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Clinical Case study presentation

BAD BLOOD: A CATASTROPHIC PRESENTATION OF ACUTE PROMYELOCYTIC LEUKEMIA

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Case: A 26-year old 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. Exam was notable for a diaphoretic man in acute distress, sluggishly reactive pupils and right-sided diminishment to light touch. Labs were significant for pancytopenia, elevated d-dimer, LDH 315 U/L, INR 1.7. CT Head showed active hemorrhage at the left posterior parietal lobe with 3 mm midline shift. He became more somnolent and confused, prompting intubation. Repeat exam revealed a newly fixed and dilated left pupil with repeat CT Head showing worsening intraparenchymal hematoma with an increased midline shift requiring an emergent decompressive hemicraniectomy. 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).

Impact/Discussion: Prompt exploration of pancytopenia and immediate intervention for unremitting headache is crucial. In our case, this was due to APL – a distinct subset of acute myeloid leukemia due to its unique pathophysiology and its high cure rate with treatment. First described in 1957 in patients with severe bleeding and fibrinolysis who had rapid deterioration of their clinical condition, this condition had the presence of increased promyelocytes. Advances in molecular pathology 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. 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. Despite the curable nature of APL with ATRA, early mortality is still prevalent due to bleeding complications.