

# **CUTANEOUS T-CELL LYMPHOMA (CTCL):**

A Rare Blood Cancer Seen on the Skin

## WHAT IS CTCL?

CTCL refers to a group of non-Hodgkin lymphomas that are derived from T-lymphocytes, a type of white blood cell. While CTCL is largely confined to the skin at diagnosis, it impacts each patient differently, with symptoms ranging from rash-like patches to thicker, itchy plaques and raised tumors. Despite ongoing research, the cause of CTCL remains unknown.<sup>1-4</sup>

- In the western world, the estimated annual incidence of CTCL is 1 in every 100,000.5
- CTCL impacts twice as many men as women.<sup>5-6</sup>
- Risk for CTCL increases with age, and patients are typically diagnosed in their mid-50s.<sup>7</sup>







## TYPES OF CTCL

CTCL includes a wide variety of different disorders. Among the various subtypes of CTCL are **mycosis fungoides** and **primary cutaneous CD30-positive lymphoproliferative disorders (LPDs)**. Overall survival rates for the disease have been shown to vary based on subtype, with 91 percent of patients with mycosis fungoides and between 90-100 percent with LPDs living for five years following diagnosis.<sup>4,8-9</sup>

**Mycosis fungoides**, named for the mushroom-like appearance of tumors, is the most common subtype and accounts for 50 to 70 percent of all cases of CTCL. Typically slow-growing, mycosis fungoides is often localized to the skin. It can be present for long periods before diagnosis due to its resemblance to other conditions. There is variability among which patients express the CD30 protein, a tumor marker in various types of lymphoma. 1-2.5,8,10-12

**LPDs** are the second largest subgroup of CTCL and include primary cutaneous anaplastic large cell lymphoma (pcALCL) and different types of lymphomatoid papulosis (LYP). These cancers are characterized by cells that highly express CD30 and manifest as raised, red skin lesions, nodules or tumors.<sup>1,13-15</sup>



### **DIAGNOSIS**

Diagnosing CTCL in its earlier stages presents challenges, as lesions often resemble benign dermatitis. From the onset of symptoms, it can take an average of five to six years for patients to receive a diagnosis, and many are diagnosed by dermatologists rather than hematologists or oncologists.<sup>1,16</sup> The following tests and procedures may be performed to diagnose CTCL:



**Physical exam** 







Blood test

**Biopsy** (skin and lymph nodes)

Screening tests (CAT scan, PET scan, MRI)

There is a need for improvements in diagnosis, including tests that can more easily identify CTCL in early stages through greater sensitivity and more specific biomarkers. Differentiating between subtypes via methods such as CD30 testing may help ensure patients receive the appropriate treatment.

### TREATMENT OPTIONS

Due to the heterogeneity of CTCL, there is no single way to manage the disease. Patients with CTCL can receive either skin-directed or systemic therapies, depending upon the stage of their disease. There is no known cure for CTCL, and few new treatment options have been introduced over the last several years.

**Skin-directed therapies**, used in earlier stage disease, include phototherapy, surgical excision, topical steroids, local radiotherapy, topical chemotherapies, and protection and moisturizing of the affected areas. In addition, topical antibiotics can be prescribed to treat infections associated with CTCL.<sup>1-2,17-18</sup>

**Systemic therapies** may be used in patients with advanced CTCL and in those who do not respond well to skindirected therapies. Biologics, immunotherapies, and chemotherapies are examples of systemic therapies, which target cancer at the cellular level.<sup>1-2,17</sup>

For systemically treated patients with progressing CTCL, complete and durable responses are rare. Therapies are currently aimed at symptom control and improved quality of life.<sup>19</sup>

<sup>19</sup> Prince HM, Whittaker S, Hoppe RT: How I treat mycosis fungoides and Sezary syndrome. Blood 114:4337-4353, 2009.



<sup>1</sup> Leukemia & Lymphoma Society. Cutaneous T-Cell Lymphoma Facts. https://www.lls.org/sites/default/files/file\_assets/cutaneoustcelllymphoma.pdf. Accessed July 5, 2016. 2 Lymphoma Research Foundation. Getting the Facts: Cutaneous T-Cell Lymphoma. http://www.lymphoma.org/atf/cf/%7Baaf3b4e5-2c43-404c-afe5-fd903c87b254%7D/LRF\_FACTSHEET\_CUTANEOUS\_TCELL\_LYMPHOMA\_CTCL.PDF. August 2015.

<sup>3</sup> Wilcox RA. Cutaneous T-cell lymphoma: 2016 update on diagnosis, risk-stratification and management. American Journal of Hematology, 2016. 91(1):152-165.

<sup>4</sup> Bekkenk MW, et al. Primary and secondary cutaneous CD30(+) lymphoproliferative disorders: a report from the Dutch Cutaneous Lymphoma Group on the long-term follow-up data of 219 patients and guidelines for diagnosis and treatment. Blood, 2000. 95(12):3653-61.

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<sup>6</sup> Agar SN, et al. Survival Outcomes and Prognostic Factors in Mycosis Fungoides/Sézary Syndrome: Validation of the Revised International Society for Cutaneous Lymphomas/European Organisation for Research and Treatment of Cancer Staging Proposal. Journal of Clinical Oncology, 2010. 28(31):4730-4739. doi:10.1200/JCO.2009.27.7665.

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<sup>9</sup> Wieser, I., Tetzlaff, M. T., Torres Cabala, C. A. and Duvic, M. (2016), Primary cutaneous CD30+ lymphoproliferative disorders. JDDG: Journal der Deutschen Dermatologischen Gesellschaft, 14: 767–782. doi:10.1111/ddg.13117.

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<sup>11</sup> Criscione VD, and Weinstock MA. Incidence of cutaneous T-cell lymphoma in the United States, 1973-2002. Archives of Dermatology, 2007. 143 (7):854-859

<sup>12</sup> Prince, Miles H. CD30 As a Target for the Treatment of Cutaneous T-Cell Lymphoma. Journal of Clinical Oncology, 2015. doi: 10.1200/JCO.2015.61.9

<sup>13</sup> Rukhsana A et al, Cutaneaous T cell lymphomas. Chapter 10. Contemporary Hematology 2013.

<sup>14</sup> Wilson LD, et al. Clin Lymphoma Myeloma Leuk 2012;12:291–6.

 $<sup>15\</sup> Cutaneous\ Lymphoma\ Foundation.\ Fast\ Facts\ About\ Primary\ Cutaneous\ Anaplastic\ Large\ Cell\ Lymphoma\ (PCALCL).\ http://www.clfoundation.org/primary\ Cutaneous\ Cell\ Ce$ 

online-learning-center/disease/calc/fast-facts-about-primary-cutaneous-anaplastic-large-cell-lymphom. Accessed July 20, 2016. 16 Mishra, A. and Porcu, P. Early CTCL diagnosis, a (miR)age no more? Blood, 2011. 118:5717-5718; doi:10.1182.

<sup>17</sup> Cutaneous Lymphoma Foundation. Cutaneous Lymphoma Fast Facts. http://www.clfoundation.org/sites/default/files/publications/CL\_Fast\_Facts.pdf. Accessed July 5, 2016.

<sup>18</sup> National Comprehensive Cancer Network. Non Hodgkin's Lymphoma: NCCN Clinical Practice Guidelines in Oncology. http://www.nccn.com. Accessed November 2016.