

CLINICAL VIGNETTE

Cutaneous Follicle Center Lymphoma in a Pregnant Patient

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Case Presentation

A 41 year old woman who was three months pregnant with her first child was referred by her primary care physician to Dermatology for evaluation of an itchy bump on her scalp. She stated that it had been present for about three months. On exam, she had a 3 cm erythematous non-scaly plaque with telangiectasias and loss of follicular ostia on the vertex scalp.



A 4 mm punch biopsy was performed for diagnosis, with a clinical differential of discoid lupus vs another inflammatory skin condition causing scarring alopecia. Histology showed an atypical lymphoid proliferation with features consistent with cutaneous follicle center lymphoma. Immunostains revealed abnormal cells with follicular pattern positive for CD20, BCL2, and CD23. She was referred to Hematology-Oncology and Surgical Oncology for further evaluation and treatment. Labs included a complete blood count with differential, comprehensive chemistry panel, lactate dehydrogenase, and peripheral blood flow cytometry, which were all unremarkable.

Staging computed tomography (CT) scan was recommended after delivery. Treatment options including surgical excision, radiation, and rituximab were discussed. Given the localized involvement of her lymphoma, surgical excision was recommended. One month later, she had a wide local excision performed but had positive margins on histology. Patient opted to wait until delivery of her baby for further treatment. Following the uncomplicated delivery of a healthy baby girl, the patient had re-excision of the lymphoma on her scalp with a focus of residual lymphoma and negative margins on histology. Staging CT was also negative. One year later, the patient continues to do well without recurrence.

Discussion

Cutaneous follicle center lymphoma is a primary cutaneous B-cell lymphoma. By definition, primary cutaneous lymphomas involve only skin at the time of diagnosis and are divided into cutaneous B-cell lymphoma (CBCL) and cutaneous T-cell lymphoma. CBCLs make up about 25% of all primary cutaneous lymphomas. Based on the 2008 World Health Organization - European Organization for Research and Treatment of Cancer joint classification, there are three types of primary CBCL: primary cutaneous follicle center lymphoma (PCFCL), primary cutaneous diffuse large B-cell lymphoma, leg type, and primary cutaneous marginal zone lymphoma. PCFCL is the most common type of primary CBCL. The median age at diagnosis is 51 years with a slight male predominance. Lesions typically present as firm erythematous to violaceous nodules or plaques with a smooth surface. The majority present as solitary or localized skin lesions on the scalp, head and neck, or trunk.^{1,2} Rarely, it may present with more diffuse inconspicuous lesions or as a scarring alopecia, making diagnosis difficult. The differential diagnosis in these cases may include inflammatory skin conditions, as in the patient in this case, and may only be diagnosed as lymphoma after biopsy.³

Diagnosis requires a punch, incisional, or excisional biopsy for histologic evaluation. Once diagnosis is confirmed, staging should be performed to evaluate for extracutaneous disease and help guide treatment decisions. Staging includes a complete blood count with differential, comprehensive chemistry panel,

lactate dehydrogenase level, and peripheral blood flow cytometry. CT or PET CT of the chest, abdomen, and pelvis should also be performed at diagnosis.¹

There are no randomized controlled trials available for treatment of PCFCL, and data supporting treatment is limited in the literature to small retrospective studies or case series. Optimal patient management requires a multidisciplinary approach, including dermatology, medical oncology, surgical oncology, and radiation oncology. Typically, patients with solitary or localized lesions, such as the patient in the case, are treated with localized low dose radiation therapy or surgical excision. Patients with generalized involvement, refractory lesions, or relapse may be treated with rituximab. Rituximab is a monoclonal antibody which binds to CD20 expressed on B lymphocytes. Patients with extracutaneous disease may additionally be treated with chemotherapy.^{1, 2, 4, 5}

PCFCL has an excellent prognosis with a 95% 5-year survival. Some patients may even experience spontaneous regression. Although it has a 30% rate of cutaneous relapse, only about 10% of patients have extracutaneous dissemination.¹

Lymphoma rarely occurs during pregnancy, and its incidence is thought to be fewer than 1 in 6,000 deliveries.⁶ PCFCL is also a rare lymphoma, and due to its sometimes nonspecific presentation it may mimic inflammatory skin conditions. Primary care physicians should be aware of this entity and promptly refer patients to dermatology if they have non-specific skin findings or presumed inflammatory conditions not responding to conventional treatment, as biopsy may be needed for appropriate diagnosis and treatment.

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