ACUTE PROMYELOCYTIC LEUKEMIA PRESENTING AS
THROMBOTIC CEREBROVASCULAR ACCIDENT

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Abstract - The usual manifestation of acute promyelocytic leukemia is hemorrhagic tendency but thrombotic manifestations may dominate the clinical picture. We report a patient with promyelocytic leukemia which presented as thrombotic cerebrovascular accident and responded to all-transretinoic acid and chemotherapy.

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INTRODUCTION

Acute promyelocytic leukemia (APL) constitutes about 10% of cases of acute myeloid leukemia (AML). Hemorrhagic manifestations are prominent and may be fatal. This disease presents with thrombocytopenia, prolongation of partial thromboplastin time (PTT) and thrombin time (TT), increased level of fibrinogen degradation product (FDP) and hypofibrinogenemia.

Coagulopathy associated with APL results from at least three distinct mechanisms: DIC, fibrinolysis and proteolysis due to release of procoagulant activities, plasmogen activators and lysosomal neutrophil enzymes.

Trombin activators may lead to diffuse intravascular coagulation, whereas leukocyte mediated proteolysis is able to clear various substance including fibrinogen, in vitro and in vivo (1-3).

CASE REPORT

Our patient was a 16 year-old girl presenting with fever, disorientation, and subacute right-sided hemiplegia, aphasia, urinary incontinence and vaginal bleeding.

Fifteen days prior to admission she had been evaluated in another center for dizziness and right-sided hemiparesis and had been discharged from the hospital because of the improvement of her symptoms.

The first laboratory results showed leukopenia (WBC = 1700/ml), anemia (hemoglobin: 7.5mg/dl), thrombocytopenia (platelet: 118000/ml), prolonged PT and PTT, increased FDP (FDP = 10) and normal fibrinogen (fibrinogen = 397).

On brain MRI, high intensity areas were seen in the left paraventricular region that were suggestive of thrombosis (Fig. 1).

On bone marrow study APL with hypergranular cells were reported and the results of flow cytometry were as follows CD3(+) CD33 (+), MPO (+) and HLA-DR(-). The patient was treated with all-transretinoic acid (ARTA) 45mg/m2. Ten days after initiation of ARTA, DIC was controlled. Because of increment in white blood cells count (>20,000/mm3), cytarabine and idarubicin were administered to induce a remission. After chemotherapy, FDP increased to 80 and fibrinogen dropped to 181. DIC was ultimately controlled and consolidation therapy was then started.

Brain CT-scan (Fig. 2), revealed that previous lesions had resolved following treatment. The patient was discharged with mild right-sided hemiparesis and resolution of the aphasia.

DISCUSSION

Review of the from 1988 to 1997 revealed only one reported case of APL presenting as thrombotic CVA in Japan (4). Several cases of APL presenting as migraine with aura have been reported.

The usual intracranial presentation of APL is intracerebral hemorrhage with fatal outcome, especially at the beginning of chemotherapy (6) but our patient presented with DIC and a thrombotic tendency. The major mechanism of coagulopathy in our patient was DIC, instead of fibrinolysis. DIC may be the predominant mechanism of coagulopathy. Although thrombotic events such as arterial occlusions, pulmonary emboli, hepatic vein occlusion and portal vein occlusion are very infrequently identified clinically (7-11), post mortem examinations show widespread thrombosis in 12-25% of patients (10-13).
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Fig. 1- Brain MRI at presentation

Fig. 2- Brain CT Scan after treatment

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REFERENCES


