

Extramedullary relapse in acute promyelocytic leukemia

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A 71-year-old woman presented with a one-week history of otorrhagia, left hypoacusia and earache having been in remission from acute promyelocytic leukemia (APL) for 17 months after the end of chemotherapy (induction phase with idarubicin 12 mg/m² day 2, 4, 6, 8 and ATRA 45 mg/m² day 1->30, then only one consolidation course of therapy with idarubicin 5 mg/m² day 1-> 4 and ARA-C 1 g/m² day 1->4 and maintenance therapy with ATRA at the dose of 45 mg/m² day for 15 days every 3 months during a period of 2 years), according to the GIMEMA AIDA protocol.

Otoscopic examination showed hyperemic tumor masses completely occluding the external left auditory canal.¹ On admission, her blood counts showed a hemoglobin level of 14.5 g/dL, a WBC count of 7.2x10⁹/L (normal blood smear) and a platelet count of 234x10⁹/L, with a normal coagulation profile. A computed tomographic (CT) scan showed contrast-enhancing masses in the left auditory canal with complete opacification of the mastoid air cells (Figure 1). A superficial biopsy showed extensive infiltration by sheets of promyelocytes (Figure 2), with characteristic granules and Auer rods and strong staining for myeloperoxidase (Figure 3), which was indicative of an extramedullary relapse of acute promyelocytic leukemia. However, a bone marrow examination showed complete morphologic remission, confirmed karyotypically and molecularly. The patient was successfully treated with local radiotherapy and chemotherapy (methotrexate 15 mg/m² weekly, 6-mercaptopurine 50 mg/m²). In the literature, there are only a few reports of extramedullary relapse of APL; to our knowledge, this is the first case of ear relapse of APL in which reverse transcriptase-polymerase chain reaction analysis of the bone marrow was negative for PML-RARalpha fusion transcripts.^{2,3}

References

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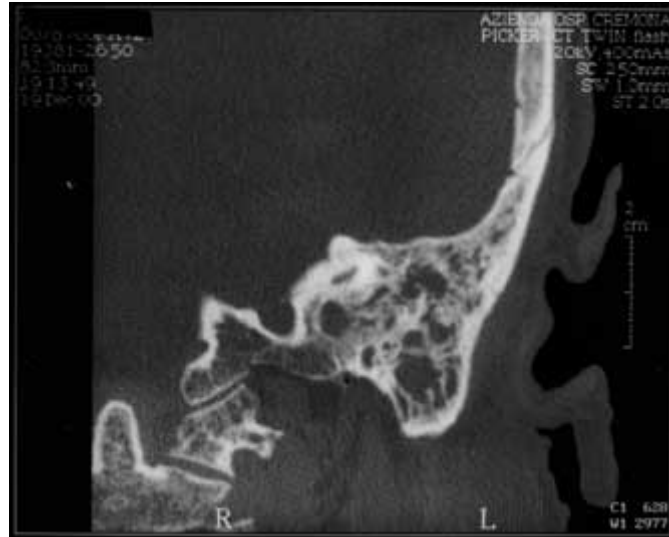


Figure 1: a computed tomographic scan showed contrast-enhancing masses in the left auditory canal with a complete opacification of the mastoid air cells.

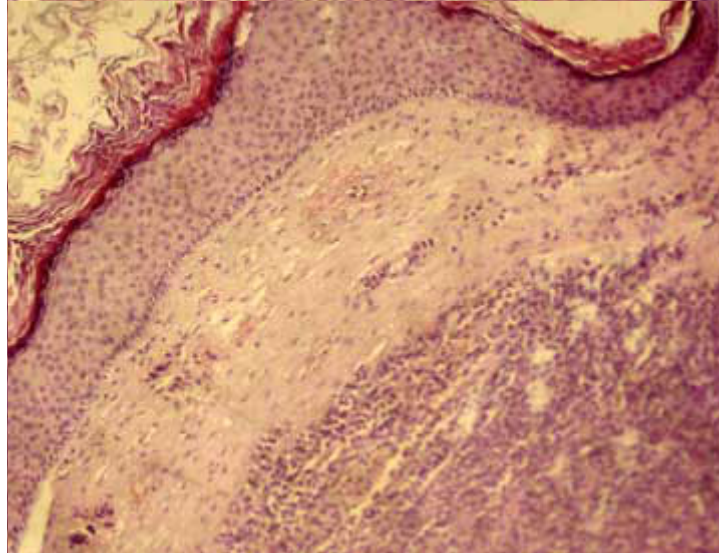


Figure 2: extensive infiltration by sheets of promyelocytes in the biopsy of left auditory canal with characteristic granules.

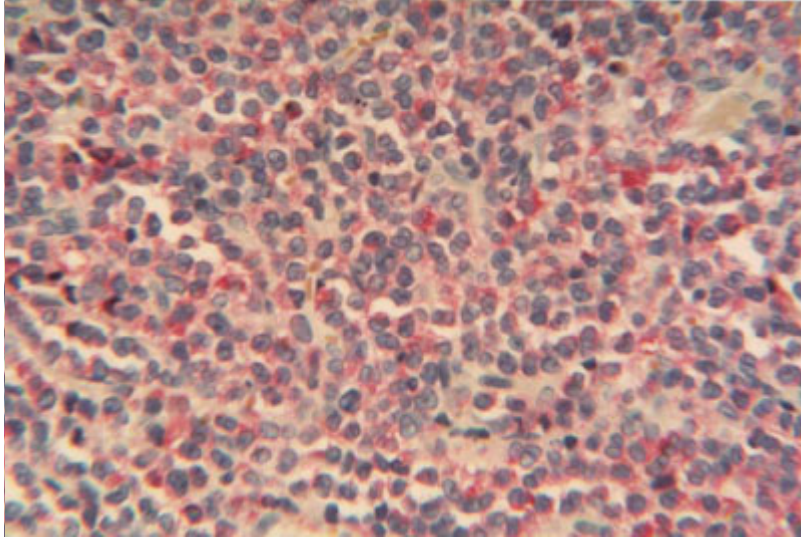


Figure 3: strong staining for myeloperoxidase by promyelocytes.