Myelomegakaryoblastic Transformation of Essential Thrombocythemia

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Introduction

Essential thrombocytherrlia, considered one of the least common of the myeloproliferative disorders[1], is being diag nosed more frequently with the increasing use of automated blood cell counters[2]. Essential thrombocythemia described by Epstein and Goendel in 1934[3] is a diagnosis of exclusion i.e., exclusion of all sy stemic conditions associated with thrombo- cytosis in an individual with a persistently elevated platelet count of more than $600x10^9$/L (600,000/ul). Prognosis of essential thrombocytherrlia is excellent, with a median sur- vival in excess of 10 years[4]. However, 10 percent of cases undergo blastic transformation.

A case of essential thrombocythemia presenting in myelomegakaryoblastic transfomlation is reported, due to its rare occurence and presentation.

Case Report

A 34 years old female presented in early 1995 with symptoms of loose motions, intermittent mild abdominal pain and malaise for the previous 2 years. The past history was significant for hemorrhoidectomy 1 1/4 years back and at that time her platelet count was $l000x10^9$/L ($1000x10^3$/ul); and low grade intermittent fever lasting for 2 to 3 months, six months ago.

At presentation the significant positive physical findings included pallor, splenomegaly 5 cm and hepatomegaly 4 cm below the respective costal margins.

Her hemoglobin was 83 gm/L (8.3 gm/dl), WBC 19. $1x10^9$/L (19.1 x 10 3/ul), Platelets $2200x10^9$/L ($2200x10^3$/ul), neutrophils 26%, lymphocytes 37%, stab/band cells 6% and blast cells 31%; 16% blast cells were morphologically myeloblasts, sudan black B negative and 15% blast cells were megakaryoblasts, acid phosphatasé, PAS and platelet glycoprotein lilA (CD 61) positive. Leucocyte Alkaline Phosphatase score was in the high normal range. Red blood cell morphology showed mild aniso- poikilocytosis, polychromasia, basophilic stippling and an occasional tear drop cell and normoblast. The platelets were markedly increased in the peripheral blood film, were in multiple large clrnps and showed giant forms anisopoikilocytosis and bare megakaiyocytic nuclei.

The bone marrow aspiration was difficult and yielded only a small amount of aparticulate marrow showing 37% blasts: 15% sudan black B negative myeloblasts and 22% acid phosphatase positive megakaryoblasts, increased megakaryocytes and depressed eiythropoiesis and myelopoiesis.

Bone marrow trephine biopsy showed a hypercellular marrow with increased megakaiyocytes present in clusters and sheets, normal myeloid etythroid ratio and focal minimal to mild fibrosis.

Anti-nuclear antibody was negative. Chromosomal study for Philadelphia chromosome was negative.

Discussion

Essential thrombocythemia, a myeloproliferative disorder, is less frequent as compared to chronic myeloid leukemia. It occurs predominantly in an older age group; however, a second population of younger patients, predominantly females, is also presents[5]. In most cases it follows a benign course,

only 10% transform into a blastic crisis[2]. Our case illustrates the point made by Kimura et al. that essential thrombocythemia is a myeloid multipotential stem cell disorder and in the event of transformation, blasts can be of any one cell type or a combination of two or all the three cell lines arising from the multipotential stem cell i.e. erythroblasts, myelomonoblasts and/or megakaryoblasts[6].

References

1. Jandl, J.H. Blood; Textbook of Hematology (1st edition)Boston/Toronto, Little, Brown and Co., 1987, p. 709.
2. Williams, W.J., Beutler, E., Erslev, A.J. et a). Hematology. 4th edition, New York, McGraw Hill, 1991, p. 234.
3. Epstein, E. and Goode), A. Haemorrhagische thrombocythamie bei vascular schrumpfmilz. Virchows Arch. (A)., 1934;293:233.
4. Murphy, S. Thrombocytosis and thrombocythemia. Clin Haematol., 1983;1 2:89.93.
5. Bellucci, S., Janvier, M., Tobelem, G. et al. Essential thrombocythemias. clinical Evolutionary and biological data. Cancer, 1986;58:2440-2485.
6. Kimura. A., Fujimoto, T., Inada, T. et al. Blastic transformation in essential thrombocythemia. In Vitro differentiation of blast cells into granulocytic, erythroid and megakatyocytic lineages. Cancer, 1990;65 :1538-44.