ISSUE
23
March
2022

QSP Monthly

HORIBA Medical

NEWSLETTER OF THE QUALITY SLIDE PROGRAM

This issue

Last Month's Slides

Slide 1
Lymphocytosis /Atypical lymphs? Monomorphic hyperlymphocytosis associated with a large number of naked nuclei. Immunophenotyping of circulating lymphocytes requested. Expert comment: Probable CLL.

Slide 2
See case study on right.

Slide 3
Intensive care unit and preoperative medicine. Multiple analyser alarms. Anisocytosis (++). Microcytosis (++). Macrocytosis (++). RBC targets (+). Expert comment: RBC targets: Underlying hepatic disorder. No microcytes.

Slide 4
Anisocytosis (++). Eosinophilia. Discrete basophil: Nothing to report.

Slide 5
Hyperleukocytosis. Balanced myelemia. Basophilia Presence of erythroblasts. Probable myeloproliferative syndrome (CML?).

Slide 6
Nothing to report.

Monthly Digital Case Study

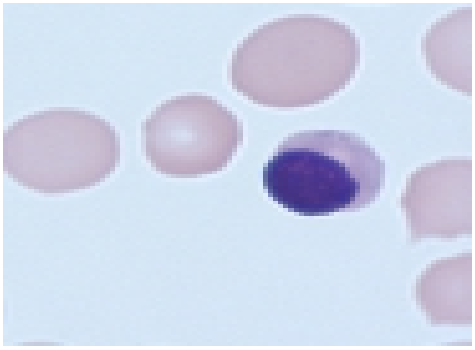
February 2022, Slide 2

Presentation

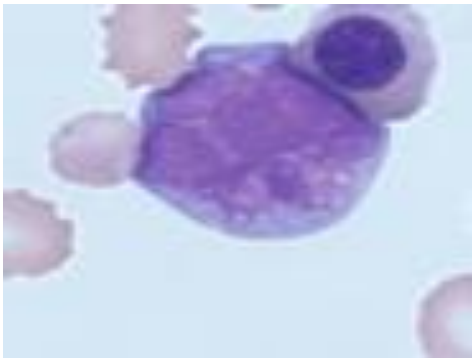
Male, 84 years old. Leucocytosis and erythroblastosis

FBC Results

WBC 84.2 (10^3/mm3)
RBC 2.82 (10^6/mm3)
HGB 8.0 (g/dL)
HCT 24.3 (%)
MCV 86.0 (fL)
MCH 28.4 (pg)
MCHC 32.9 (g/dL)
PLT 54 (10^3/mm3)
Neutrophils 10.6
Lymphocytes 39.0
Monocytes 2.4
Eosinophils 1.6
Basophils 0.8



Erythroblast



Blast cell

Slide Review

Anisocytosis (++). Poikilocytosis (+). Echinocytes (+). Hypogranulation of neutrophils (+). Blast population estimated at 40% of leukocytes. To be compared with the rest of the clinical assessment. Expert comment: Indicative of AML.

AML is the most common type of acute leukaemia in adults, and progresses quickly if left undiagnosed. AML results when myeloid stem cells develop into immature white cells called MYELOBLASTS. The myeloblasts in AML are abnormal and do not develop into healthy cells.

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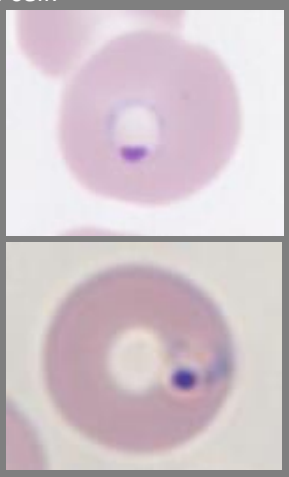
Cell Quiz:

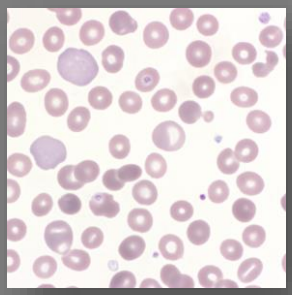
What is the feature within the red cell:



A) Howell Jolly Body
B) Ring form of P. Falciparum
C) Platelet

Last Month's Cell Quiz:

The film below is from a patient with Haemolytic Anaemia. What feature do you see?



A) Target cells

B) Spherocytosis

C) Poikilocytosis

Answer: B
The film shows SPHEROCYTOSIS, and is from a patient with haemolytic anaemia. Spherocytes appear as round red cells that lack the central area of pallor. They appear darker and smaller than a normocytic red cell. A raised MCHC is noted in the FBC result.

These leukaemic cells build up in the bone marrow and blood, preventing healthy WBC's platelets doing their job. This results in infection, anaemia and bleeding tendency.

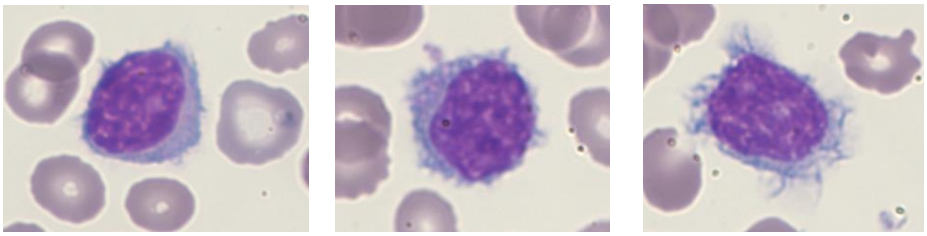
There are sub-types of AML, which are based on the maturity of the leukaemic cells at the time of diagnosis, and their variation from normal cells.

Acute Promyelocytic Leukaemia is a sub-type of AML which occurs when there is a switching of the gene on chromosome 15 with genes on chromosome 17, resulting in an abnormal gene, called *PML-RARA*. This abnormal gene sends a message which stops promyelocytes maturing.

Rare Lymphoid Clonal Abnormalities

Hairy Cell Leukaemia (HCL)

HCL is a rare clonal chronic lymphoproliferative disorder of B Cell Lymphocyte origin which accounts for about 2% of all leukaemias. In the 1920's it was originally described as histiocytic leukaemia, malignant reticulosis and lymphoid myelofibrosis. The disease was first officially described in 1958 by cancer researcher Bertha Bouroncle from the Ohio state University USA. The term Hairy Cell Leukaemia was first used to describe the appearance of hair like projection of the mononuclear cells in the disorder in 1966 by Shrek and Donelly. See below:



Hairy Cells showing fine cytoplasmic projections, larger than anormal lymphocyte, weakly basophilic cytoplasm.

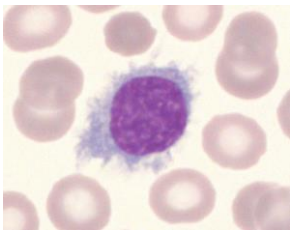
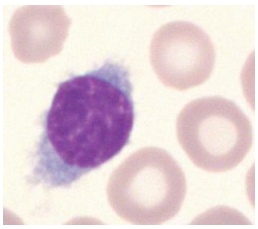
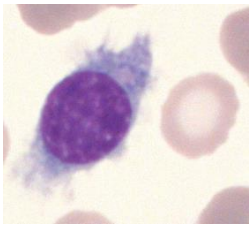
The incidence of HCl is most common in elderly males (median age at diagnosis 55), the commonest clinical symptoms being Pancytopenia and Splenomegaly (Splenomegaly due to extramedullary haematopoiesis) without lymphadenopathy are the common clinical presentation. Monocytopenia is almost always present, few abnormal lymphocytes may be present in the blood film. The abnormal cell population has slightly basophilic cytoplasm, and the cell nucleus is round slightly indented, or reniform, nucleoli are indistinct or absent. Bone marrow biopsy and Immunophenotyping is essential for correct diagnosis. Marrow aspiration may not be possible because of increased reticulin fibrosis. The recently identified mutation BRAFV600E has been identified in 100% of classical HCL patients.

HCL Variant

HCL Variant (HCL-V) accounts for about 10 % of all HCL cases and has features of both HCl and B-Prolymphocytic Leukaemia (B-PLL), the WBC is usually elevated as in B-PLL with abnormal cells having the cytoplasmic projection as in HCL but with the addition of a prominent central nucleolus similar to that in B-PLL, abnormal cells are often numerous in number, the BRAF V600E mutation is absent in variant HCL. Presenting symptoms are similar to those of classical HCL.

Splenic Lymphoma with Villous Lymphocytes (SLVL)

SLVL is a rare disorder accounting for less than 1% of lymphoid neoplasms. It is the leukemic counterpoint of splenic marginal zone lymphoma (SMZL) and is characterised by splenomegaly, often with no lymphadenopathy moderate lymphocytosis and the presence of villous (hairy), like lymphocytes in the blood film. The morphology of the cells is slightly different to HCL in that the nucleus is round with chromatin clumping, the cytoplasm is scanty to moderate in amount and weakly to moderate basophilic. The cytoplasmic projections are located at the poles of the cell (see below):

Differential Diagnosis

	HCL	HCL-V	SMZL (SLVL)
Gender M:F	4:!	M > F	1:1
Age	Median : 55	Middle Aged - elderly	> 60 yrs
Immunophenotype	CD20 bright +	CD20 bright +	CD20 bright +
	CD103+	CD103 +	CD103 -
	CD25 +	CD25-	CD25 -/+
	CD27 -	CD27 +	CD27 +
	CD11c+	CD11c+	CD11c +/-
	CD123+	CD123-	CD123-
	DBA 44+	DBA44 +	DBA44 +
	Annexin A1+	Annexin A1-	Annexin A1-
Molecular Genetics	BRAF V600 E mutation	BRAF V600E not mutated. MAP2KI mutations	Del7q (40%), NOTCH2 and KLF2 mutations

Prognosis and treatment of HCL, HCL-V and SLVL

Generally HCL is a less aggressive disease than is HCL-V with 95% of HCL patients surviving for more than 5 years post diagnosis. SLVL has a median overall median survival of 10 years, approx. 30% of these patients will develop an aggressive form which greatly reduces the median survival. Treatment can take the form of – watchful waiting with regular medical reviews, chemotherapy (Cladribine and Pentostatin), immunotherapy (recombinant interferon alpha-2b), targeted therapy (Monoclonal antibody Rituximab, Moxetumomab pasudotox-tdfk if relapsed), and surgery (removal of enlarged spleen).

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Bibliography

https://www.sciencedirect.com/topics/nursing-and-health-professions/hairy-cell-leukemia

BJH Guidelines for diagnosis and management of HCL and HCL-V

Essential Haematology
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