Review of contemporaneous mycosis fungoides and B-cell malignancy at Mayo Clinic

Elise Herro, MD,[e] David J. DiCaudo, MD,[a,b] Mark D. P. Davis, MD,[d] Amy L. Weaver, MS,[c] and David L. Swanson, MD[a]
Scottsdale, Arizona, and Rochester, Minnesota

Background: Having either mycosis fungoides or B-cell lymphoma may predispose a patient to the other.

Objective: We sought to determine whether the contemporaneous occurrence of the two malignancies is greater than chance and to investigate possible risk factors for the second malignancy.

Methods: We retrospectively reviewed the records of patients with contemporaneous mycosis fungoides and B-cell lymphoma seen between 1990 and 2007 at Mayo Clinic, Rochester, MN, or at Mayo Clinic, Scottsdale, AZ.

Results: In all, 23 patients had contemporaneous mycosis fungoides and B-cell malignancy. The first diagnosis was mycosis fungoides in 10 patients and B-cell lymphoma in 7; in 6 patients, the diseases were diagnosed simultaneously. No therapeutic factors could account for a predisposition to a second malignancy.

Limitations: Retrospective design, referral center, and small sample size are limitations.

Conclusion: Mycosis fungoides and B-cell lymphoma are unlikely to occur contemporaneously by chance, but no factor obviously predisposes a patient with one malignancy to development of the second (J Am Acad Dermatol 2009;61:271-5.)

Key words: B-cell lymphoma; cutaneous T-cell lymphoma; mycosis fungoides; Sézary syndrome.

Mycosis fungoides, a T-cell lymphoma, is the most common cutaneous lymphoproliferative disorder. Some authors have speculated that patients with mycosis fungoides are at increased risk for development of a secondary malignancy, including another non-Hodgkin lymphoma.[1-3] This speculation is based primarily on anecdotal reports or small case series. We reviewed our experience with patients who had received diagnoses of contemporaneous mycosis fungoides and B-cell malignancy to determine whether the dual occurrence of these malignancies was by chance. In addition, we attempted to identify cause and effect relationships between the two malignancies or their therapies. For example, we attempted to identify whether specific treatment of one malignancy predisposed a patient to the development of the second.

METHODS

After obtaining approval for our study from the Mayo Clinic Institutional Review Board, we identified records of patients who had received a diagnosis of mycosis fungoides, B-cell malignancy, or both at Mayo Clinic, Rochester, MN, or at Mayo Clinic, Scottsdale, AZ. We conducted a computerized search of diagnoses in the medical records that dated from 1990 to 2007. Our search terms related to mycosis fungoides included "mycosis fungoides," "cutaneous T-cell lymphoma," "Sézary syndrome," and "parapsoriasis." Other T-cell lymphoproliferative disorders, such as peripheral T-cell lymphoma, CD30+ anaplastic large-cell lymphoma, and subcutaneous panniculitis-like T-cell lymphoma, were excluded. For B-cell malignancy, our search terms

From the Department of Dermatology[a] and Division of Anatomic Pathology,[b] Mayo Clinic, Scottsdale; Division of Biostatistics[c] and Department of Dermatology,[d] Mayo Clinic, Rochester; and College of Medicine, Mayo Clinic, Scottsdale (visiting medical student).[e]
Funding sources: None.
Conflicts of interest: None declared.
Accepted for publication March 23, 2009.
Reprint requests: David L. Swanson, MD, Department of Dermatology, Mayo Clinic, 13400 E Shea Blvd, Scottsdale, AZ 85259. E-mail: swanson.david@mayo.edu.
Published online May 29, 2009.
0190-9622/$36.00

doi:10.1016/j.jaad.2009.03.033

were "lymphocytic leukemia," "lymphoma," "multiple myeloma," "monoclonal gammopathy of undetermined significance," and "amyloidosis." For the purposes of data analysis, we considered chronic lymphocytic leukemia and small lymphocytic lymphoma as a single entity, on the basis of the 2001 World Heath Organization grouping.[4] A dermatopathologist and the other members of our research team verified diagnoses by reviewing the written clinical notes and pathologic diagnoses in the medical record. B-cell diagnoses were consolidated into 3 groups: high-grade B-cell lymphomas, low-grade B-cell lymphomas, and proliferative disorders with plasmacellular infiltrates. In our review, we noted year of diagnosis, method of diagnosis, type of treatment, outcome, and order of malignancy (which malignancy preceded the other).

CAPSULE SUMMARY

- The contemporaneous occurrence of mycosis fungoides and B-cell malignancy should be rare, but one disease may predispose a patient to the other.
- Between 1990 and 2007, 23 patients seen at Mayo Clinic, Rochester, MN, and Mayo Clinic, Scottsdale, AZ, had contemporaneous mycosis fungoides and B-cell malignancy—an incidence greater than that expected by chance.
- No therapeutic factors could account for a predisposition to a second malignancy.

Because Mayo Clinic is a tertiary referral center, the patient records represented not only residents of the local regions (Olmsted County, Minnesota, and Maricopa County, Arizona) but also these two centers' referral populations. Mayo Clinic's referral population is primarily from North America, although some international patients are included among the referred patients.

RESULTS

During the 18-year period studied, we identified 7395 patients who had received a diagnosis of B-cell malignancy and 533 who had received a diagnosis of T-cell malignancy. Of those patients, 23 had contemporaneous mycosis fungoides and B-cell malignancy. The specific B-cell and T-cell neoplasm diagnoses and the consolidated diagnoses for these 23 patients are listed in Table I. All 23 patients resided in the United States, 21 of whom were from the Midwest or Arizona.

Of those who received diagnoses of both malignancies, 10 were women and 13 were men. The median age at diagnosis of the first malignancy was 69 years (range, 28-84 years). In 10 of the patients, the first diagnosis was T-cell malignancy (mycosis fungoides or Sézary syndrome) and in 7 patients the first diagnosis was B-cell lymphoma (Table II). In 6 patients a simultaneous diagnosis of mycosis fungoides and B-cell lymphoma was made at the time of initial consultation with a physician. Among the patients who received an initial diagnosis of mycosis fungoides only, the median age was 62 years (range, 28-84 years) and the median time to second malignancy diagnosis was 4 years (range, 1-48 years). Among the patients who received an initial diagnosis of B-cell lymphoma, the median age was 69 years (range, 41-77 years) and the median time to diagnosis of the second malignancy was 3 years (range, 1-5 years).

Among the 7 patients who received a diagnosis of B-cell malignancy first, 3 received treatment with alkylating agents and 4 did not (Table III). Among the 10 patients who received a diagnosis of mycosis fungoides first, 4 received treatment with psoralen plus ultraviolet A (PUVA) and 6 did not. The numbers of patients were too small to allow statistical power for comparisons.

Among the 7 patients who received a diagnosis of B-cell malignancy first, 4 patients had follow-up beyond 5 years, one of whom died at 7 years. Among the 10 patients who received a diagnosis of T-cell lymphoma first, 9 patients had follow-up beyond 5 years, 3 of whom died: two patients at 6 years and one at 15 years. No patients were lost to follow-up.

DISCUSSION

One aim of this investigation was to determine whether substantially more patients at our institution had diagnoses of both mycosis fungoides and B-cell malignancy than would be expected by chance. The overall age-adjusted incidence of mycosis fungoides in the United States is 6.4/million persons/year.[5] In addition, the US age-adjusted incidence of B-cell malignancy is 26.13/100,000 persons/year.[6] If contemporaneous mycosis fungoides and B-cell malignancy occurred by chance, the incidence of both would be 1.67/billion persons/year. Therefore, it would be improbable for a large enough patient series to have been accumulated at a single institution.

According to the US Census Bureau, as of 2008 more than 304 million people live in the United States.[7] Mayo Clinic, Scottsdale, AZ, and Mayo Clinic, Rochester, MN, combined, have 260,000 individual patient visits/year. If both malignancies occurred by chance alone, we would expect a combined annual