

Non-Hodgkin Lymphoma

Examining mycosis fungoides and Sézary syndrome in the context of oncology nursing

Karla V. Ow, MSN, FNP-C, and Jeannine M. Brant, PhD, APRN, AOCN®, FAAN



BACKGROUND: Mycosis fungoides and Sézary syndrome are the most common non-Hodgkin lymphomas that manifest primarily in the skin. Although early-stage disease has an excellent long-term survival rate, advanced disease carries a poor survival rate. Given the lengthy and complex clinical course, nurses are at the forefront of education and supportive care management for patients and caregivers.

OBJECTIVES: This article aims to provide an overview of mycosis fungoides and Sézary syndrome and to highlight practice considerations for optimal nursing care.

METHODS: Clinical presentation, diagnosis, management, and nursing consideration are discussed.

FINDINGS: Oncology nurses have a vital role in educating patients and their caregivers about the side effects of cancer treatment, appropriate skin care, and infection risk.

KEYWORDS

mycosis fungoides; Sézary syndrome; supportive care; pruritus; skin infections

DIGITAL OBJECT IDENTIFIER

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PRIMARY CUTANEOUS LYMPHOMAS (PCLs) ARE A RARE TYPE of malignant non-Hodgkin lymphomas that initially present in the skin. The World Health Organization–European Organisation for Research and Treatment of Cancer classification for cutaneous lymphomas serves as the gold standard for diagnosis and classification of PCLs. Under the umbrella of PCLs are cutaneous T-cell lymphomas (CTCLs) and cutaneous B-cell lymphomas (CBCLs) (Willemze et al., 2019).

CTCLs include a spectrum of diseases that are characterized by malignant T-cell lymphocytes infiltrating the skin (Larocca & LeBoeuf, 2019). The incidence rate of CTCLs is approximately 10.2 cases for every 1 million people in the United States (Larocca & Kupper, 2019). Mycosis fungoides and Sézary syndrome are the two classic subtypes of CTCLs (Berg et al., 2017).

Mycosis fungoides accounts for about 55% of all CTCLs, and Sézary syndrome accounts for about 2%–3% of all CTCLs (Trautinger et al., 2017). The incidence of mycosis fungoides and Sézary syndrome is about 2,500–3,000 new cases per year and represent only about 0.4% of all non-Hodgkin lymphomas per year (Alpdogan et al., 2019). Although mycosis fungoides can occur in children and young adults, it is predominantly seen in men older than age 60 years, and higher incidence is seen in Black people as compared to White people (Photiou et al., 2018). Sézary syndrome is a leukemic variant and an extremely rare subtype. It is characterized by a more aggressive behavior with diffused pruritic erythroderma, lymphadenopathy, and blood involvement (Pulitzer, 2017). Sézary syndrome can evolve from preexisting mycosis fungoides or present de novo (Virmani et al., 2017).

Etiology

CTCLs are presumed to arise from a background of chronic inflammation because of a persistent antigen exposure in genetically susceptible individuals (Duvic, 2013; Wilcox, 2017). Although risk factors associated with mycosis fungoides and Sézary syndrome have not been clearly established, environmental and occupational exposures (e.g., chemicals, solvents), infections (e.g., *Staphylococcus aureus*), viruses (e.g., cytomegalovirus, human T-cell lymphotropic virus type 1, Epstein–Barr virus), and immunosuppressive