Background: The 2008 WHO classification of MPNs includes the category "Myeloid and lymphoid neoplasms associated with eosinophilia and abnormalities of PDGFRA, PDGFRB, or FGFR1"—rare entities united by constitutive overexpression of a tyrosine kinase gene product and by substantial and durable clinical responses to tyrosine kinase inhibitors. We describe the clinical, pathologic and cytogenetic features and follow-up of our patient with CMPD associated with t(5;12)(q33;p13). Methods and results: A 19 year old man was sent to our department with suspicion of chronic myeloid leukemia on November 2010. CBC showed a significant leukocytosis with a prominent left shift, mild anemia, and normal platelet count. Notably, there was no eosinophilia or monocytosis. The peripheral blood was negative for BCR-ABL gene rearrangement and JAK2 mutation. No hepatosplenomegaly was detected. The bone marrow biopsy specimens were hypercellular with marked myeloid hyperplasia. Conventional cytogenetic studies were performed on bone marrow aspirate samples and t(5;12)(q33;p13) was confirmed. Additional cytogenetic aberrations were not identified. Fluorescent in situ hybridization (FISH) for rearrangement of the PDGFRB gene showed separation of the 5' and 9' PDGFRB signals in 78% of nuclei. On the basis of the morphologic findings, cytogenetic changes with t(5;12)(q33; p13), and FISH analysis (ETV/PDGFRB), patient was classified as CMPD with t(5;12)(q33;p13), ETV/PDGFR beta. The treatment has begun with imatinib at a daily dose of 400 mg orally. The patient achieved complete hematologic and cytogenetic remission after 3 months and molecular (MR) response after 6 months since the start of imatinib treatment and has remained in MR for 44 months to the time of the last control (June, 2014). Conclusion: We describe the patient who exhibited features that are somehow unusual for this type of MPN, namely, lack of eosinophilia and splenomegaly. Because recent studies have shown that CMPD with t(5;12) can respond to imatinib mesylate, recognition of this neoplasm has important clinical implications.