INTRODUCTION

Hematologic malignancies have the highest rate of intracranial hemorrhage (ICH) amongst all cancers, however there is limited data on the incidence of ICH in childhood acute myeloid leukemia (AML).

BACKGROUND:

- Acute myeloid leukemia (AML) in pediatrics consists of a heterogenous group of diseases. Acute promyelocytic leukemia (APML), characterized by t(15;17) translocation, accounts for only 5%–10% of pediatric AML and increases in prevalence with age. ²
- A presenting feature of APML is profound coagulopathy comparable to disseminated intravascular coagulation (DIC) and may be an early indicator of APML. ³
- In adults there is a high incidence of intracranial hemorrhage (ICH) in individuals with AML, especially if AML presents early (within 7 days of diagnosis). Fatality rate reported to be as high as 75% in the study. ⁴
- Hemorrhagic stroke in patients with acute leukemia is highly fatal, especially if ICH presents early. ¹
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CASE PRESENTATION:

DT is a previously healthy 7-year-old female who presented to the ED for evaluation:

- 3 days of fever
- 2 days of fatigue and increased sleepiness
- 1 day of headache
- 1 day of abdominal pain
- 3 episodes non-bloody, non-bilious emesis

Initial Vital Signs, Exam and Labwork:

- T: 37.3 °C, HR 105, RR 20, BP 99/59, SpO2 100% on RA
- Patient noted to be lethargic but opened eyes and looked around with stimuli. Able to answer simple questions.
- Pupils 4 to 2 mm and equal bilaterally, reactive to light
- Abdominal tenderness in the periumbilical area, no guarding.

Blasts 93% 1
PT 20.3, INR 1.9, PTT 27
Fibrinogen 131

While in the ER, the patient clinically decompensated with bradycardia, widened pulse pressure, and worsening mental status and pupillary changes prompting concern for increased ICP.

Head CT obtained showing extensive intracranial hemorraghes in the bilateral frontal lobes with extensive local mass effect and changes of early herniation requiring emergent neurosurgical intervention.

Hospital Course:

- DT remained in critical condition status for 1 day of headache
- There is limited recognition and management of complications related to chemotherapy but was eventually able to be discharged home in stable condition with mild left-sided weakness.

DISCUSSION:

- Given the severe coagulopathy present on presentation and because the initial flow cytometry seemed most consistent with APML, patient was empirically started on APML therapy to reduce the risk of immediate morbidity or mortality from bleeding episodes. She was closely monitored for differentiation syndrome while being treated with ATRA.
- Confirmatory testing was negative for the necessary t(15; 17) and RT-PCR in blast cells thus DT was ultimately diagnosed with AML and started on the appropriate therapy.
- Notably, DT presented with an initial WBC of 319 x 10^9 WBC in addition to significant thrombocytopenia.
- Leukocytosis is a pathological process where WBCs accumulate in small blood vessels. Studies have shown an association between hyperleukocytosis (>100K WBCs) and CNS ischemia and hemorrhage. ³
- AML has more frequent leukostasis than ALL owing to larger and lesser deformable blasts compared with lymphoblasts. The overcrowded blasts in the capillaries furthermore release cytokines inducing direct endothelial cell damage of blood vessels. ⁴

CONCLUSION:

- Given the severity of initial presentation, DT’s overall outcome was favorable, highlighting the importance of early recognition and treatment in ICH on improved patient outcome.

REFERENCES: