoma. Megakaryocytes were adequate in number and no evidence existed of granuloma or peritrabecular lymphoid infiltrates in the bone marrow biopsy. A core biopsy specimen was 20–30% cellular and therefore inconclusive for the presence of iron. Peripheral blood was unremarkable and the B-cell leukemia/lymphoma panel was negative.

A thorough systematic examination of the patient's skin was unremarkable for ulcerations, scaling plaques, or microabscesses. The vulvar lesion was the only one observed. A right hemivulvectomy with bilateral groin lymph node dissection was performed. The redundant labium was excised in a deep-wise fashion to the perineal fascia.

The surgical specimen was sent for regular histology, flow cytometry, PCR and Southern Blot studies. Histological examination revealed an atypical lymphoid lesion made of predominantly small monotocous lymphocytes (Fig. 1) with round or convoluted nucleus and scanty clear cytoplasm. Those cells are arranged as dense interstitial, perivascular or lichenoid patterns (Fig. 2). The epidermis has psoriasiform hyperplasia. Solitary units of lymphocytes at the basal layer of the epidermis, the epidermis and adnexae with lymphocyte infiltrate associated with scant spongiosis (Fig. 3), papillary dermis stuffed with lymphocytes and scattered lymphocytes between wiry bundles of collagen in the superficial dermis (Fig. 1) are observed.

Flow cytometry study gated those small lymphocytes and revealed predominant T-cell (CD2, CD3, and CD5 positive) population that expresses CD 4 and negative for CD8. Immunohistochemical study on paraffin sections revealed again T-cell population (CD3, CD45RO, CD5, CD43 positive) that is negative for natural killer cell marker, CD56. There is minor lymphoid component in the lesion that is positive for CD20, B-cell marker. Study of the gene rearrangement of T-cell receptor (T-gamma chain) by PCR and Southern blot with JB1/EcorRI showed an evidence of monoclonality. The final diagnosis read as mycosis

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http://dx.doi.org/10.1016/j.gore.2015.01.006

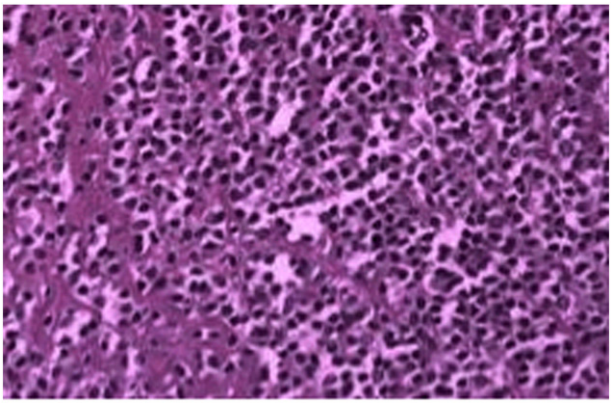

Fig. 1. Many small lymphocytes as sheet between wiry bundles of collagen.

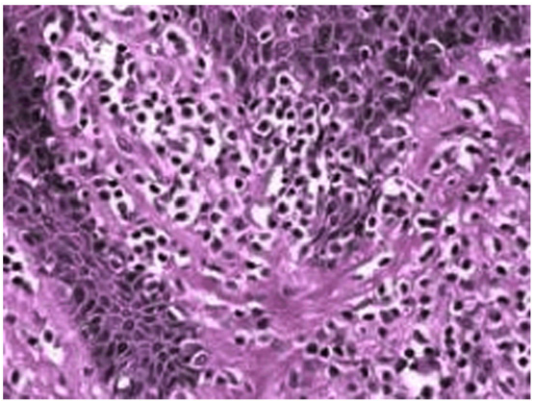

Fig. 3. Solitary units of small lymphocytes lined at the basal layer of the epidermis and epidemotrophism of lymphocytes with scant spongiosis present.

fungoides. The diagnosis was concurred by outside pathology consultation. Lymph nodes were negative for malignancy.

A consulting medical oncologist recommended that additional treatment was unnecessary. The first post-operation appointment was two weeks following surgery, followed by appointments every three months for the first two years, every six months for the following three years, and then annually thereafter. Annual CT scans of the chest, abdomen, and pelvis showed no evidence of any pathologic processes. Based on periodic patient complaints and physical examination findings, periodic biopsies and symptomatic treatment occurred without histological evidence of recurrent lymphoma. The last PET with CT scan, seventy-two months after surgery, was negative. Patient remained well with no evidence of recurrence 13 years after surgery.

Comment

Rare, case reports of lymphoma involving the vulva exist (Vang et al., 2000, 2001; Iczkowski et al., 2000; Marcos et al., 1992; Nam et al., 1992; Ferrando-Marco et al., 1992). MF is the most common lymphoid neoplasm involving the skin (Vang et al., 2001) however, the most common vulvar primary lymphoid neoplasm is diffuse large cell lymphoma. Vang et al. reviewed non-Hodgkin's lymphoma involving the gynecologic tract. Of the 21 cases of vulvar NHL reported over a 20-year period only 8 were localized to the vulva (Vang et al., 2001). Only one was identified histologically as MF. The previously seen vulvar MF was identified 2.5 years after initial diagnosis MF, thus this is likely a metastatic lesion and not the primary malignancy.

To our knowledge, this patient represents the first reported case of primary MF of the vulva to date. The vulva is widely thought to be a secondary site of lymphoma; however, a diagnostic evaluation in search of a primary site elsewhere in this patient revealed no other lesion. The case reported within has the histopathological characteristics of MF according to the WHO-EORTC criteria (Willemze et al., 2005), including the appearance of small-to medium-sized T lymphocytes aligned at dermo-epidermal junction, the overabundance of lymphocytes in the dermal papillae, bundles of collagen arranged haphazardly in zones of lymphocytic infiltrate. In other parts of the body, MF has an indolent clinical course with slow progression and skin-targeted chemo- or radiation-therapies are indicated for MF lesions restricted to the skin. However, there is reportedly low efficacy of radiation therapy to the vulva, therefore surgical removal of the lesion is the sole treatment.

Unfortunately, lymphoma of the vulva has an exceptionally high mortality rate, likely because it clinically mimics benign disease process and usually has a delayed diagnosis. An accurate diagnosis and early intervention likely contributed to the favorable outcomes for this patient. Therefore, all suspicious lesions of the vulva should be aggressively investigated to reduce morbidity and mortality.

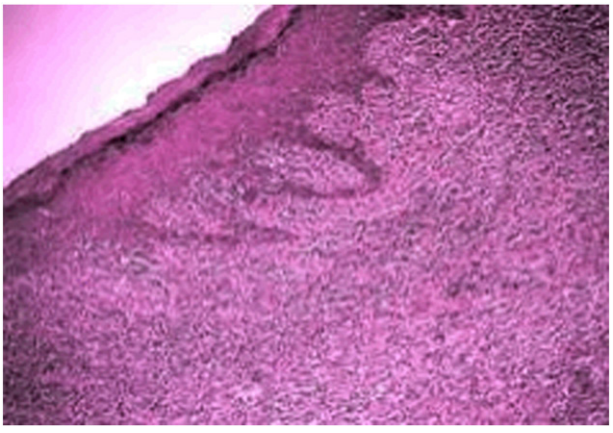

Fig. 2. Dermis is infiltrated by dense lymphoid cells as interstitial, lichenoid and perivascular pattern. The overlying epidermis shows psoriasiform hyperplasia.

Disclosure

No financial disclosures.

Conflict of interest

The authors have no conflicts of interest to disclose.

Acknowledgments

The authors would like to thank Christine M Dugan and Carl Reisdorff.

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