PB2069 A CASE OF HAIRY CELL LEUKEMIA PRESENTED AS HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS

Topic: 18. Indolent and mantle-cell non-Hodgkin lymphoma - Clinical

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Background:

Hairy cell leukemia (HCL) is an indolent and uncommon chronic B cell lymphoproliferative disorder. The clinical picture is mainly dominated by cytopenias and splenomegaly, with some patients only receiving a watch and wait approach. It has generally a very favorable prognosis.

Hemophagocytic lymphohistiocytosis (HLH) is a rare syndrome of uncontrolled inflammation that occurs due to severe systemic immune activation, mainly secondary to infection, autoimmune diseases, and malignancies (mostly aggressive T/NK-cell or B-cell lymphomas) and carries a dismal prognosis.

Aims:

To present a case of a newly diagnosed HCL that manifests as HLH.

Methods: Clinical registry review.

Results:

Female, 62 years old, ECOG-PS 0, with an unremarkable medical history. She presented to the emergency department with a two-week history of constitutional symptoms (fever, marked weight loss and night sweats) and skin lesions. Physical examination revealed hepatosplenomegaly and no adenomegalies. Blood tests showed bicytopenia (hemoglobin 10.8 g/dL normocytic/normochromic and leukocyte 2.1x10^9/L), an elevated C-reactive protein (14 mg/dl) and an elevated erythrocyte sedimentation rate (85 mm/h); lactate dehydrogenase (LDH), kidney function and liver function analysis were unremarkable. She was then admitted as an inpatient to the hospital for additional etiologic investigation. An extensive screening for infectious agents and autoimmune diseases was performed but showed no alterations. Skin lesion biopsy was not diagnostic, for two consecutive times, suggesting the possibility of erythema nodosum or vasculitis, with normal immunophenotyping.

Bone marrow trephine (dry tap for aspirate) showed a small focal invasion by B-cells with HCL phenotype (CD20+/Annexin A+/ CD23+/Cyclin D1+). PET/CT scan showed infra-diaphragmatic hypermetabolic adenopathies (maxSUV 15.1) and splenic involvement.

Intraabdominal lymph node biopsy showed a diffuse involvement by HCL clonal B-cells (CD20+/CD10-/CD25+/CD5-/Annexin A+). New peripheral blood immunophenotyping showed 0.4% B-clonal cells with HCL phenotype. The BRAF V600E gene mutation was positive in both products. The diagnosis of HCL (classical) was performed.

Throughout the next month as she remained at the hospital for investigation, she had been febrile with no germs identification and no improvement despite multiple antibiotic courses.

Despite the usage of corticosteroids and later life-saving rescue with cladribine 0.12 mg/kg 5 days, she developed severe multiorgan dysfunction - pancytopenia (hemoglobin 6.9 g/dL, leukocyte 0.6x10^9/L, platelet 10x10^9/L), liver dysfunction (bilirubin 24.7 mg/dL, gamma-glutamyl transferase 1071 U/L, alkaline phosphatase 707 U/L, LDH 1771 U/L), kidney failure (creatinine 3.14 mg/dL) and coagulopathy (fibrinogen 111.0 mg/dL). Markedly elevated serum...
ferritin (10271 ng/mL), triglycerides (618 mg/dL) and soluble CD25 (5119 pg/mL) supported a diagnosis of HLH (6 of 8 HLH-2004 criteria and HScore 268 points) the patient immediately started treatment with dexamethasone 10 mg/m² per day. Due to respiratory failure, she was admitted in ICU, where she unfortunately died due to *Klebsiella pneumoniae* pneumonia, a month and a half after the beginning of the clinical symptoms.

**Summary/Conclusion:** Conclusion: HCL is usually associated with an indolent clinical picture and with a good prognosis, while HLH is mainly associated with aggressive lymphoma. This case illustrates a case of HCL with a difficult diagnosis and highlights the importance of considering HCL as a triggering factor for HLH.