

THEY ARE TRYING TO SICKLE

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School of Medicine
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CASE HISTORY

- 21 yo Black Female Track and Field Multi athlete with c/o painful lumps in both armpits
 - Day 3 of symptoms, Team Physician contacted by Athletic Trainer
 - Presumptive Lymphadenitis
 - Doxycycline 100mg po bid prescribed empirically
 - Day 4 developed fever, Tmax 100.5
 - Increasing pain, not improved with ibuprofen
 - Athlete presented to the ER; Took first dose of doxycycline in the waiting area
 - ER diagnosed: Bilateral Axillary Abscesses
 - Prescribed:
 - Clindamycin 450mg po tid; Stop Doxycycline
 - Ketorolac 40mg IM x 1
 - Day 5 seen by Team Physician in clinic
 - Pain 7/10 both axillae; unable to lower arms completely
 - No further fevers
 - Denied night sweats, unexplained weight loss or other lymphadenopathy
- PMHx: Iron Deficiency Anemia, Vitamin D Insufficiency; Sickle Cell Trait Negative (Mandatory NCAA screening)
- PSHx: Inguinal hernia repair as a child
- Meds: Ferrous Gluconate 324mg po bid, q other day with Ascorbic Acid 500mg; Cholecalciferol 2000IU/d; Nexplanon; Clindamycin; Ibuprofen
- Social: No tobacco, drugs
- Fam Hx: No known hemoglobinopathies; Mother unsure of family ancestry details
- ROS: No history of jaundice, splenomegaly, cramping with exertion

INITIAL PHYSICAL EXAMINATION

- T 98.1 P79 RR 18 BP 110/70 BMI 22.2
- Gen: A&O x 3; NAD, but holding arms away from body
- HEENT, Resp, CV: Unremarkable
- Both Axillae:
 - Visible axillary lymph node enlargement b
 - Right: 2 large firm nodes and at least 5 smaller nodes
 - Left: 2 large firm nodes and at least 4-5 smaller nodes
 - All enlarged nodes, exquisitely tender
 - No fluctuance of any nodes consistent with abscess
- Lymphatics: No other LAN to include cervical, supraclavicular, epitrochlear, submandibular or inguinal; no splenomegaly
- Skin: No discharge from nodes; No erythema of axillae
- MSK: Unremarkable

DIFFERENTIAL DIAGNOSIS

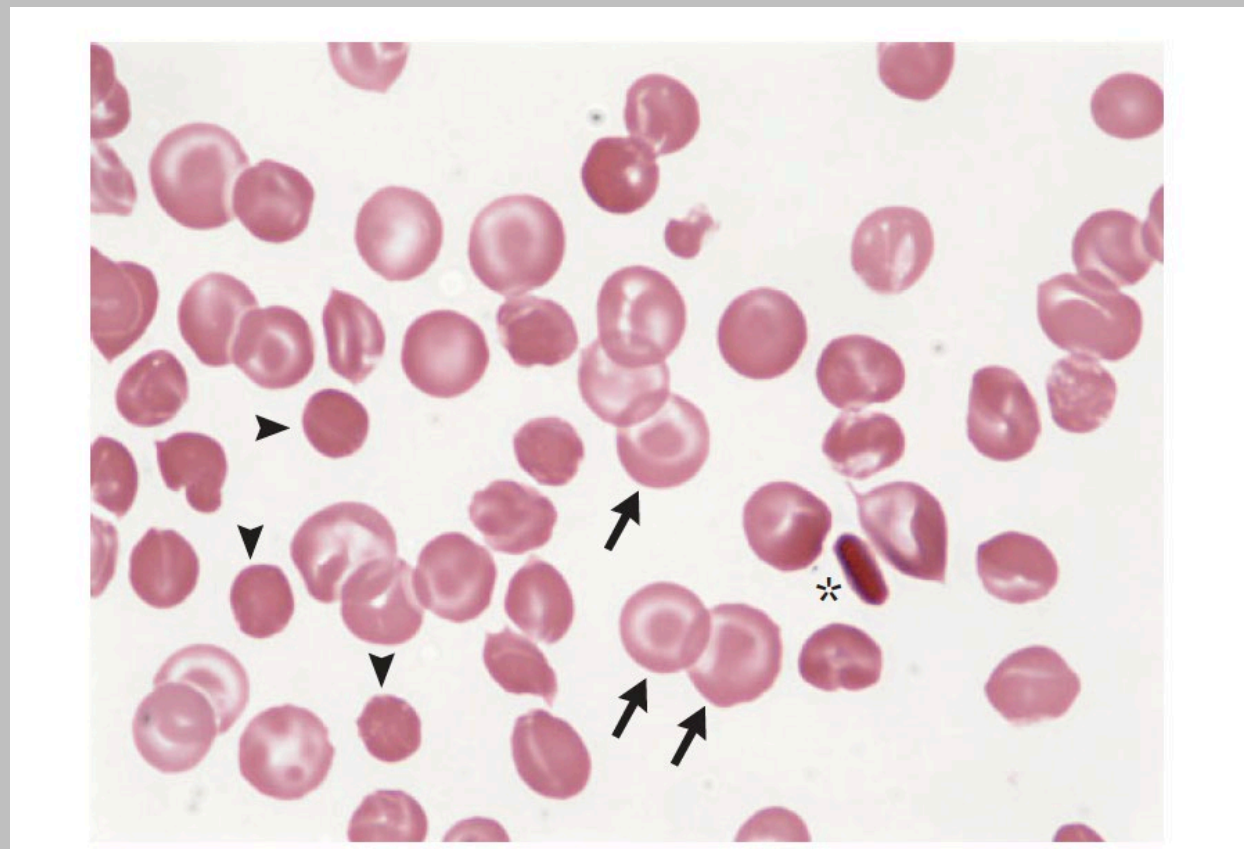
Bilateral Axillary Lymphadenitis
Methicillin Resistant Staph Aureus Infection
Lymphoma
Hidradenitis Suppurativa
Bilateral Axillary Abscesses

LABWORK AND IMAGES

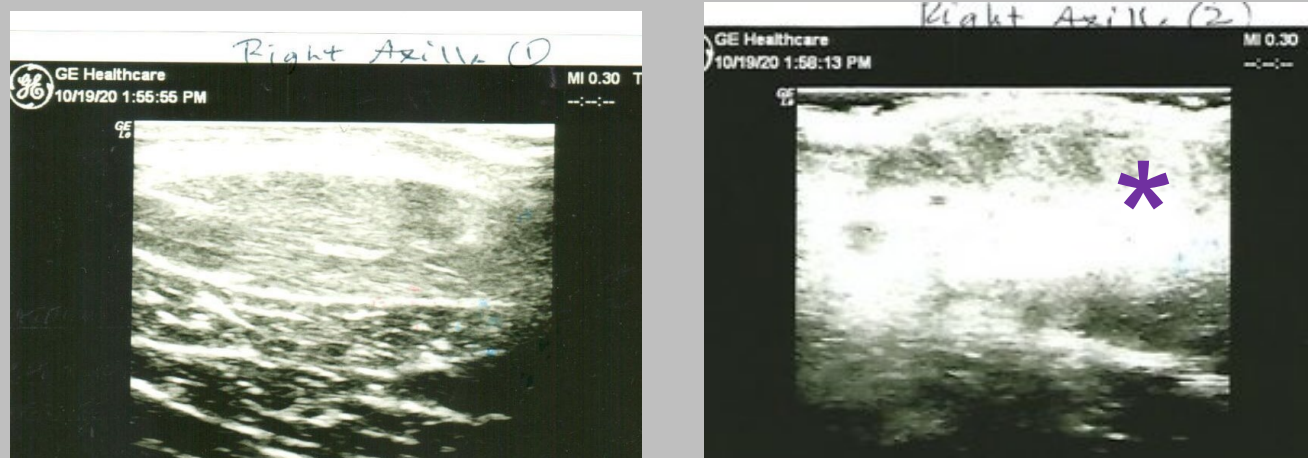
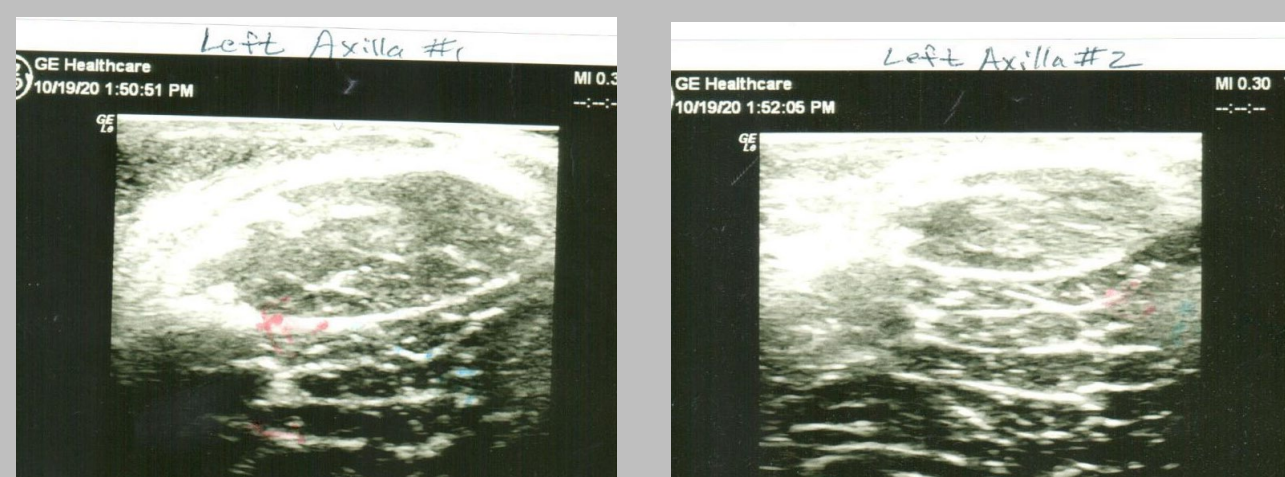
- Sofia Antigen COVID-19 Screening Test: Negative
- Comprehensive Metabolic Profile, unremarkable
- Monospot: Negative
- CBC due to infection concern; other labs done due to abnormal indices and abnormal peripheral smear showing:

- Target Cells 3+
- Microcytosis 3+
- Anisocytosis 2+
- Poikilocytosis 2+
- Schistocytosis 1+
- Hypochromasia 1+

Comment from Lab Technician:
“The red blood cells look like they are trying to sickle.”



- Typical Peripheral Smear
- Hemoglobin C Disease:
 - * HbC Crystal
 - Arrowhead: Microspherocyte
 - B-Thalassemia:
 - Pappenheimer Bodies
 - Poor Hemoglobinization
 - Both Conditions:
 - Arrows: Target Cells



Day 5 US images with enlarged lymph nodes (*), but no fluid collections essentially ruling out Axillary Abscesses

Hemoglobin Electrophoresis			
	Pt. Values	Normals	
Hgb A	16.7	95 - 98	Low
Hgb A2	4.5	2 - 3	High
Hgb F	9.1	0.8 - 2	High
Hgb C	69.7	Abnormal	

Hgb Electrophoresis consistent with Hemoglobin C / B-Thalassemia

Labs:	Patient Value	Normals	
WBC	8.2	4.8 - 10.8	Normal
RBC	5.2	4.2 - 5.4	
Hgb	10.9	12 - 16	Low
HCt	32.5	37-47	
PLT	288	130 - 400	
MCV	62.4	81 - 99	
MCH	20.9	27 - 31	
MCHC	33.4	32 - 36	
RDW	18.1	11.5 - 15.5	High
Retic Count	1.92%	.5 - 1.8	
Iron	48	37-145	
TIBC	365	250-450	
Ferritin	8.2	>40	
CRP	24 - 47	<6	
Folate	8.4	4.6 - >20	

FINAL DIAGNOSIS

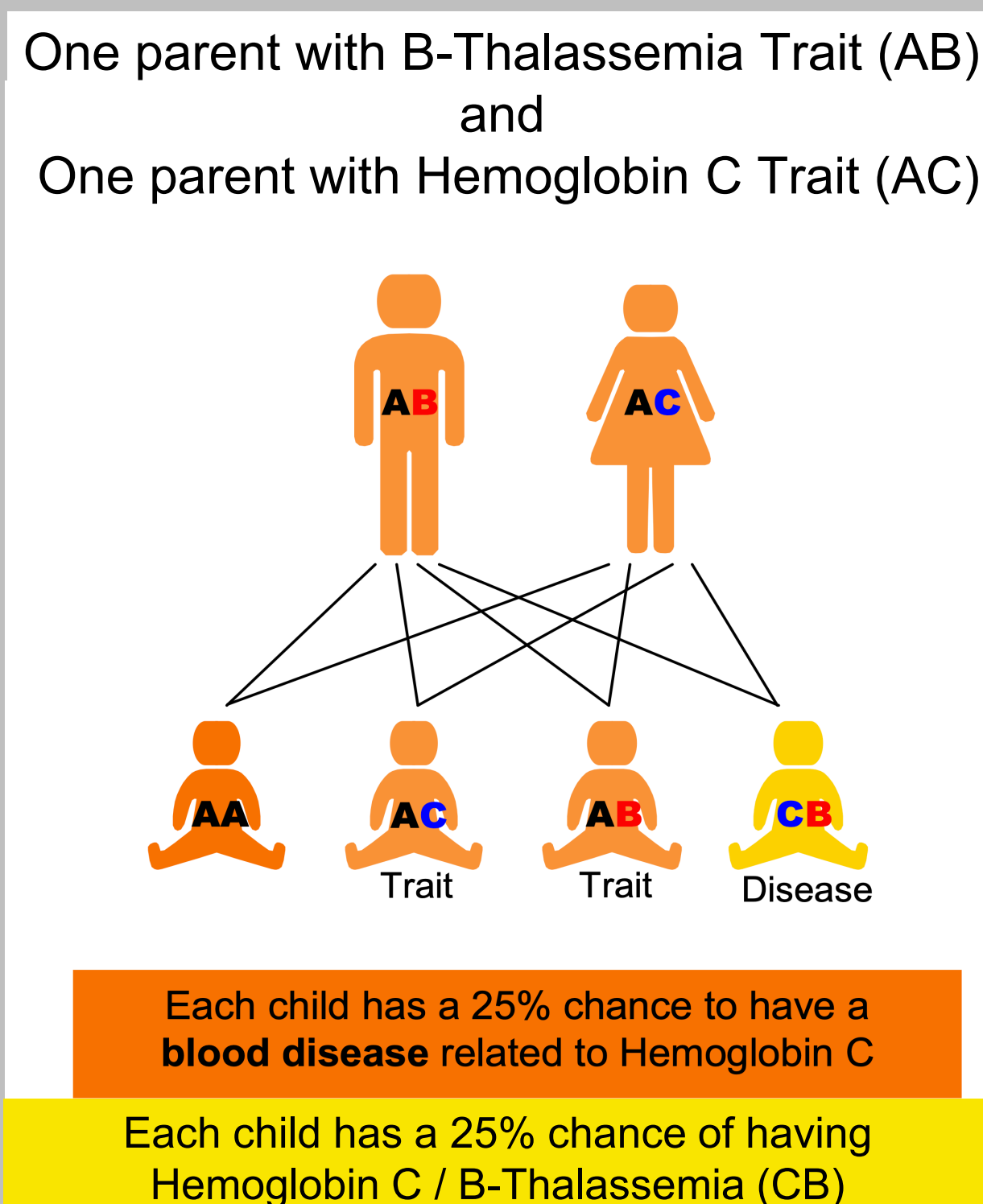
Bilateral Axillary Lymphadenitis
with incidental discovery of
Hemoglobin C / B-Thalassemia

CLINICAL COURSE

- While reviewing the peripheral smear, the lab technician noted multiple atypical shaped red blood cells with some appearing to almost sickle.
- The team physician was contacted to ask if the patient has Sickie Cell Trait; screening was negative during her PPE
- Hemoglobin Electrophoresis was then ordered
- Day 8: pain resolved, Lymphadenitis resolving
- Day 13: Lymphadenitis clinically resolved
- Patient counseled regarding the new hemoglobinopathy dx

DISCUSSION/SIGNIFICANCE

- Hemoglobin C is one of the structural variants of normal hemoglobin
- Hgb C Trait (Hb AC), typically is asymptomatic, but does not carry Oxygen as well as normal hemoglobin; Poor Quality Hemoglobin
- B-Thalassemia arises from deficient or absent synthesis of B globin chains leading to excess alpha chains; Low Quantity
 - Patient typically asymptomatic or mild anemia
 - Low MCV; RDW high or normal
- Hemoglobin C / B-Thalassemia:
 - One Hgb C gene from one parent and
 - One Thalassemia gene from the other parent (see figure)
 - Mild to moderate anemia
 - Typically does not cause serious health problems
 - Low MCV and low Hemoglobin *always* seen
- Mutations seen in hemoglobinopathies protect carriers from malarial diseases
- Preconception genetic counseling is important for those with hemoglobinopathies.
- Any offspring of this individual will inherit either a gene for Hgb C or for Beta-thalassemia.
- If her partner has abnormal hemoglobin the offspring will have a high likelihood of having abnormal hemoglobin rather than just being a carrier.
- Those at risk of being carriers for beta-thalassemia include populations in the Mediterranean basin, West Africa, and South Asia.
- Prior to any decision regarding conception, she should strongly consider partner carrier testing.



RETURN TO ACTIVITY/FOLLOW-UP

- With the lymphadenitis resolved, the patient returned to full activity workouts.
- It was recommended her siblings, her parents, and their siblings should consider Hgb electrophoresis testing.
- She was counselled regarding potential future genetic issues with her offspring depending on partner's hemoglobin components.

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