

## Introduction

- Juvenile myelomonocytic leukemia (JMML) is a rare childhood hematopoietic disorder classified as a mixed myeloproliferative neoplasm and myelodysplastic syndrome.
- JMML typically presents with hepatosplenomegaly, lymphadenopathy, pallor, fever, and cutaneous findings. Here we discuss a unique presentation in the form of a subdural hematoma (SDH).
- Infantile SDH should immediately raise suspicion for accidental and non-accidental head trauma, however mental note must also be paid to the less common etiologies such that appropriate workup proceeds in a timely and organized manner should initial findings be unremarkable.

## Case Description

- A seven-month-old male with a recent history of influenza was referred for neurosurgical evaluation of a full fontanelle and increasing head circumference in the setting of recurrent low grade fevers, rhinorrhea, and an intermittent cough.
- Subsequent MRI evaluation (Fig. 1) demonstrated chronic, bilateral holo-hemispheric subdural collections.
- The patient tested positive for adenovirus, but trauma, infectious, and coagulopathy workups were otherwise unrevealing.

- Operative management was pursued in the form of bilateral burr holes to wash out the subdural collections, which were determined to be chronic hematomas. A sample of the fluid was collected and sent for analysis.
- Postoperative imaging (Fig. 2) demonstrated an acute right frontal convexity and parafalcine extra-axial hemorrhage.
- Concurrently, the patient developed splenomegaly, severe anemia, and thrombocytopenia (platelet count of ~10,000), necessitating multiple transfusions of packed red blood cells and platelets.
- Cytologic analysis of the initial subdural fluid demonstrated atypical mononuclear cells (Fig. 3) with subsequent flow cytometry describing a 2% population of CD34+ blasts with immunophenotypical features of myelomonocytic and T-lineage differentiation.
- Bone marrow aspirate demonstrated hypercellular marrow, decreased megakaryocytes, and a 1-2% population of abnormal CD34+ blasts.
- Next-generation sequencing demonstrated an NRAS Q61R mutation, a germline mutation of which was subsequently ruled out.
- The combination of clinical (splenomegaly), hematologic (peripheral monocytosis with <20% peripheral blood and marrow blasts), and genetic features (absence of a BCR-ABL1 fusion and presence of a somatic NRAS mutation) met diagnostic criteria for JMML.<sup>1</sup>

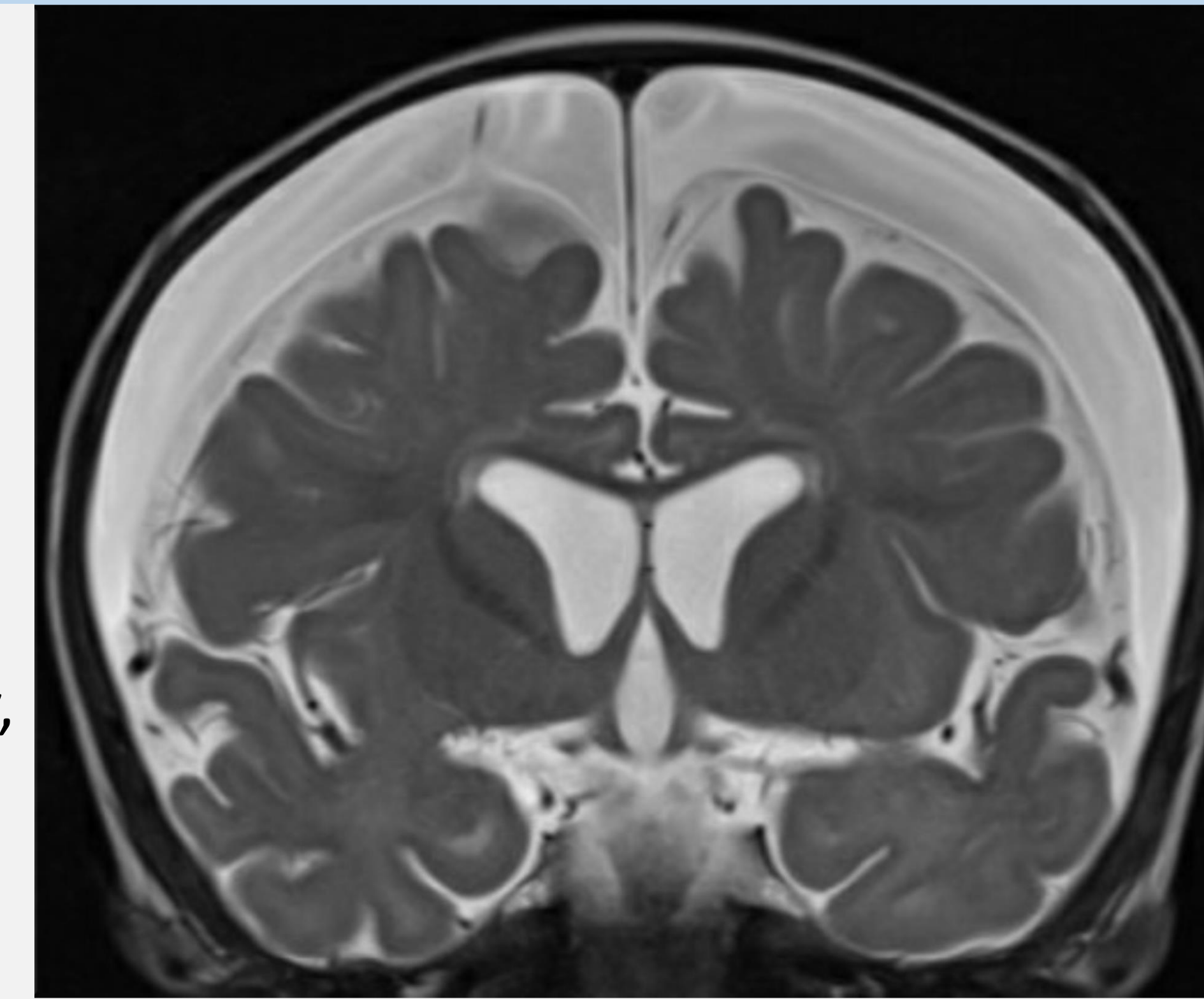


Figure 1: Pre-Operative MRI – T2 Coronal

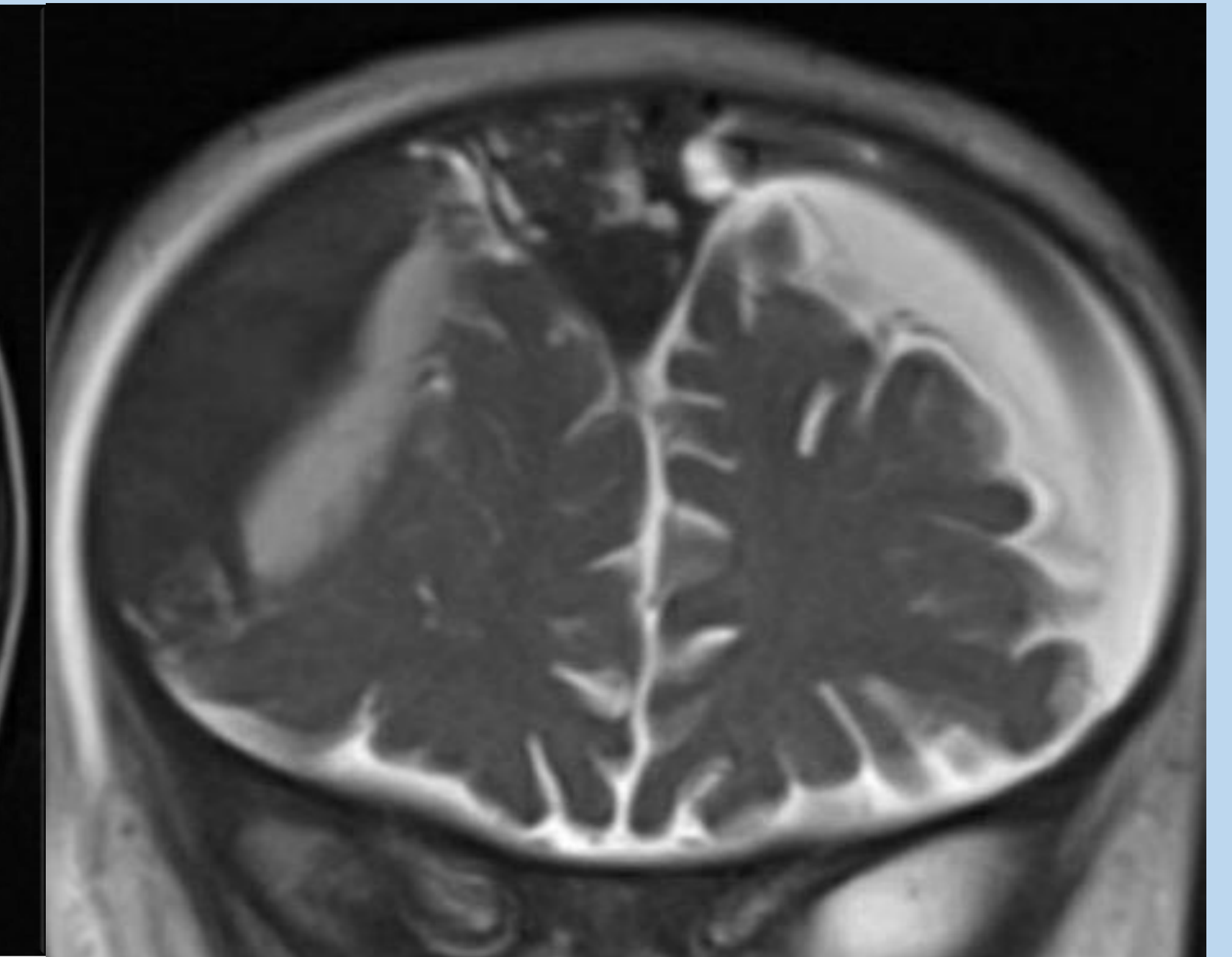


Figure 2: Post-Operative MRI – T2 Coronal

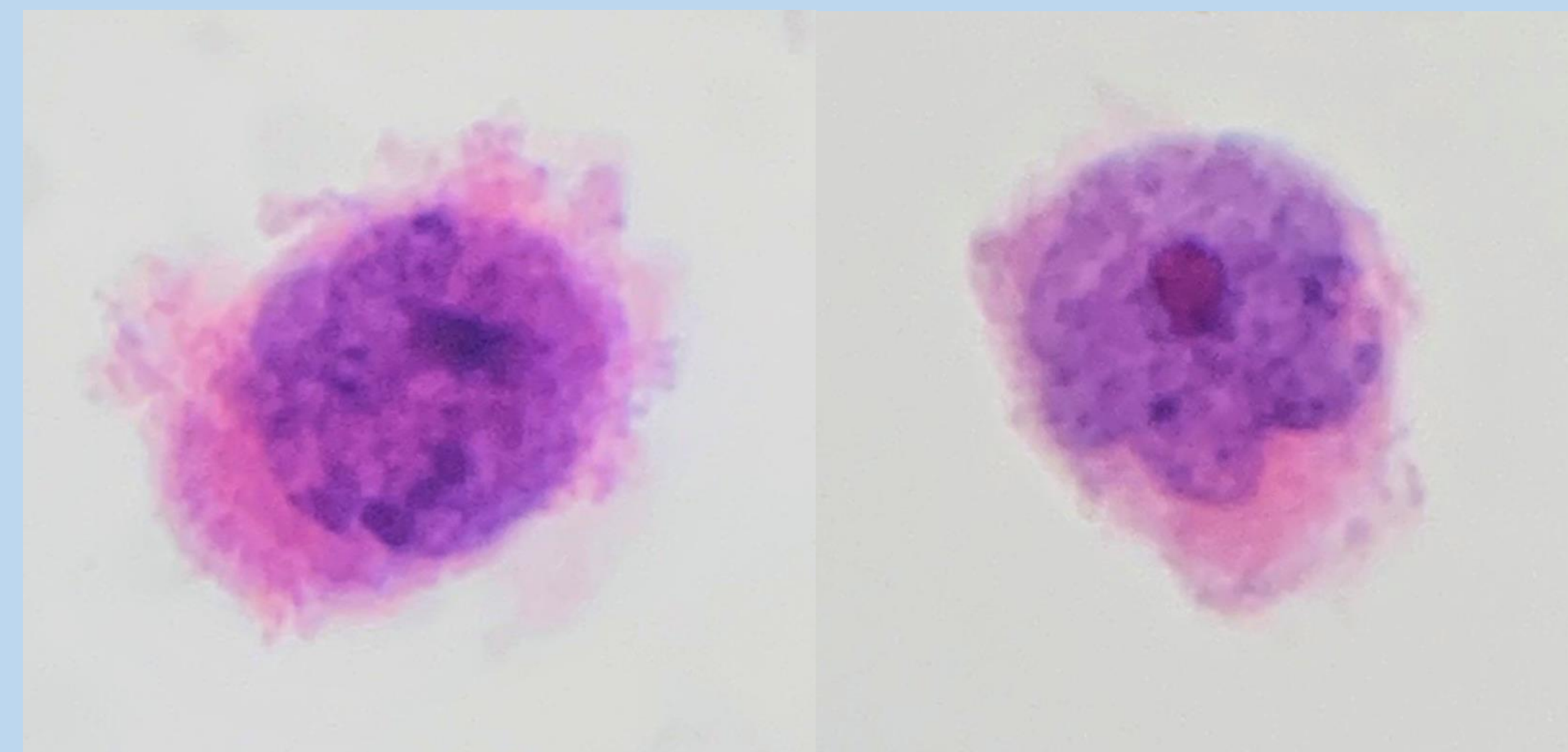


Figure 3: Atypical mononuclear cells from subdural fluid sample

## Discussion

- The case presented herein serves to review a unique presentation of a rare disease, as well as a reminder to maintain a broad differential even in pathologies with predominant etiologies.
- Had the clinical team not maintained neoplasia in the differential for this SDH then the subdural fluid may not have been collected and sent for analysis, which would likely have delayed accurate diagnosis and treatment of JMML.

## References:

1. Locatelli F, Niemeyer CM. How I treat juvenile myelomonocytic leukemia. Blood. 2015;125(7):1083-1090.