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Acute promyelocytic leukemia with persistently positive central nervous system involvement: a case report

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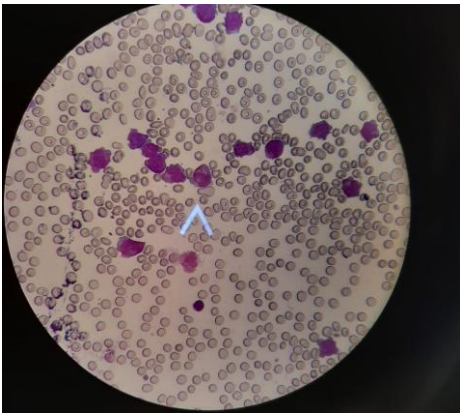
INTRODUCTION:
Central nervous system (CNS) involvement in acute promyelocytic leukemia (APL) is rare and occurs mainly in relapses. Risk factors include high leukocyte count and CNS bleeding at diagnosis. This report seeks to describe the case of an adolescent with APL and a persistently positive CNS involvement.

CLINICAL SUMMARY:
A 17-year-old female sought medical attention for epistaxis and bruises. The initial workout showed anemia, thrombocytopenia, leukocytosis (94.000 - 82% blasts[1]) and coagulopathy. Due to the suspicion of APL, ATRA was started. A subsequent bone marrow analysis confirmed the diagnosis. The patient evolved with hemodynamic instability, disseminated intravascular coagulation, an intraparenchymal bleeding in the CNS and a subdural hematoma.
On day 45 of ATRA she achieved clinical stability that enabled the performance of a lumbar puncture. The analysis of the cerebrospinal fluid (CSF) revealed the presence of 69% blasts, and it was decided to start intrathecal chemotherapy twice a week. Even though morphological and molecular remission of APL was achieved after consolidation I of the CLEHOP protocol without ATO, the CSF remained with positive oncotic cytology. An alternative chemotherapy regimen with high dose cytarabine and doubled dose of intrathecal cytarabine was chosen for follow up treatment. The CSF was cleared of blasts after 3 weeks and the patient was switched to the CLEHOP protocol with ATO for consolidation.



1. Photo of the myelogram, showing the morphology of blasts with Auer rods.

DISCUSSION:
Even though the best way to treat CNS involvement in APL is not well-established, the use of systemic treatment with high dose cytarabine and an optimized intrathecal chemotherapy with doubled dose cytarabine may be an option.