Bad Blood: A Catastrophic Presentation of Acute Promyelocytic Leukemia

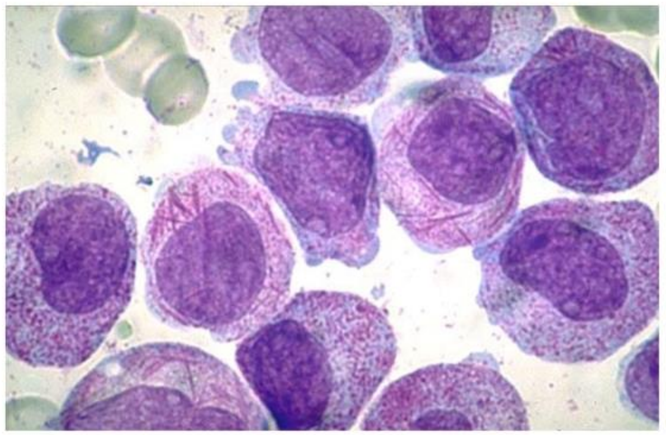
Sarah Bentil-Owusu, Vanessa Soetanto, Hyein Jeon, Joshua Kra

Department of Internal Medicine, Rutgers University, Newark, New Jersey 07103

Learning Objectives

1. Recognize intracranial hemorrhage as a fatal and often overlooked complication of Acute Promyelocytic Leukemia (APL)

2. Treat patients with high suspicion for APL with immediate administration of all-trans retinoic acid (ATRA) as a potential life-saving treatment



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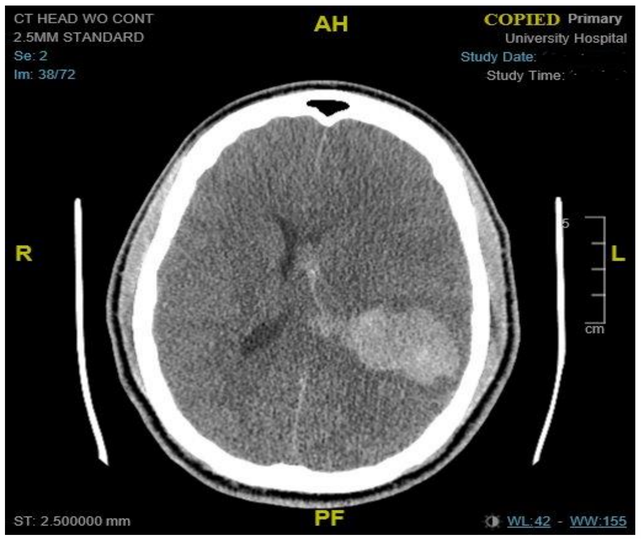
Hospital course

Peripheral smear showed hypergranular promyelocytes without Auer rods. Despite initiation of all-trans retinoic acid (ATRA), continued administration of blood products, hypertonic solutions, and initiation of pressors, patient clinically deteriorated and was terminally extubated after pronouncement of brain death. Postmortem, FISH confirmed the diagnosis of APL with translocation of PML (chr 15) and RARA gene (chr 17).

Case

A 26-year old Haitian man presented with 3 days of acute, left-sided, non-radiating headache with no known exacerbating or remitting factors. His headache worsened 5 hours prior to presentation with nausea, vomiting, photophobia, and phonophobia. He denied fever, neck stiffness, visual changes, recent travel or sick contacts. Labs were significant for WBC 1.1 x10 /µL, Hgb 7.7 g/dL, Plt 8 x10 /µL, elevated d-dimer, LDH 315 U/L, INR 1.7. Exam was notable for a diaphoretic man in acute distress, sluggishly reactive pupils and right-sided diminishment to light touch.

CT Head showed active hemorrhage at the left posterior parietal lobe with 3 mm midline shift. Repeat exam revealed a newly fixed and dilated left pupil with repeat CT showing worsening intraparenchymal hematoma with an increased midline shift requiring an emergent decompressive hemicraniectomy.



Impact and Discussion

APL – a distinct subset of acute myeoid leukemia with presence of increased promyelocytes was first described in 1957 in patients with severe bleeding and fibrinolysis with rapid deterioration of their clinical condition. Advances led to the understanding of the pathognomonic translocation between genes on chr 15 and 17 (PML-RARA) that causes developmental arrest at the promyelocytic stage. ATRA is highly effective at releasing this block leading to maturation of the leukemic cells.

Prompt exploration of pancytopenia and immediate intervention for unremitting headache is crucial. Despite effective treatment, hemorrhagic complications account for the major cause of morbidity and mortality, due to increased thrombin generation, activation of coagulation, and abnormal fibrinolysis. Early introduction of ATRA at the first suspicion of a diagnosis of APL can prevent these complications in most cases and should be administered even prior to confirmation of the diagnosis.

Conclusion

1. This case illustrates the importance of prompt diagnosis and recognition of hematological malignancies, specifically APL.

2. Despite the curable nature of APL with ATRA, early mortality is still prevalent due to bleeding complications