

HAIRY CELL LEUKEMIA

Epidemiology

Hairy cell leukemia (HCL) represents 2% of adult leukemias. Approximately 1600 new cases per year are diagnosed in Europe, with a median age of 52 years at the time of diagnosis. HCL occurs more often in men.

Signs and symptoms

HCL may be asymptomatic in some cases. On contrary, some people experience signs and symptoms like enlargement of the spleen that can lead to a feeling of fullness in the abdomen, easy bruising, recurring infections, fatigue, weakness and weight loss.

Diagnosis and risk stratification

The diagnosis relies on blood tests and a bone marrow biopsy. HCL is characterized by the presence of BRAF mutation. Radiological examinations such as CT-scan or US -tomography are helpful to assess the presence of splenomegaly or lymph nodes enlargement. There is no international prognostic system for risk stratification, however there are some clinical variables that are considered at diagnosis: degree of cytopenias and the presence of lympho-adenopathies. Mutations of TP53 and lack of somatic mutation of IGHV correlate with a more aggressive course.

Treatment

A treatment is not always required. If treatment is required, chemotherapy is effective at putting hairy cell leukemia in remission for years. Our treatment approach relies on national and international guidelines, i.e. ESMO guidelines (<https://www.esmo.org/guidelines/haematological-malignancies/hairy-cell-leukaemia>), Consensus guidelines for the diagnosis and management of patients with classic hairy cell leukemia (Blood, 2017 DOI: [10.1182/blood-2016-01-689422](https://doi.org/10.1182/blood-2016-01-689422)).

Two chemotherapy drugs are used in hairy cell leukemia: cladribine and pentostatin. Cladribine is widely used as first line treatment and is administered daily over several days and does not require hospitalization. Side effects of cladribine may include infection and fever. In some cases, cladribine may be followed by biological therapy with Rituximab, a monoclonal antibody. Pentostatin is more often used in patients with unsatisfactory responses to cladribine. The remission rates and side effects are similar between these two drugs.