THEY ARE TRYING TO SICKLE

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TEXAS TECH UNIVERSITY HEALTH SCIENCES CENTER™
School *of* Medicine
Family and Community 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

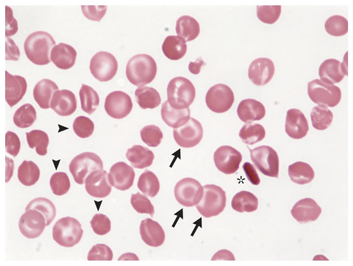
DIFFERENTIAL DIAGNOSIS

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."



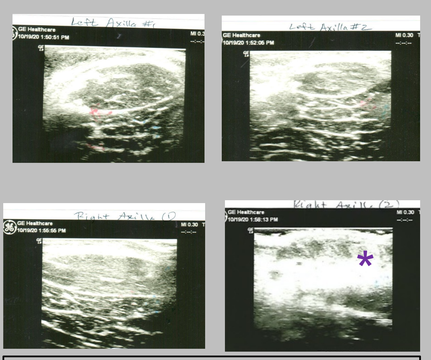
Typical Peripheral Smear
- Hemoglobin C Disease:
  - * HbC Crystal
  - Arrowhead: Microspherocyte
- $\mathcal{B}$-Thalassemia:
  - Pappenheimer Bodies
  - Poor Hemoglobinization
- Both Conditions:
  - Arrows: Target Cells

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 / $\mathcal{B}$-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	



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

FINAL DIAGNOSIS

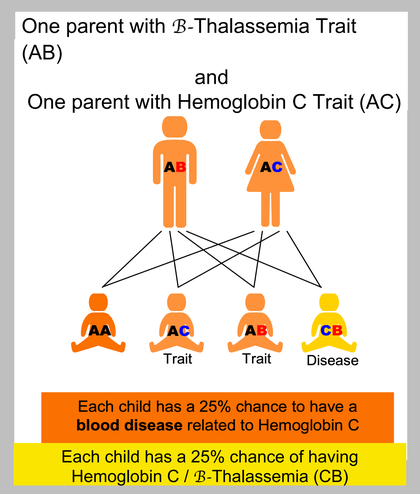
Bilateral Axillary Lymphadenitis with incidental discovery of Hemoglobin C / $\mathcal{B}$-Thalassemia

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 Sickle 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
- $\mathcal{B}$-Thalassemia arises from deficient or absent synthesis of $\mathcal{B}$ globin chains leading to excess alpha chains; Low Quantity
  - Patient typically asymptomatic or mild anemia
  - Low MCV; RDW high or normal
- Hemoglobin C / $\mathcal{B}$-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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