

Hairy Cell Leukemia

Hairy cell leukemia is a rare but distinctive mature B cell tumor that constitutes about 2% of all leukemias. This disease predominantly affects middle aged adults with a median age of 55. It is also more common in males with a male to female ratio of 5 to 1. Hairy cell leukemia derives its name from the appearance of the cancerous cells, which have fine filamentous hair-like projections. The diagnosis can also be confirmed by viewing the cells with a special stain known as TRAP stain, which stands for tartrate resistant acid phosphatase. Because these tumors originate from B cells, they typically express B cell markers including CD 19 and CD 20. Clinical manifestations mostly reflect infiltration of the bone marrow, liver and spleen. Splenomegaly is particularly massive and is the most common and sometimes only abnormal physical finding.



PLAY PICMONIC

Pathophysiology

Mature B Cell Tumor

[Mature \(B\) Cell Basketball with Tumor-guy](#)

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Signs and Symptoms

Splenomegaly

[Spleen-balloon](#)

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Diagnosis

Flow Cytometry

[Side-toe-meter Measuring-cells](#)

Flow cytometry is a study done by suspending cells in a stream of fluid and passing them by an electronic detection apparatus. The cells are then analysed using laser or impedance based technology, evaluating cell size, count and biomarker detection. Flow cytometry is used to detect hairy cell leukemia's distinctive immunophenotype staining with antibodies to CD5 (negative), CD10 (negative), CD23 (negative), CD20 (abnormally bright), CD22 (abnormally bright), CD11c (abnormally bright), CD25 (abnormally bright), CD103 (positive), and CD123 (positive).

Cells Have Filamentous Hair-Like Projections

[Hairy-guy](#)

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Stains TRAP Positive

[Bear TRAP](#)

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Dry Tap on Bone Marrow Aspiration

[Dry Tap from a Bone Marrow Aspirating-ss](#)

In hairy cell leukemia, there is extensive bone marrow effacement, with replacement of hairy cells. There is interstitial or patchy infiltration of the marrow with widely spaced lymphoid cells, as well as increased amounts of reticulin fiber deposition. Because of these bone marrow changes, bone marrow aspiration leads to a "dry tap."

Treatment

Cladribine

[Cladribine](#)

Cladribine, which is also known as Leukostatin, is used to treat hairy cell leukemia and multiple sclerosis. It works as a purine (adenosine) analog, inhibiting adenosine deaminase. This interferes with the cell's ability to process DNA and destroys them.

Pentostatin

[Pentostatin](#)

Though cladribine is the first line treatment for hairy cell leukemia, pentostatin is equally effective in terms of response rate and remission duration. Patients taking pentostatin must have adequate renal function, as pentostatin is cleared through the kidneys and cannot be used in those with renal dysfunction.