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Letters to the Editor

Abolishment of Pleural Effusion as the Initial Manifestation of Chronic Myelomonocytic Leukemia without Chemotherapy

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To the Editor

A 66-year-old male was admitted to our hospital with chest pain. The white blood cell count was 15.8 x 10^9/L and hemoglobin was 12.0 g/dL. A peripheral blood smear revealed 22.0% mature monocytes and 0.5% promonocytes. A bone marrow examination revealed increased mature monocytes (18.4%) without an increase in immature cells (monoblast : 0.4%, promonocyte : 0.4%). Chromosome analysis using bone marrow blood revealed a normal karyotype. Chest computed tomography and X-ray photography showed right pleural effusion with the following features : cell count, 2.0 x 10^9/L (mature monocytes : 56.5%, immature monocytes 1.0%) ; protein, 4.9 g/dL ; LDH, 54 U/L and glucose 142 mg/dL, which suggested exudates. The phenotypes of the monocytes in the peripheral blood, the bone marrow, and the pleural effusion were identical ; cells were positive for CD13, CD14, CD33, and HLA-DR and negative for CD2, CD10, CD19, and CD34. The patient was diagnosed with chronic myelomonocytic leukemia (CMML) and leukemic cell infiltration seemed to have caused the pleural effusion. Because immediately life-threatening symptoms were absent, the patient was followed up as an outpatient without chemotherapy. The pleural effusion decreased after 6 months and then disappeared within one year, but monocytosis and mild anemia persisted.

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The French-American-British (FAB) classification has categorized CMML as a subtype of myelodysplastic syndrome (MDS). The 1990 WHO classification re-categorized CMML as myelodysplastic/myeloproliferative disease (MDS/MPD) and classified it into CMML-1 and CMML-2 according to the blast ratio.[1] One report indicated that MPD-like CMML was associated with increased blasts in the peripheral blood, manifested as dysplasia, and was frequently associated with hepatosplenomegaly and adenopathy.[2] Furthermore, leukemic cell invasion could be combined pleural effusion, ascites, and pericardiac effusion in this type of CMML.[2] Such pleural effusion usually occurs in aggravated CMML,[3-5] but is extremely rare at the onset of CMML as in our patient.[6]

Although treatments with hydroxyurea, 6-mercaptopurine and low dose cutaneous cytosine arabinoside have been met with some success, the most effective agent remains obscure and an optimal management strategy for CMML has not been established. Asymptomatic CMML patients with slight monocytosis should remain under observation without treatment. As to pleural effusion of CMML, steroid pulse therapy and low doses of etoposide are effective.[7,8] However, when should we start chemotherapy for CMML if the pleural effusion is accumulated? In our case, the monocytes, which manifest the same phenotype, were observed in the pleural effusion as well as in the bone marrow and peripheral blood. These results suggest the pleural effusion which we observed in this case might be associated with the involvement of CMML, whereas the pleural effusion spontaneously disappeared in the space of a year without undergoing chemotherapy. Controlling pleural effusion and/or ascites does not necessarily improve the prognosis. Prognostic factors of CMML have not been clearly delineated, but Fenaux *et al*. reported that prognosis could be anticipated from standard parameters at diagnosis : namely, bone marrow blasts, hemoglobin and blood monocytosis.[6] Actually, almost all patients

with disease that transforms into AML die early. Thus, Voglova *et al*. proposed that patients should be monitored while they lack other signs of myeloproliferation such as peripheral immature granulocytes, splenomegaly, lymphadenopathy, skin involvement and pleural or peritoneal effusion,[2] suggesting that the myeloproliferative features of CMML have an important impact on the choice of treatment. Pleural effusion without respiratory failure might not necessarily require an active treatment in CMML. Data regarding patients with CMML should therefore be accumulated, and prognostic factors should be defined before appropriate treatment strategies can be agreed upon.

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