HAEMA 2019; **10**(1):60-61

Images in Haematology

A stubborn lady with pancytopenia

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A beautiful and chic woman 62 years-old comes to your office complaining of easy tiredness and fatigue.

Her past medical history is uneventful apart from chronic low back pain after a serious car accident 6 years ago, at the age of 56. At that time she was told that she had "low levels of white cells but nothing to worry about". She provides the blood counts: WBC 1.46, neutros 0.43, lymphs 0.99, monos 0.14, eos 0.14, Hb 8.2, Hct 27.6%, MCV 99.6, MCH 29.6, MCHC 29.7, RDW 19.4, NRBC 2, Plts 80.0. 4 years later she complained for pain in the left upper quadrant which resolved with mild analgesics. The family doctor advised her to consult a Hematologist but she denied claiming that she felt absolutely healthy living a fully active life.

Now she comes to your office for fatigue. She denies infections, bleeding episodes, fever, weight loss or night sweats.

The physical examination reveals paleness, mild ankle swelling, hardly palpable liver, moderate splenomegaly, no palpable lymph nodes and no signs of bleeding or bruises. Auscultation of heart and lungs is unremarkable.

Blood counts: WBC 1.63, neutros 0.33, lymphs 1.14, monos 0.13, eos 0.02, Hb 7.4, Hct 22.6%, MCV 98.9, MCH 30.1, MCHC 30.4, RDW 21.3, NRBC 2, Plts 54.0. The blood film, meticulously searched, is unremarkable and she is persuaded to have a bone marrow aspiration and biopsy performed.

The aspiration is dry tap, however a tiny drop of material was spread on a glass slide (Fig. 1-2).

We note a population of medium size round cells with rounded nucleus and mature chromatin, obviously belonging to the lymphocyte series. The chromatin is fine, less dense than that of a normal lymphocyte and there are no nucleoli. The nucleus is surrounded by abundant grayish cytoplasm with short, pale "hair-like" projections with a pointed end.

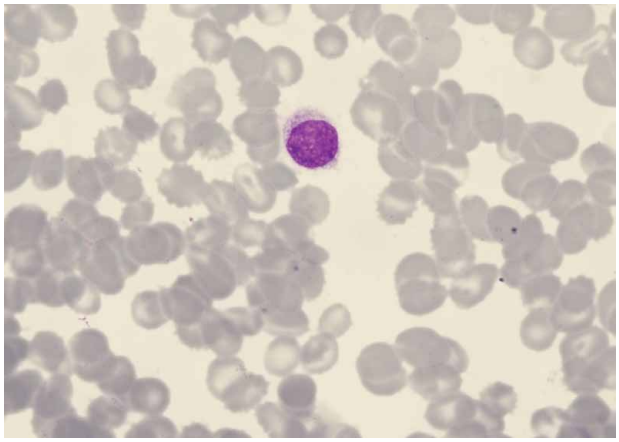

Figure 1. Peripheral blood. Normal red cells and a normal lymphocyte.

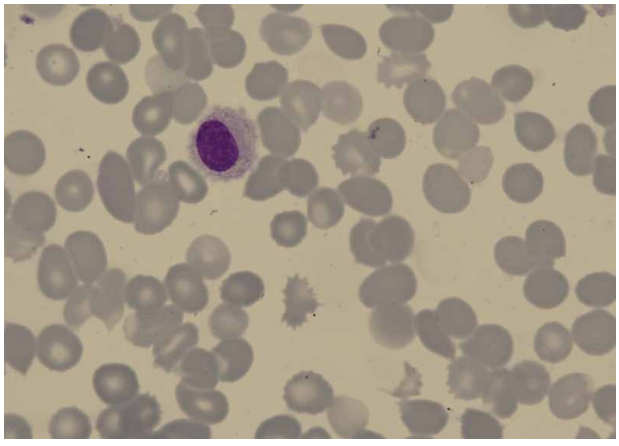

Figure 2. One cannot estimate the bone marrow cellularity since there are no fragments. There are a few erythroblasts, confirming that it is bone marrow material and not only peripheral blood.

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Received 1 Sep 2018; Accepted 26 Sep 2018

The immunophenotype - strong positivity of CD20, CD22, CD25, CD11c, CD103 and CD123, negativity of CD10, CD5, confirms the diagnosis of Hairy Cell Leukemia (HCL). In addition Annexin A1 (ANXA1) is positive, distinguishing classic HCL, which stains positive, from both splenic marginal zone lymphoma and HCL-variant, which are always negative.

COMMENTS

This case shows that HCL is a slowly progressing disease. As in most low-grade B-cell neoplasms treatment may be delayed and the strategy of "wait and watch" is absolutely warranted. In asymptomatic patients there is no general agreement when to start treatment and -as we can see in this case – blood counts falling below the threshold of 1.0 neutrophils and 100.0 platelets does not make mandatory the initiation of treatment.

Monocytopenia, a distinctive characteristic of classical HCL was absent in this case since in every blood film a few monocytes were always present. Perhaps this is the explanation why this lady with long-term severe neutropenia did not present any significant infections.

Although the diagnosis was clear, the meticulous search of peripheral blood film for abnormal "hairy" lymphocytes was unfruitful. Indeed, HCL often does not have circulating hairy lymphocytes and the diagnosis is made on bone marrow aspiration or, more often, on bone marrow biopsy, since aspiration is often "dry tap".

END OF STORY

The patient was treated with rituximab and pentostatin. The spleen regressed but cytopenia persists until now, 3 years later. The patient is treated with erythropoietin and transfusions of red blood cells regularly every 2 months. She fiercely denies further treatment, claiming that she leads an almost normal life. She also has skeletal problems attributed mainly to the car accident and severe osteoporosis, but the role of HCL cannot be excluded. She still has no infections at all.

The patient confessed that she knew she had a kind of blood cancer at the time of the accident but she kept it secret from her family fearing she would receive chemotherapy and lose her hair!

Conflict of Interest: None.