

Acute Spontaneous Intracranial Epidural Hematoma and Disseminated Intravascular Coagulation in a Pediatric Sickle Cell Patient: Case Report

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Introduction

Intracranial epidural hematomas (EDH) typically occur in the setting of trauma. Acute non-traumatic intracranial epidural hematomas are very rare, occurring in the contexts of pregnancy, dialysis, Paget's disease, coagulopathy, metastases to dura, infection, vascular malformation, and sickle cell disease. While epidemiologically the incidence of combined ischemic and hemorrhagic strokes in the pediatric population is about 1 case per 100,000 children, in SCD patients the frequency is about 200 per 100,000 children. We report a rare case of a spontaneous EDH complicated by the development of disseminated intravascular coagulopathy (DIC) in a pediatric sickle cell disease (SCD) patient.

Methods

We present the first ever reported case of spontaneous EDH complicated by DIC in the setting of pediatric SCD. We discuss possible mechanisms and report on a review of the pertinent literature.

Results

An 18-year-old African American male with SCD, in the midst of an an acute sickle cell crisis, spontaneously developed acute bifrontal EDHs that required emergent evacuation. However, he also subsequently developed DIC. Despite successful evacuation of the EDH, the patient's EDH reaccumulated. After multiple transfusions, the patient underwent a repeat EDH evacuation along with craniectomy and placement of an intracranial pressure monitor. With medical therapy and ICP management, the patient's neurologic status improved slightly. Due to family's wishes, no further intervention was pursued, and the patient expired from respiratory complications.

Literature describes (1) acute vasoocclusive infarction of a craniofacial bone, and (2) acute bone marrow hyperplasia from acute anemia, as mechanisms of EDH in SCD.

Conclusions

SCD presents a unique hematological landscape by which hemorrhagic sequelae such as intracranial EDH can occur more frequently and by atypical mechanisms. In our case report, we

Learning Objectives

By the conclusion of this session, participants should be able to: 1) understand the epidemiology of acute cerebral events specific to SCD, 2) describe the proposed underlying mechanisms of spontaneous EDH formation specific to SCD, 3) identify the grave complications that a coagulopathy such as DIC can cause in a SCD patient.

References

Available Upon Request



This CT scan is obtained prior to the patient's first surgery.

Re-accumulation of left acute epidural hematoma



This CT scan was obtained after the first surgery, before the second surgery. At this time the patient was in DIC, and his neurological exam was poor.

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