A 45-year-old woman was admitted with fatigue, icterus, thrombocytopenia (77 × 10⁹/L), normocytic anemia (9.8 g/dL), and a normal white blood cell count (8.5 × 10⁹/L). The peripheral blood smear showed metamyelocytes, myelocytes, monocytes, erythroblasts, and atypical cells with bilobulated nuclei resembling basophil granulocytes (left panel of figure). In the bone marrow there was a nearly complete infiltration with atypical, partly degranulated mast cells expressing surface markers (CD2, CD25, and CD117; right panel of figure). These cells were also positive for c-KIT mutation (D816V). Tryptase levels in serum were elevated (719 μg/L, normal value: 0-11.4 μg/L) in the absence of symptoms arising from mediator release.

A diagnosis of aleukemic variant of mast cell leukemia was made because < 10% mast cells could be detected in peripheral blood. She had no skin lesions. Four weeks after starting treatment with high-dose chemotherapy, the patient died with *Klebsiella pneumonia* sepsis and multiorgan failure. Mast cell leukemia is a rare and very aggressive form of systemic mastocytosis. The prognosis is poor with a median survival of < 2 months.
Aleukemic variant of mast cell leukemia
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