Mast cell leukemia

Mast cell leukemia is a particularly aggressive subtype of acute chronic myelocytic leukemia that sometimes occurs de novo but can, rarely, evolve from transformation of chronic chronic myelocytic leukemia into the more aggressive acute chronic myelocytic leukemia. During a small proportion of cases, acute mastocyte leukemia may evolve from a more progressive sort of systemic mastocytosis. The diagnosis of acute mastocyte leukemia by the WHO criteria includes the need for a prevalence of 20% neoplastic mast cells in marrow and 10% in blood. If the mast cells represent but 10% of blood cells, the tumor is named "aleukemic" mastocyte leukemia.

Signs and symptoms

Acute mastocyte leukemia may be a rapidly progressive disorder with leukemic mast cells in blood and in large numbers in marrow. The common signs and symptoms include fever, headache, flushing of face and trunk. the standard cutaneous mastocyte infiltrates of urticaria pigmentosa are usually not present before, during, or after diagnosis in patients who have mastocyte leukemia. Symptoms include abdominal pain, bone pain, and peptic ulceration which are more prevalent than in other subtypes of acute chronic myelocytic leukemia. These former symptoms are thanks to release of a substance called histamine from neoplastic mast cells. Enlargement of the liver and spleen, or hepatosplenomegaly is characteristic. The mast cells release also many anticoagulants like heparin which may cause serious bleeding. Liver and splenic dysfunction also contributes to hemorrhage. Involvement of the bone can cause osteoporosis. Abdominal ultrasound or computed tomography (CT) scanning is employed to seem for hepatosplenomegaly and lymphadenopathy. Plain radiography and bone densitometry are often wont to assess bone involvement and therefore the presence of osteoporosis. Endoscopy and biopsy are often useful if gut involvement is suspected.

Diagnosis

Cytochemistry

Cytochemical properties of the leukemic cells must be typical of mastocyte derivation (presence of metachromatic granules staining with alpha-naphthyl chloroacetate esterase, but not with peroxidase). mastocyte tryptase is an enzyme contained in mastocyte granules. mastocyte numbers are best estimated by tryptase immunostaining because very poorly granulated cells may stain very weakly if in the least for alpha-naphthol chloroacetate esterase

Tumor markers

The leukemic cells usually are strongly positive for CD13, CD33, CD68, and CD117. Characteristically, basophil (e.g. CD11b, CD123) and monocyte markers (CD14, CD15) are absent. The cells usually express CD2 and CD25. Malignant mast cells overexpress the anti-apoptosis gene, bcl-2. A mutation called KIT mutation is detected in most patients.

Biochemistry

Total serum tryptase is elevated in mastocyte leukemia. Normal total (alpha + beta) serum tryptase is approximately 6 micro g/L (range 0 to 11 micro g/L). Values of several hundred micro g/L are characteristic of mastocyte leukemia. Plasma and urinary histamine levels are frequently elevated in mastocyte leukemia. Histidine decarboxylase (HDC) is that the enzyme that catalyzes the reaction which produces histamine from histidine. Measurement of histidine carboxylase within the marrow cells of patients with mastocyte leukemia may be a very sensitive marker of mast cells.

Treatment

Immunoglobulin E (IgE) is vital in mastocyte function. Immunotherapy with anti-IgE immunoglobulin raised in sheep resulted during a transient decrease within the numbers of circulating mastocyte s in one patient with mast cell leukemia. Although splenectomy has led to brief responses in patients with mastocyte leukemia, no firm conclusions on the efficacy of this treatment are possible.

Chemotherapy with combination of cytosine arabinoside and idarubicin, daunomycin, or mitoxantrone as for acute chronic myelocytic leukemia has been used. Somatic cell transplantation is an option, although no experience exists concerning responses and outcome

Prognosis

Acute mastocyte leukemia is extremely aggressive and features a grave prognosis. In most cases, multi-organ failure including bone marrow failure develops over weeks to months.[16] Median survival after diagnosis is merely about 6 months.