Response to STI571 in Chronic Myelomonocytic Leukemia with Platelet Derived Growth Factor Beta Receptor Involvement: A new case report

Based on its ability to inhibit the tyrosine kinase activity of ABL, as well as the c-kit and the Platelet Derived Growth Factor Receptor tyrosine kinases, the spectrum of diseases that may respond to STI571 is increasing. A recently recognized subgroup of myeloproliferative disorders/myelodysplastic syndromes (MPD/MDS) has a t(5;12)(q33;p13) with the activation of the gene for PDGFRB which encodes a receptor tyrosine kinase. Here, we present the case of a patient, with MPD/MDS, and eosinophilia, carrying a translocation t(5;12)(q33;p13) who achieved a complete remission following treatment with STI571, 400 mg daily. At the time of writing he still remains in complete remission with an excellent performance status. There is clearly a need for further studies of STI571 in MPD/MDS with chromosomal translocations involving PDGFRB to confirm this promising initial result.

Recent cytogenetic and molecular studies suggest the existence of some new single subset of patients with Myeloproliferative Disorders with defined cytogenetic abnormality. The most common abnormality is the t(5;12)(q33;p13),1 which fuses the ETV6/TEL gene to the hybridization and reverse transcriptase at the molecular level have been examined. Here, we present the case of a patient with MPD/MDS, and eosinophilia, carrying a translocation t(5;12)(q33;p13) who achieved a complete remission following treatment with STI571, 400 mg daily. At the time of writing he still remains in complete remission with an excellent performance status. There is clearly a need for further studies of STI571 in MPD/MDS with chromosomal translocations involving PDGFRB to confirm this promising initial result.

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Figure 1. G-banded karyotype of bone marrow cells showing 46, XY, t(5;12)(q33;p13).
typical features of myeloproliferative disorders with translocation t(5;12)(q33;p13), involvement of PDGFRB and response to STI571 treatment. Interestingly, all cases are male, hypereosinophilia, monocytosis and splenomegaly appear as characteristic features. In view of the availability of STI571, a potent inhibitor of several tyrosine kinases, there is clearly a need for further studies of STI571 in this setting to confirm these promising initial results.

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References