

Sezary Syndrome: A Case Study of Cutaneous T-Cell Lymphoma

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Cutaneous T-cell lymphomas (CTCLs) are a diverse group of neoplasms that affect the skin. Mycosis fungoides and Sezary syndrome are the most common forms of CTCL, with Sezary syndrome being the more aggressive type (Leukemia and Lymphoma Society, 2008). A total of 4,783 new cases of CTCL were identified from 1973–2002; 72% were mycosis fungoides and 2.5% were Sezary syndrome (Criscione & Weinstock, 2007). The incidence of CTCL is increasing by 2.9 cases per million people per decade; the increase could be a result of changes in classification or improvements in diagnosis. CTCL incidence is about 50% greater in African Americans than Caucasians, and men are affected twice as often as women (see Figure 1). Unlike other forms of CTCL, the incidence of Sezary syndrome is higher among Caucasians than African Americans (Criscione & Weinstock). CTCL incidence increases with age, with the greatest increase occurring at age 70 or older (Criscione & Weinstock) (see Figure 2). This column will use a case study of a patient with Sezary syndrome to focus on the background of the syndrome, signs and symptoms, and new approaches to treatment.

Signs and Symptoms

The initial onset of CTCL often is difficult to diagnose because of its similarity to many other nonmalignant skin pathologies (Trautinger et al., 2006). Sezary syndrome is characterized by an extensive red pruritic rash and, sometimes, sloughing of the skin (Leukemia and Lymphoma Society, 2008). Palms and soles often are thickened, scaly, and fissured. In addition, patients with

advanced disease may develop alopecia, nail dystrophy, and eye changes (Hwang, Janik, Jaffe, & Wilson, 2008). Sezary syndrome also has been characterized by the presence of 5% or more malignant T cells with cerebriform nuclei or Sezary cells of peripheral blood lymphocytes. In May 2007, the International Society for Cutaneous Lymphoma proposed that the diagnosis of Sezary syndrome should be made with molecular and flow cytometry based on a large clonal population of abnormal T cells in the blood in addition to erythroderma of 80% of the body or more (Hwang et al.).

Treatment

CTCL treatment is determined by the type and stage of the disease. Treatments are divided into two types: skin-directed and systemic treatments. Options include phototherapy, radiation, topical therapy, systemic single-agent chemotherapy, combination chemotherapy, and combined therapies. Commonly used topical therapies include chemotherapeutic agents (e.g., nitrogen mustard, retinoids). Topical treatments such as phototherapy and corticosteroids generally are used in early stages; systemic treatments, combination therapies, and chemotherapy are reserved for advanced-stage disease. The U.S. Food and Drug Administration has approved the following systemic agents for treatment of CTCL: denileukin, bexarotene, and vorinostat. Patients

with localized early-stage disease typically are treated with topical agents, skin-softening agents, anti-itch agents, and phototherapy (Trautinger et al., 2006). Clinical trials should be considered for patients with CTCL.

Case Study

M.O., a 28-year-old African American woman, presented to the National Cancer Institute (NCI) with a history of stage IV CTCL. Her symptoms began in 2003 when she experienced itching on her hands and feet during her second pregnancy. Each week, more skin became affected and progressed to peeling, with increased edema and scalp alopecia. In September 2003, M.O. was given a high-dose steroid regimen by her family practitioner to resolve the edema of her hands and feet. Her symptoms increased when the steroids were tapered, so the steroid dose was escalated. Two months later, M.O. developed significant edema to the hands and feet. She then consulted a dermatologist and was diagnosed and treated for severe eczema. Treatment proved ineffective, as M.O. developed staphylococcal infections. The infections caused oozing ulcerations that were localized to her hands and feet but soon spread to the legs, causing widespread bright-red skin and edema. Multiple skin biopsies were inconclusive. Despite her condition, M.O. successfully delivered a full-term baby by cesarean section in 2004.

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