

## Sezary Syndrome and T – Cell Lymphoma

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### Abstract

Lymphoma is a cancer of the white blood cells; the body has two main types of lymphocytes: B lymphocytes, or B-cells, and T lymphocytes. T-cell lymphoma is a disease in which T lymphocyte cells become cancerous. One of the most common forms of T-cell lymphoma is cutaneous, or skin, lymphoma, because it starts in the lymphocytes in the skin. Cutaneous lymphoma actually describes many different disorders with various signs and symptoms, outcomes and treatment considerations. Sézary Syndrome (SS) is characterized by erythroderma, generalized lymphadenopathy, and the presence of circulating atypical lymphocytes, which are difficult to identify by morphologic data.

**Keywords:** Sezary Syndrome (SS); T-lymphocyte; Blood cancer

### Introduction

Sezary syndrome is a wild form of blood cancer which called cutaneous T-cell lymphoma. Cutaneous T-cell lymphomas take place when T cell of white blood cells, become malignant and migrate in to other organs [1].

### Epidemiology

Sezary disease is more common in males as compare to women with a ratio of 2:1 and the mean age of diagnosis is between 55 and 60 years of age. Affected individuals survived an average of 2 to 4 years after development of the condition, although survival has improved with newer treatments [2].

### Causes

The reason of Sezary Syndrome (SS) is unknown. Most of people suffer from SS have chromosomal abnormalities, which include the loss or gain of genetic material. These abnormalities occur in the DNA of cancerous cells. Although, abnormalities have been found on most chromosomes, but there found in some regions compare to others. The most affected regions which have losses of DNA from regions of chromosomes 10 and 17 or additions of DNA to regions of chromosomes 8 and 17 [3].

### Sign and Symptom

In Sezary syndrome, Sezary cells are migrate in to skin, lymph nodes, and blood. These cells lead to enlargement of organs, including the lymph nodes, liver, spleen, and bone marrow. Affected People with Sezary syndrome suffer from itchy rash (erythroderma) that covers large portions of their body. People with Sézary syndrome also have enlarged lymph nodes (lymphadenopathy). Other common signs and symptoms of this condition include hair loss, palm plantar keratoderma, ectropion. Some people with Sézary syndrome are less able to control their body temperature than people without the condition [4].

### Diagnosis

There are different methods for diagnosis of Sezary syndrome, which including a physical exam and history; blood tests to identify antigens, or markers, on the surface of the cells in the blood by using flow cytometry technique; a skin and/or lymph node biopsy; and a series of imaging tests such as CT (computerized axial tomography), MRI (magnetic resonance imaging) and/or PET (positron emission tomography) scans to determine if the cancer has spread to lymph nodes or other organs. In addition to these diagnostic tests, occasionally a bone marrow biopsy may be necessary to verify complete staging [5].

### Treatment

There are many effective therapies available to treat Sézary syndrome. Second-line drug for Sezary syndrome include Romidepsin, Mogamulizumab, and Vorinostat. Treatments are often used in combination between phototherapy and chemotherapy. The specific treatment for individual patients is based on a variety of factors, including the patient's general health and stage of the disease [6].

### There are several types of standard treatment for Sézary syndrome

- Biologic, or immunotherapy, therapy is a treatment used to stimulate a patient's own immune system to fight the cancer.
- Chemotherapy, a drug given either orally or through an infusion in a vein, to stop the growth of rapidly dividing cancer cells.
- Extracorporeal photopheresis (ECP), a procedure used to expose the blood to ultraviolet light.
- Histone deacetylase inhibitors, a class of drugs that cause a chemical change that stops tumor cells from dividing.
- Phototherapy, for example, the drug psoralen and ultraviolet-A light radiation (PUVA) directed to the skin or skin- directed ultraviolet-B (UVB) or narrow band ultraviolet-B (NBUVB).
- Radiation therapy, which uses high-energy X-rays or other types of radiation to kill cancer cells or keep them from growing.

- Retinoids, which are drugs related to vitamin A and can slow certain types of cancer cells [7].

#### Some specific drugs include

- Alemtuzumab (Campath), a monoclonal antibody.
- Interferon alfa or interleukin-2, immune stimulants that bind to specific cell- surface receptors.
- Liposomal doxorubicin (Doxil), a chemo therapy that binds to DNA.
- Methotrexate (Trexall), an antimetabolite chemotherapy, which blocks the metabolism of cells.
- Vorinostat (Zolinza), a histone deacetylase inhibitor.

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