We report a 63-year-old woman with myeloproliferative neoplasm (MPN), who presented with inflammatory involvement of the skin. The white cell count was 31,800/μL and the platelet count was 709×10^3/μL. The positivity of neutrophil alkaline phosphatase was 100% and the score was 281. The bone marrow was hypercellular with enlarged megakaryocytes with mature cytoplasm and multilobulated nuclei (Picture 1A; May-Giemsa staining, ×40 objective). The myeloid-lineage cells contained coarse toxic granules (Picture 1B; May-Giemsa, ×100), probably reflecting the cutaneous complication, while they were entirely negative for peroxidase activity except for eosinophils (Picture 1C; peroxidase staining, ×100). Peripheral blood pictures confirmed the lack of activity of monocytes (Picture 1D; peroxidase, ×100). On the other hand, the periodic acid-Schiff reaction and naphthol AS-D chloroacetate esterase positivity of neutrophils as well as the α-naphthyl butyrate esterase positivity of monocytes were normal. The patient has been treated with hydroxyurea for cytoreduction.

Primary myeloperoxidase (MPO) deficiency is caused by...
a mutation within the MPO gene, while the expression of eosinophil peroxidase, the gene of which is independent of MPO, is normal (1, 2). It remains to be determined whether the association of MPO-deficiency with MPN in the current patient is coincidental or whether there is a causal relationship between the two conditions.

References