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JAMA Oncology Clinical Challenge

A Lesion on the Scalp

Zaid Abdel-Rahman, MD; Kedar Inamdar, MD; Haythem Ali, MD



Figure 1. A, Clinical image of the patient at presentation. B, Biopsy specimen from the skin lesion is shown.

A 67-year-old man presented with a rapidly enlarging scalp lesion (Figure 1A) first noticed 3 months earlier as a pimple. The lesion was itchy but without generalized pruritus. The patient reported a weight loss of 10 pounds over the preceding 3 months. In addition, he reported a longer history of vague, nonspecific, intermittent abdominal pain without nausea, vomiting, or fever starting about 3 years prior to presentation. Previous work-up had revealed minimal abdominal adenopathy, but the patient declined further workup. On examination, he had no other cutaneous lesions, but he did have cervical lymphadenopathy. All other physical findings were within normal limits. Laboratory findings were unremarkable, save for a mild normochromic normocytic anemia and a mildly elevated levels of lactate dehydrogenase. A biopsy specimen from the skin lesion is shown in Figure 1B.

WHAT IS YOUR DIAGNOSIS?

- A. Mycosis fungoides
- B. Follicular lymphoma
- C. Lichen planus
- D. Marginal zone lymphoma

Diagnosis

B. Follicular lymphoma

Clinical Course

Scalp lesion, lymph node, and bone marrow biopsy demonstrated involvement by CD10-positive, low-grade B-cell non-Hodgkin lymphoma (NHL), consistent with follicular lymphoma. A positron emission tomography (PET) scan revealed extensive hypermetabolic activity in the scalp soft tissue (standardized uptake value [SUV], 7.0) (Figure 2) and in the cervical (SUV, 9.1), axillary, upper thoracic, and abdominal lymph nodes (SUV, 3.9) compatible with lymphoma with a FLIPI score (Follicular Lymphoma International Prognostic Index) of 5. The patient underwent 6 cycles of R-CHOP chemotherapy (rituximab plus cyclophosphamide, doxorubicin, vincristine, and prednisone) and achieved a complete response verified by PET scan after cycle 4.

Discussion

Follicular lymphoma is one of the most common subtypes of NHL and is the most common of the clinically indolent NHLs. In the United States, follicular lymphoma accounts for approximately 35% of NHLs, the majority of which present with diffuse lymphadenopathy. In the present case, the patient had clear evidence of systemic disease, but the skin involvement was the most prominent presenting feature. Skin involvement is uncommon in follicular lymphoma, reported in fewer than 5% of the patients.

Histopathologic examination of the skin lesion exhibited a predominant follicular pattern of small B cells that tested positive for CD20, CD10, Bcl2, and Bcl6 and negative for CD3, CD5, TdT, cyclin D1, and CD21, consistent with follicular lymphoma. The proportion of centroblasts was fewer than 15 per high-power field, and there were no morphologic areas of transformation to large B-cell lymphoma. However, the lymphoma exhibited some

atypical features, demonstrating lymphoid cells with blastoid cytomorphologic characteristics rather than classic centrocytes. In addition, there were focal areas of increased mitotic activity. While overall Ki-67 nuclear proliferation rate was not substantially elevated, in some foci it was as high as 40%. Follicular lymphoma with blastoid features, or a "blastoid variant" of follicular lymphoma, is not a defined category owing to a lack of a reproducible definition of blastoid features in the literature.¹⁻⁴

At first glance the scalp lesion in the present case is suggestive of cutaneous T-cell lymphoma, especially mycosis fungoides. However, the predominant presence of monoclonal B cells in the scalp biopsy (Figure 1B), the lack of epidermotropism and Pautrier microabscesses, and the absence of immunophenotypic evidence of a T-cell neoplasm argue against this diagnosis.

The location and size of the lesion would also be atypical for lichen planus, which, in addition, is an unlikely cause of diffuse lymphadenopathy. On histopathologic analysis, lichen planus usually demonstrates a bandlike infiltrate at the dermal-epidermal interface with extension of lymphocytes into basal and parabasal keratinocytes. The epidermis may demonstrate prominent acanthosis and hyperkeratosis.

Marginal zone lymphoma B cells do not exhibit a germinal center profile, and they test negative for CD10 and Bcl6. They also characteristically exhibit small lymphoid cells with abundant pale cytoplasm and irregular nuclei with mature chromatin, thus show-

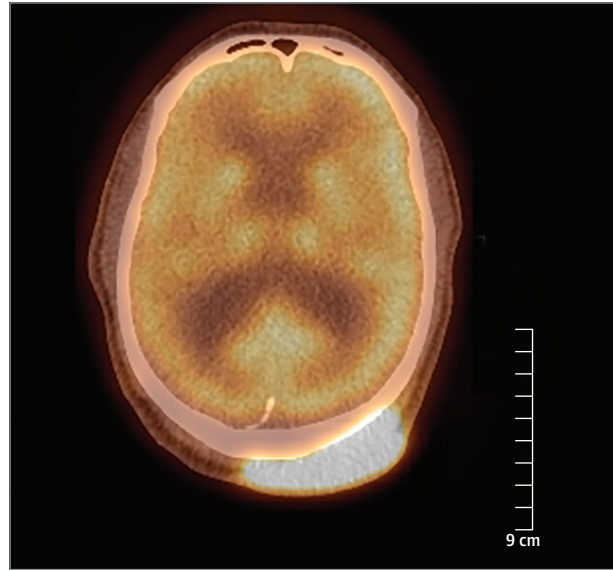


Figure 2. Positron emission tomography reveals extensive hypermetabolic activity in the scalp soft tissue.

ing "monocytoid appearance" of the lymphoid cells. In addition to this, the presence of centrocytes and centroblasts in the present case argue against a diagnosis of marginal zone B-cell lymphoma.

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