When a Sickle Cell Crisis is Not a Sickle Cell Crisis

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LEARNING OBJECTIVES

1. Recognize clinical and laboratory findings consistent with sickle cell disease
2. Utilize in-hospital transitions of care between providers as an opportunity to consider all aspects of the clinical picture.

CASE INFORMATION

HPI

28-year-old woman with PMH significant for sickle cell disease presents with diffuse body pain reported to be consistent with her typical vaso-occlusive pain crises, triggered by cold weather. Generalized pain, myalgia. No recent acute illness.

PMH/PSH

Two lifetime hospitalizations for vaso-occlusive pain crisis
s/p appendectomy
s/p cholecystectomy

MEDS

Tylenol and non-steroidal medications

EXAM

Intermittently on O2
Lungs clear to auscultation bilaterally
No hepatomegaly or splenomegaly

LABS

Admission Labs		Sickle Cell Workup Labs	
Hb / Hct	11.8/37	Hb / Hct	11.5/38
MCV	72.1	MCV	75.9
Retic count/absolute	0.5%/30	Retic count/absolute	0.7%/40
Creatinine Kinase	54	Peripheral Smear	No hemolysis
Pregnancy	Neg	Haptoglobin	normal
Troponin	15.3	Ferritin	normal
Respiratory viral panel	neg	Iron Binding Panel	normal
Creatinine	0.7	Folate	normal
Urinalysis	Ketones, glucose, mucus	Hb Electrophoresis	normal
		Indirect Bilirubin	1.5, mild elevation

IMAGING

- Chest x-ray: no acute findings.
- CT chest: no pulmonary embolism, + for multiple small scattered nodules <4mm

HOSPITAL COURSE

- Admitted and started on hydromorphone PCA
- Multiple titrations with multiple providers
- After transition of care, noted unusual lab findings for patient with sickle cell disease, completed appropriate workup with normal laboratory values
- Patient determined not to have sickle cell as etiology of acute pain.
- Laboratory values at Hematology clinic follow-up remained within normal limits, not consistent with a diagnosis of sickle cell disease

WHAT IS SICKLE CELL DISEASE?

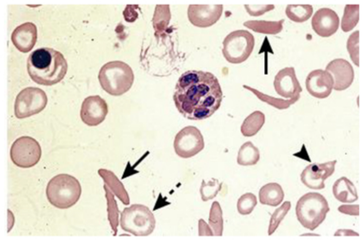
Sickle Hemoglobin S (HbS) gene mutation on at least one beta globulin gene

AND

A second beta globulin gene mutation

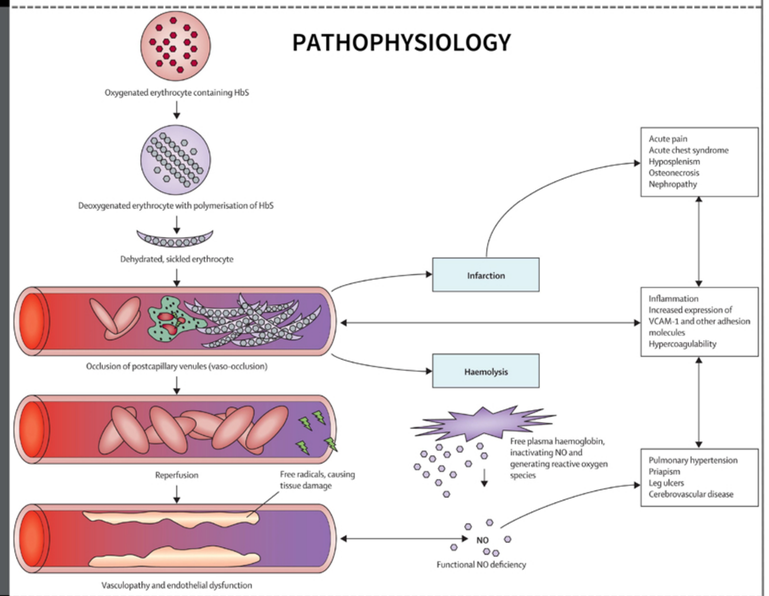
→ Sickling → Vaso Occlusive Crisis And Hemolytic Anemia

PERIPHERAL BLOOD SMEAR



Arrows = multiple sickle cells
Upper left = Nucleated RBCs
Arrowhead = Howell-Jolly body
Dashed arrow = target cells

PATHOPHYSIOLOGY



TYPICAL LABORATORY FINDINGS AND CLINICAL EXAM
Mild to moderate anemia (hematocrit 20-30%)
Reticulocytosis
Unconjugated hyperbilirubinemia
Elevated serum lactate dehydrogenase
Low serum haptoglobin
Hyposplenism (function) and splenomegaly (size)
Confirmatory test is typically hemoglobin electrophoresis

IMPLICATIONS/DISCUSSION

Type of Bias	Description
Anchoring bias	Implicit reference point of first data
Attribution bias	Attempts to discover reason for observations
Search-satisficing bias	Tendency to believe that our current knowledge is sufficient and complete
Confirmation bias	Favor information confirming first belief
Framing bias	Favor based on presentation of information in negative or positive context
Status quo bias	Favor of options supporting current scientific dogma
False consensus bias	Tendency to overestimate how much others agree with us
Blind spot bias	Tendency to believe one is less biased than others
Not-invented-here bias	Bias against external knowledge

While there is an increase in in-hospital mortality associated with transitions of care between providers on service teams, this is also an opportunity to re-evaluate patient care plan for anchoring bias, confirmation bias, and status quo bias.

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