A 32-year-old man presented at the hematology department with bilateral testicular enlargement and multiple, painless, purple cutaneous nodules on both upper extremities and the trunk (Figure 1). He had been diagnosed with acute promyelocytic leukemia (APL) 7 years earlier. Complete remission was achieved with all-trans-retinoic acid (ATRA) and idarubicine therapy, and maintenance therapy had continued for 2 years since remission.

Complete blood count and biochemical test results were normal, and peripheral blood smear (PBS) revealed no specific features. Bone marrow biopsy was performed, and did not demonstrate any leukemic infiltration. Real time polymerase chain reaction (RT-PCR) assay to detect t(15;17) (PML-RAR alpha) in blood samples was negative. Skin biopsy, however, revealed acute myeloid leukemic infiltration. Skin sample evaluated using RT-PCR for t(15;17) was positive. Bilateral, heterogeneous, hypervascular, irregular limited area was detected with testicular ultrasonography, indicative of testicular infiltration. Two weeks after confirmation of diagnosis, increase in D-dimer (>5000 μg/mL) and hypofibrinogenemia had developed. At the start of induction therapy, idarubicin (12 mg/m²/day, D2, 4, 6, 8), ATRA (45mg/m²), arsenic trioxide (ATO) (0.15 mg/kg/day for 22 days) was administered and methylprednisolone was added for ATRA syndrome prophylaxis. On the 20th day of therapy, after observing leukocytosis in his blood count, PBS and flow cytometry were reperformed. PBS demonstrated promyelocytes, and flow cytometry showed cell population CD13, CD33 positive, CD14 dim positive, and CD34 and HLA-DR negative. On the 30th day of therapy, ATRA and ATO therapy was interrupted due to respiratory distress, prolonged QT, and fever. Dexamethasone 20 mg/day and intravenous imipenem 2 g/day was initiated for differentiation syndrome and pneumonia. Bilateral, subpleural consolidation areas; peribronchovascular inflation; and ground glass opacity in the right middle lung zone was seen on tho-

Figure 1. Purple papulonodular lesions on the trunk.
rax computed tomography image. Even after addition of trimethoprim-sulfamethoxazole and amphotericin-B, the patient did not respond to the therapy. Since the patient’s dyspnea was worsening, he was taken to the intensive care unit. On the third day of intensive care unit follow-up, the patient passed away.

Even though the application of ATRA and anthracycline-based chemotherapy has increased complete remission rates to more than 90% and improved survival in newly diagnosed APL, relapse currently occurs in 10% to 15% of APL patients.[1, 2] APL relapse in the bone marrow is common; however, an increasing number of extramedullary (EM) recurrences have also been reported.[3, 4] Central nervous system, skin, testicle, vascular access, external ear, and auditory canal are the most common EM relapse sites.[3–8] Before the ATRA era, the large majority of first relapses in APL occurred within 3 years of complete remission, and only 2% to 3% of patients relapsed after 4 years.[9]

Disclosures
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References