CLINICAL IMAGE

Ear involvement in acute promyelocytic leukemia

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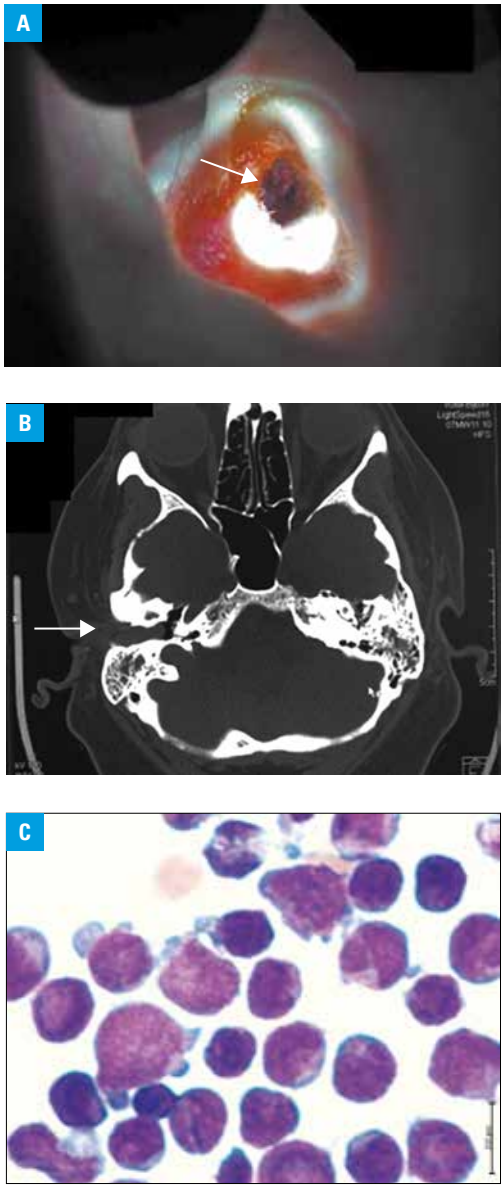

FIGURE 1 **A** – tumor mass in the external acoustic meatus (arrow); **B** – tumor mass in the auditory apparatus (arrow); **C** – leukemic cells in cerebrospinal fluid (Wright–Giemsa staining)

A 44-year-old woman with acute promyelocytic leukemia (APL) presented to our department in an overall bad condition with hearing loss, severe headache, and severe weakness. Two years earlier, she had been diagnosed with APL with 46,XX, t(15;17)(q24;q21), del (16)(q11;q22), add(7)(p21), add (X)(p22), and detectable PML/RARα and FLT3-ITD mutations, and had been successfully treated with the PETHEMA protocol including all-trans retinoic acid (ATRA) with anthracyclines and cytarabine. She had remained in complete remission for 15 months.

We performed laboratory tests, which revealed moderate anemia (hemoglobin, 10.6 g/dl) and leukopenia (3.4×10^9/l) with normal white blood cell differential. Platelet count and coagulation test results were normal. A neurological examination revealed right-sided paresis and nystagmus. An otoscopic examination was remarkable for the masses in the external auditory canals (FIGURE 1A). A biopsy revealed dense infiltration of immature myeloid progenitors. A head computed tomography scan showed tumor infiltrates in the mastoid air cells, middle ear, and external auditory canals (FIGURE 1B). Brain parenchyma was leukemia-free. A bone marrow aspirate demonstrated blast cells and atypical promyelocytes with detectable PML/RARα fusion on reverse-transcriptase polymerase chain reaction. Cerebrospinal fluid contained leukemic cells (FIGURE 1C). The patient received arsenic trioxide and ATRA with intrathecal chemotherapy but died a few weeks later.

APL is a subtype of acute myeloid leukemia characterized by unique chromosomal abnormality, response to ATRA, and disseminated intravascular coagulation.[1] Extramedullary relapse of APL occurs rarely and involves the skin, lymph nodes, and central nervous system.[2] Ear involvement at relapse of APL is extremely rare, and only single cases have been reported to date. It is noteworthy that some patients with ear involvement at relapse of APL may have simultaneous involvement of other sites (eg, bone marrow or

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the central nervous system, as in our patient).[3] Arsenic trioxide therapy is known to be highly effective in newly onset and relapsed APL; however, its efficacy in extramedullary relapse requires further studies.[4] The pathogenetic mechanism of extramedullary relapse in APL remains to be elucidated. The potential role of prior ATRA treatment and its impact on the leukemic cell adhesion to the endothelium and the subendothelial matrix has been reported.[5]

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