Myelomegakaryoblastic Transformation of Essential Thrombocythemia

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Introduction

Essential thrombocythemia, considered one of the least common of the myeloproliferative disorders\(^1\), is being diagnosed 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 systemic conditions associated with thrombocytosis in an individual with a persistently elevated platelet count of more than \(600 \times 10^9/L\) (600,000/ul). Prognosis of essential thrombocythemia is excellent, with a median survival in excess of 10 years\(^4\). However, 10 percent of cases undergo blastic transformation.

A case of essential thrombocythemia presenting in myelomegakaryoblastic transformation is reported, due to its rare occurrence 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 \(1000 \times 10^9/L\) (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 phosphatase, PAS and platelet glycoprotein liI (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 clumps and showed giant forms anisopoikilocytosis and bare megakaiyocytic nuclei.
The bone marrow aspiration was difficult and yielded only a small amount of apatriculate marrow showing 37% blasts: 15% sudan black B negative myeloblasts and 22% acid phosphatase positive megakaryoblasts, increased megakaryocytes and depressed eithyropoiesis and myeloipoiesis. Bone marrow trephine biopsy showed a hypercellular marrow with increased megakaiyocytes present in clusters and sheets, normal myeloid erythroid 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 present\(^5\). 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