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Born to Bleed: Fatal Intracranial Hemorrhage in a Patient with a Rare Sickle Cell Variant

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Introduction

Sickle cell disease is associated with increased rates of hemorrhagic stroke, often with patients presenting with severe headache and altered neurologic exam. Due to its high mortality, rapid recognition and diagnosis are necessary for quick intervention.

Case Summary

- A 25-year-old African American male with SO-Arab sickle cell disease presented with an acute, severe headache, extremity pain, and auditory hallucinations. He presented distressed and tearful, stating that his pain was 10/10.
- Vitals: T (36.5°F), BP (123/83), HR 71 bpm, RR 18 bpm, SpO2 96% on RA
- PE: slowed cognition and emotional lability without focal neurologic deficit
- He was treated with hydromorphone, ondansetron, and intravenous fluids as he awaited chest roentgenography (x-ray) and computed tomography (CT) of his head.
- During his work-up and prior to head CT, he was found to be unconscious. He was hypoxic with a saturation in the 30s with good waveform. Bag-mask ventilation was initiated and 6 mg of IV Narcan were administered. Despite these interventions, there was no improvement in his mental status, so he was intubated for airway protection.
- His arterial blood gas revealed an acute hypercapnic respiratory acidosis with a pH 7.18, PCO2 71, PO2 260, and HCO3 25.
- Non-contrast Computed Tomography (CT) of the Head: large cerebellar hematoma with obliteration of the fourth ventricle, obstructive hydrocephalus, as well as both transtentorial and tonsillar herniation.
- Neurosurgery was consulted who recommended no treatment due to the catastrophic, non-survivable bleed

Table 1: Laboratory Values

White blood cell count (g/dL)	14.4
Hemoglobin (g/dL)	10.9
MCV (fL)	103.1
Platelets (g/dL)	445
Bilirubin, Total (mg/dL)	2.8
Bilirubin, Direct (mg/dL)	0.6

Images

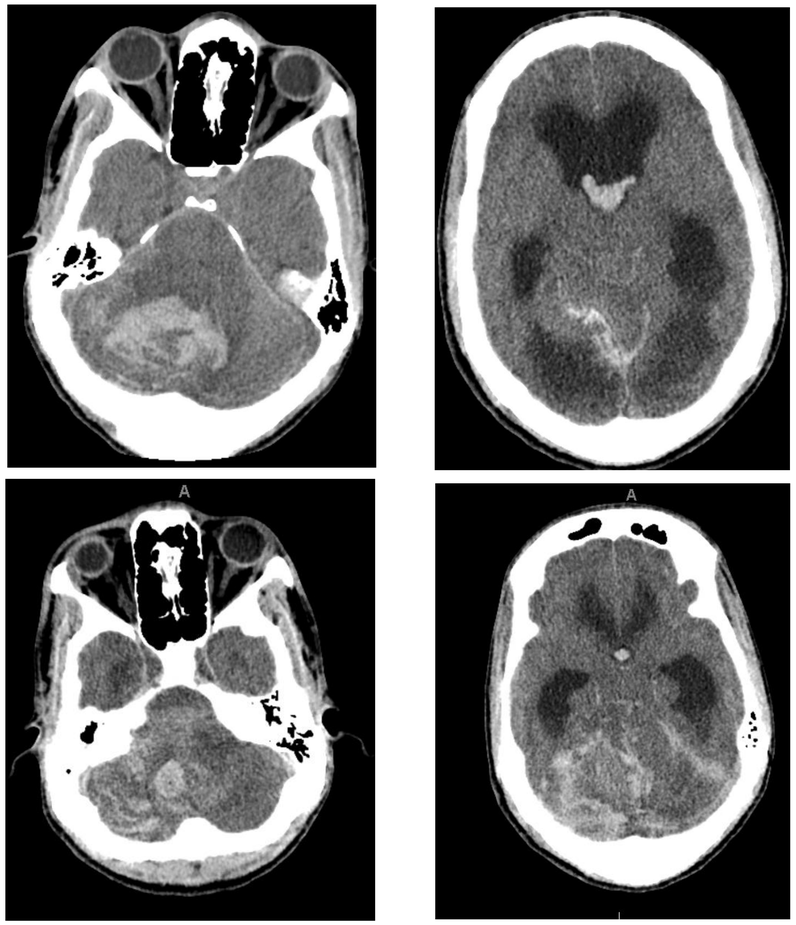

Figure 1.Noncontrast CT of the Head showing hemorrhage, hydrocephalus, and herniation

Table 2: Risk Factors for Hemorrhagic Stroke in Sickle Cell Disease

- Age*
- Low steady-state hemoglobin concentration*
- High steady-state leukocyte count*
- Acute chest syndrome^
- Previous history of stroke^
- Recent transfusion^
- Recent corticosteroid use^

*Proven associations
^ Proposed from case report/series associations

Discussion

Hemorrhagic stroke in sickle cell disease is rare compared to ischemic stroke, but accounts for the mortality from stroke [1]. The mortality is high, ranging from 25% to 50%, with most of these patients dying within 2 weeks of the hemorrhagic event [1]. The peak incidence occurs between the ages of 20 and 29 [2, 3]. Common presenting symptoms include severe headache, nuchal rigidity, coma, and focal neurologic deficits [1]. Risk factors include age, low steady-state hemoglobin concentration, high steady-state leukocyte count, acute chest syndrome, and previous history of stroke [1,2].

Our patient was in the age group of the peak incidence of hemorrhagic stroke. He presented with a severe headache, rare neurological changes including auditory hallucinations, and leukocytosis. He had previous pain crises, eight in the last year, but never had acute chest syndrome. He had no history previous stroke. However, he had a rare variant of sickle cell disease, the SO-Arab form. This form has been shown to be associated with severe sickling sequelae including acute chest syndrome, recurrent painful vasocclusive pain crises, and dactylitis [3]. Incidence of stroke has not been studied in this variant of sickle cell anemia, highlighting the uniqueness of this case. In conclusion, given the high mortality of hemorrhagic stroke in sickle cell disease and its unique presentation, rapid recognition and diagnosis are necessary for quick intervention.

Take Home Points

- There are many variants of sickle cell disease that vary in severity of disease presentation and outcomes
- Consider hemorrhagic stroke risk in patients with sickle cell disease
- Neurologic signs and symptoms are clues for early recognition and life-saving workup for hemorrhagic stroke

References

1. Strouse, J.J., et al., *Primary hemorrhagic stroke in children with sickle cell disease is associated with recent transfusion and use of corticosteroids.* Pediatrics, 2006. **118**(5): p. 1916-24.
2. Ohene-Frempong, K., et al., *Cerebrovascular accidents in sickle cell disease: rates and risk factors.* Blood, 1998. **91**(1): p. 288-94.
3. Zimmerman, S.A., et al., *Hemoglobin S/O(Arab): thirteen new cases and review of the literature.* Am J Hematol, 1999. **60**(4): p. 279-84.