Sézary syndrome

Description

Sézary syndrome is an aggressive form of a type of blood cancer called cutaneous T-cell lymphoma. Cutaneous T-cell lymphomas occur when certain white blood cells, called T cells, become cancerous; these cancers characteristically affect the skin, causing different types of skin lesions. In Sézary syndrome, the cancerous T cells, called Sézary cells, are present in the blood, skin, and lymph nodes. A characteristic of Sézary cells is an abnormally shaped nucleus, described as cerebriform.

People with Sézary syndrome develop a red, severely itchy rash (erythroderma) that covers large portions of their body. Sézary cells are found in the rash. However, the skin cells themselves are not cancerous; the skin problems result when Sézary cells move from the blood into the skin. People with Sézary syndrome also have enlarged lymph nodes (lymphadenopathy). Other common signs and symptoms of this condition include hair loss (alopecia), skin swelling (edema), thickened skin on the palms of the hands and soles of the feet (palmoplantar keratoderma), abnormalities of the fingernails and toenails, and lower eyelids that turn outward (ectropion). Some people with Sézary syndrome are less able to control their body temperature than people without the condition.

The cancerous T cells can spread to other organs in the body, including the lymph nodes, liver, spleen, and bone marrow. In addition, affected individuals have an increased risk of developing another lymphoma or other type of cancer.

Sézary syndrome most often occurs in adults over age 60 and usually progresses rapidly; historically, affected individuals survived an average of 2 to 4 years after development of the condition, although survival has improved with newer treatments.

Although Sézary syndrome is sometimes referred to as a variant of another cutaneous T-cell lymphoma called mycosis fungoides, these two cancers are generally considered separate conditions.

Frequency

Sézary syndrome is a rare condition, although its prevalence is unknown. It is the second most common form of cutaneous T-cell lymphoma after mycosis fungoides, accounting for approximately 3 to 5 percent of cases of cutaneous T-cell lymphoma.

Causes

The cause of Sézary syndrome is unknown. Most affected individuals have one or more chromosomal abnormalities, such as the loss or gain of genetic material. These abnormalities occur during a person's lifetime and are found only in the DNA of cancerous cells. Abnormalities have been found on most chromosomes, but some regions are more commonly affected than others. People with this condition tend to have losses of DNA from regions of chromosomes 10 and 17 or additions of DNA to regions of chromosomes 8 and 17. It is unclear whether these alterations play a role in Sézary syndrome, although the tendency to acquire chromosomal abnormalities (chromosomal instability) is a feature of many cancers. It can lead to genetic changes that allow cells to grow and divide uncontrollably.

Inheritance

The inheritance pattern of Sézary syndrome has not been determined. This condition occurs in people with no history of the disorder in their family and is not thought to be inherited in most cases.

Other Names for This Condition

- Sezary erythroderma
- Sezary syndrome
- Sezary's lymphoma

Additional Information & Resources

Genetic Testing Information

- Genetic Testing Registry: Sezary syndrome (https://www.ncbi.nlm.nih.gov/gtr/conditions/C0036920/)

Genetic and Rare Diseases Information Center

- Sézary syndrome (https://rarediseases.info.nih.gov/diseases/7629/index)

Patient Support and Advocacy Resources

- National Organization for Rare Disorders (NORD) (https://rarediseases.org/)

Clinical Trials

- ClinicalTrials.gov (https://clinicaltrials.gov/search?cond=%22Sézary syndrome%22)

Catalog of Genes and Diseases from OMIM

- MYCOSIS FUNGOIDES (https://omim.org/entry/254400)

Scientific Articles on PubMed

- PubMed (https://pubmed.ncbi.nlm.nih.gov/?term=%28Sezary+Syndrome%5BMAJR%5D%29+AND+%28Sezary+syndrome%5BTIAB%5D%29+AND+english%5Bla%5D+AND+human%5Bmh%5D+AND+%22last+720+days%22%5Bdp%5D)

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Last updated May 17, 2021