Cutaneous T-cell Lymphoma

What is cutaneous T-cell lymphoma? Cutaneous T-cell lymphoma (CTCL) is a cancer in which there is a proliferation of abnormal T-cell lymphocytes (a type of white blood cell) in the skin and blood. Occasionally, it can spread to the lymph nodes and internal organs. CTCL usually appears after age 50, and men are more commonly affected than women.

What causes CTCL? The cause is currently unknown. Fortunately, it is a rare disease, affecting only 5-10 persons per million.

What does CTCL look like? There are several different types of CTCL:

- **Mycosis fungoides** – This is the most common type of CTCL in which the lymphocytes infiltrate the skin. The lesions can be flat, red and scaly patches, thicker plaques, or large tumors. Itching is variable. The cancer usually remains confined to the skin and has a slow progression. Spread to other organs is more common with tumor development.

- **Sézary syndrome** - The three characteristic features of Sézary syndrome are generalized red skin, enlarged lymph nodes, and atypical lymphocytes (Sézary cells) in the blood. The skin on the entire body becomes red, thickened, dry or scaly and is usually very itchy. This is a more serious and aggressive form of CTCL.

- **Primary Cutaneous T-Cell Lymphomas** - These lymphomas present as solitary or localized skin lesions that have a tendency to ulcerate and may spontaneously regress. Certain types have an indolent course, others are aggressive in nature.

- **T-cell leukemia** – In leukemia, the atypical cells are multiplying in the blood.

- **Subcutaneous T-Cell Lymphoma** - Typically, patients present with nodules in the deeper fat, usually on the legs, accompanied by weight loss, fever, and fatigue.

How is CTCL diagnosed? The diagnosis of CTCL is made with a skin biopsy; characteristic changes in the affected skin are seen under the microscope. The diagnosis is often delayed for months or years since early CTCL can be difficult to tell apart from other skin conditions, particularly eczema. Multiple and repeated skin biopsies are sometimes necessary to confirm the diagnosis. If the lymph nodes are affected and enlarged, they may also be biopsied to see if cancer is present. The blood count is normal in most patients with CTCL, but the presence of Sézary cells is characteristic of Sézary syndrome. Patients with advanced CTCL may have CT or MRI scans to determine whether the internal organs are affected.

How is CTCL treated? Treatment of individual patients varies, and depends on the disease progression, local expertise, and available drugs and technologies. Although there is no cure for CTCL, the following therapies have been shown to be helpful in reducing symptoms and the number of cancerous cells:

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- Topical steroids – reduces inflammation
- Topical nitrogen mustard or BCNU – chemotherapy kills cancerous cells
- Systemic chemotherapy and interferon – kills and controls the growth of cancerous cells
- Topical or oral retinoids – vitamin A derivatives that interfere with cell growth
- PUVA or UVB phototherapy – specialized light treatment slows the rapid growth of skin cells
- Photopheresis – blood is removed from the body, treated with light therapy and returned to the circulation
- Localized radiotherapy and electron beam radiotherapy – kills cancerous cells

What is the prognosis of CTCL? The course of CTCL is unpredictable and varies from patient to patient. Fortunately, unlike some other lymphomas, the outlook is generally good if it is caught and treated early. In most patients, CTCL remains confined to the skin for many years and does not interfere with daily function. Those with early disease involving less than 10% of their body have a normal life expectancy. Ten percent of patients will develop progression of disease, with involvement of the lymph nodes and internal organs, or other serious complications.