

Acute Onset Symptomatic Polycythaemia Vera

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Letter to Editor

Polycythaemia vera (PV) is a myeloproliferative neoplasm (MPN) that is chronic and insidious in its evolution. The diagnosis of MPNs is based on criteria set out by the Polycythaemia Vera Study Group (PVSG) and the WHO^{1,2,3,4,5}. These diagnostic criteria are directed primarily at the exclusion of other myeloproliferative neoplasms (MPN) and of other clinical conditions that could account for the cytopenias. It is well documented that iron deficiency could mask the presentation of PV, but I here present a case of PV that was masked by a low grade lymphoma and presented acutely within 24 hours of a splenectomy in a dramatic fashion.

A 70 year old lady presented with the following blood count : WBC $17.2 \times 10^9/l$ (L $12.3 \times 10^9/l$, N $3.28 \times 10^9/l$, M $0.72 \times 10^9/l$, E $0.16 \times 10^9/l$, B $0.06 \times 10^9/l$) Hb 12.4 g/l, MCV 86.5 fl, Platelets $172 \times 10^9/l$, Hct 38.85 %. The marrow aspirate revealed 94% infiltration by small lymphocytes which were CD 20 / FMC 7 positive but CD5 /CD23 negative and exhibited kappa light chain restriction. Residual trilineage haemopoiesis was markedly reduced. Radiological splenomegaly was detected. A diagnosis of marginal zone B cell lymphoma was made. The patient developed progressive symptomatic splenomegaly even though the blood count did not significantly change. She was treated with one cycle of fludarabine/cyclophosphamide. Although the blood count normalised there was no effect on the massive splenomegaly. So, a splenectomy was performed. Histology of the spleen showed diffuse expansion of the red pulp by an infiltrate of small lymphoid cells, the immunohistochemistry being consistent with splenic marginal zone lymphoma.

In the first postoperative week despite thromboprophylaxis with enoxaparin, the patient developed a pulmonary embolism, splenic and portal vein thrombosis. The post operative blood count was WBC $25.9 \times 10^9/l$ (N $16.7 \times 10^9/l$, L $6.04 \times 10^9/l$, M $1.82 \times 10^9/l$, E $1.13 \times 10^9/l$, B $0.15 \times 10^9/l$) Hb 14.4g/l, MCV 63.2 fl, Hct 46.4%, Platelets $1492 \times 10^9/l$. The blood count changes were attributed to a post splenectomy redistribution effect. She received 6 months of warfarin switching to aspirin as the thrombocytosis persisted.

One year later, the patient presented with a new pulmonary embolism. The blood count at this stage was WBC $40 \times 10^9/l$ (N $26.6 \times 10^9/l$, L $8.45 \times 10^9/l$, M $2.4 \times 10^9/l$, E $1.7 \times 10^9/l$, B $1.12 \times 10^9/l$), Hb 16.7g/l, Hct 57.9%, MCV 62 fl, Platelets $1663 \times 10^9/l$. The bone biopsy showed residual patchy low grade infiltration with lymphoma (10-20% overall). However, the sample was hypercellular with marked increase of megakaryocytes and hyperplasia of both erythroid and granulocytic lineages. There was grade I reticulin fibrosis. The serum ferritin was 17 ng/ml, the BCR/ABL was negative but the JAK 2 mutation was detected. A diagnosis of polycythaemia vera was made. After being managed with venesections, hydroxyurea and rivaroxaban, there was a dramatic relief of the pruritus the patient had had over the previous months.

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Erythrocyte microcytosis is a common feature in overt untreated PV attributed to the depletion of iron stores consequent on the augmented erythroid activity in the marrow. In fact, the MCV normalised (85 fl) as did the serum ferritin (125 ng/ml) just two months after suppressing the marrow with hydroxyurea.

It is obvious the myeloproliferative neoplasm became clinically apparent immediately after the spleen had been removed. However, it did not fulfil even the recently revised WHO diagnostic criteria for a definitive diagnosis in the context of the splenectomy at the time. Cognizant of the biology of MPNs, it is plausible to deduce that the MPN had been present for a number of years before the splenectomy but was completely masked by the concurrent low grade B cell lymphoma. Molecular confirmation of the diagnosis was made after one year only because of the persistence of dramatic clinical features. Awareness of the possibility of "Acute onset" MPN post splenectomy would encourage a more molecular investigative approach that might avert the serious complications of the MPN. However, even if these investigations were done in the early post splenectomy period, it may have been difficult to distinguish between JAK 2-mutated essential thrombocythaemia(ET) and masked PV.

5. Barbui T, Thiele J, Vannucchi AM, Tefferi A. Rationale for revision and proposed changes of the WHO diagnostic criteria for polycythaemia vera, essential thrombocythaemia and primary myelofibrosis. *Blood Cancer Journal* (2015) 5, e337.

REFERENCES

1. Murphy S, Illand H, Rosenthal D & Laszlo J. Essential thrombocythaemia: an interim report from the Polycythaemia Vera Study Group. *Seminars in Haematology*. 1986,23,177-182.
2. Thiele J, Kvasnicka HM. A critical appraisal of the WHO classification of the chronic myeloproliferative disorders. *Leuk Lymphoma*. 2006; 47:381-396.
3. Vardiman JW, Brunning RD, Harris NL. WHO histological classification of chronic myeloproliferative diseases. In: Jaffe ES, Harris NL, Stein H, Vardiman JW, editors. World Health Organisation Classification of tumours of the Haemopoietic and Lymphoid Tissues, Lyon: International Agency for Research on Cancer (IARC) Press; 2001. Pp 17-44.
4. Pearson TC, & Messinezy M. The diagnostic criteria of polycythaemia rubra vera. *Leukaemia & Lymphoma*. 1996, 22 (suppl. 1), 87-93.