Chronic myelomonocytic leukemia developed 2 years after the onset of immune thrombocytopenic purpura like syndrome

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Abstract—An 80-year old man was diagnosed as having immune thrombocytopenic purpura based on epistaxis, purpura and by the platelet count \(8 \times 10^9/1\). Prednisolone and gamma globulin were administered and the platelet count had been kept around \(50 \times 10^9/1\) during his follow up. Two years from the onset of immune thrombocytopenic purpura he was admitted because of leukocytosis \((79 \times 10^9/1\) with 79% monocytes), anemia and thrombocytopenia. Hypercellular bone marrow with dysplasia of three lineages was observed. In the bone marrow cytogenic analysis, a -6, clonal cytogenic abnormality was observed, 45XY, der(6), t(6;6)(q16;q23). He was diagnosed as having chronic myelomonocytic leukemia. This is a difficult case in which it was diagnosed as refractory thrombocytopenia as a subgroup of myelodysplastic syndrome, rather than immune thrombocytopenic purpura, which might have preceded the development of chronic myelomonocytic leukemia.

Key words: Chronic myelomonocytic leukemia; immune thrombocytopenic purpura.

INTRODUCTION

Both immune thrombocytopenic purpura (ITP) and myelodysplastic syndrome (MDS) have a typical concept in terms of hematopoietic disorders, but there can be difficulty encountered in differentiating ITP from MDS because of both causing megakaryocytic hyperplasia and thrombocytopenia. The most common cytopenias in MDS are, in decreasing order of prevalence, monocytopenias, bicytopenias, and pancytopenia [1]. Of the monocytopenias, anemia is the most common. However, thrombocytopenia may be presenting as a cytopenia of MDS and is categorized as
refractory thrombocytopenia (RTC) [2–4]. The morphologic identification of MDS presenting initially as thrombocytopenia is difficult because morphologic features of dysplasia may not be evident and megakaryocytic hyperplasia may be confused with ITP [5]. So, it is important to keep in mind RTC as the cause of an isolated thrombocytopenia on the basis of clinical history and laboratory testing to avoid the inappropriate therapy. Here, we report on one case whom developed chronic myelomonocytic leukemia (CMMoL) after two years after the onset of RTC.

PATIENT AND METHODS
An 80-year old man was diagnosed as having ITP from his symptoms of epistaxis, purpuric lesions, and a low platelet count of $8 \times 10^9$ /l without anemia or leukopenia. Other laboratory values including hepatic and renal function tests, antibody titers against rubella, toxoplasmosis, CMV, HSV, EBV, HCV, HIV and HBV, as well as the VDRL test, Melitensis, Wright and Gruber Widal agglutinations, prothrombin and partial thromboplastin times were all within normal limits. Examination of his bone marrow smears revealed a prominent increase of micromegakaryocytes in normocellular bone marrow without dyserythropoiesis or dysgranulopoiesis. Platelet associated IgG and serum anti-platelet antibodies were not assessed due to technical limitations. The patient when questioned gave no indication that there was any history of familial thrombocytopenia or drug administration known to be potentially causing the thrombocytopenia and the patient was not in a high risk category for HIV infection. Prednisolone and gamma globulin were administered and the platelet count had been kept around $50 \times 10^9$ /l during his follow up. A splenectomy was suggested but he refused this intervention. After two years from the onset of ITP, he was admitted because of epistaxis, dyspnea, and precordial pain. The white cell count was $79 \times 10^9$ /l with 79% monocytes, 9% lymphocytes, 9% myeloblasts, 3% normoblasts. The hemoglobin was 5.6 g /dl and the platelet count was $14 \times 10^9$ /l. The peripheral smear revealed small and large red cells with pale cytoplasm and polychromasia; basophilic stippling and dysmyelopoiesis were also noted. Hypercellular bone marrow with dysplasia of three lineage’s such as dyserythropoiesis, Pelger-Huet abnormality, and micromegakaryocytes were observed. The cytogenetic analysis of bone marrow cells revealed 45XY, der(6), t(6;6)(q16;q23), -6, clonal abnormality. The patient was diagnosed as having CMMoL and received low dose Ara-C. Due to a sudden increase in white blood cell count ($103 \times 10^9$ /l) with 52% myeloblasts observed under low dose Ara-C, the therapy was switched to mitoxanthrone 12 mg/m² 1, 2, 3, days and etoposide 100 mg/m² 5 days. He is still in remission one year into the follow-up period.

DISCUSSION
The original FAB classification of 1982 focuses mainly on refractory anemia (RA), and thrombocytopenia or leukopenia without anemia have also been classified