Mass General Brigham

Leuk What You Made Me Do

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November, 2022

60 y/o M w/ HTN, HLD, and afib presents with a new "lump" on his neck and afib w/ RVR

- Over the course of the last 4 weeks, he noticed the development of multiple "lumps" in his axilla, groin, and most prominently right side of his neck
- During the same time period, he noticed daily fevers, fatigue, mild dyspnea, and poor PO intake
- He is a prisoner, and is transported to an OSH where he is found to be hypotensive and in atrial fibrillation with RVR
- CT neck, chest, and abdomen showed diffuse adenopathy, and given all the above, plan was made to transfer to BWH for further evaluation

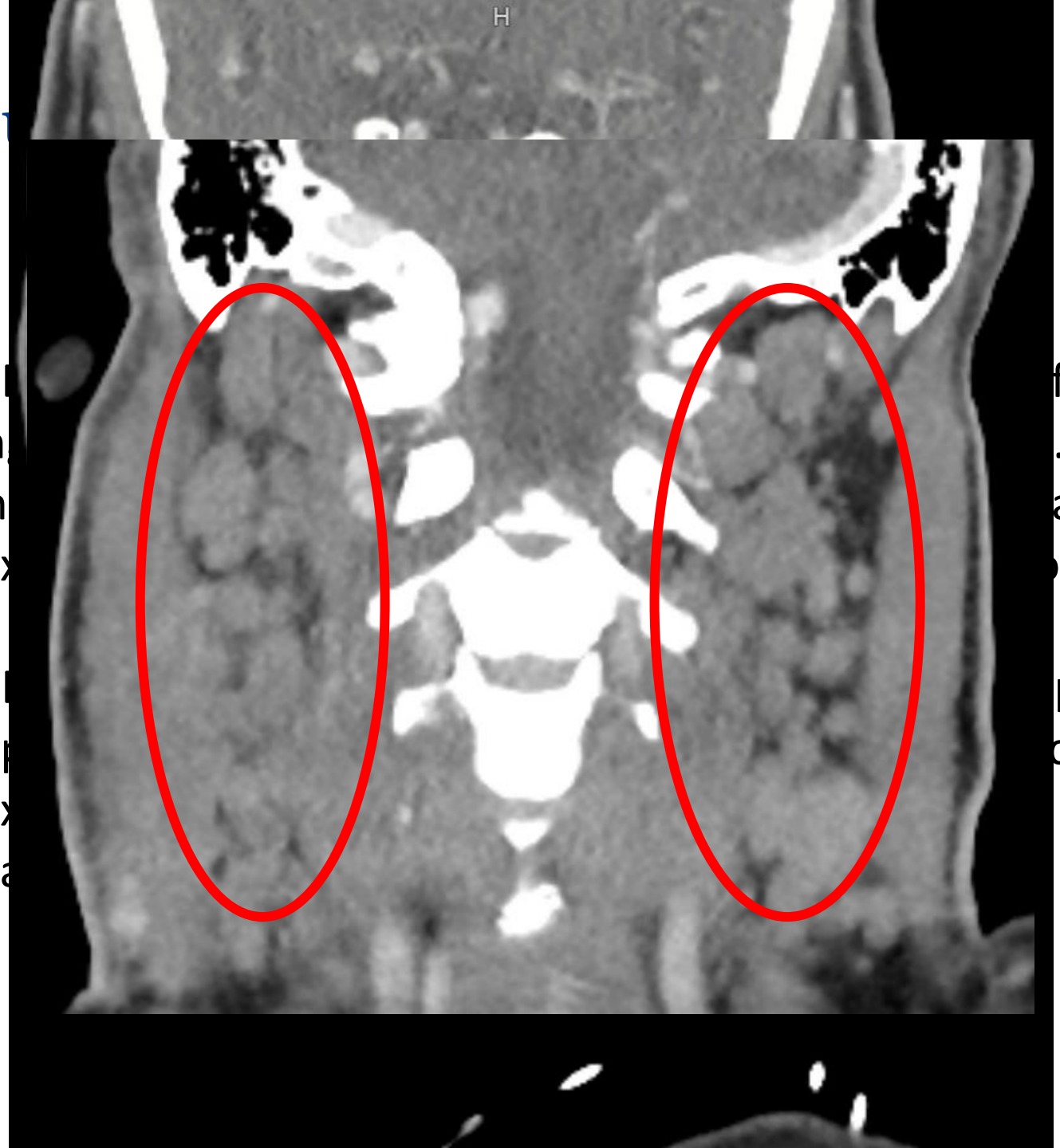
Past Medical History

- PMH: HTN, HLD, reported history of atrial fibrillation but has never been on any medications
- PSH: None
- Meds: None
- NKDA
- SH: Has been incarcerated in Massachusetts since 2018. No known tobacco, alcohol, or other recreational drug use

Physical Exam

- Vitals: 97.9 F, HR 129, BP 149/86, RR 41, SpO2 94% on 6L NC
- Gen: Patient is alert and conversant, AAOx3
- HEENT: Normal conjunctiva, icteric sclerae, EOMI
- Neck: Supple, notable cervical lymphadenopathy R>L, trachea midline
- CV: Tachycardic, irregular, elevated JVD, no LE edema
- Pulm: Increased respiratory effort, CTA bilaterally, no w/r/r
- Abd: Soft, TTP RUQ, ND; No masses or organomegaly
- Neuro: Moves all extremities, alert and communicative
- Skin: Warm and dry; no rash

Lymphatic Bu



CT Neck:

- Lymph Nodes: [redacted] ft level 2 lymph node measurin[redacted].2 x 1.7 cm, left level 5 lymph n[redacted]ar lymph node measuring 1.9 x[redacted]des.

CT Abdomen:

- Lymph Nodes: [redacted]ph nodes; portocaval lym[redacted]c lymph node measuring 2.2 x[redacted]
- Spleen: Modera[redacted]

What's the Differential for Diffuse Lymphadenopathy?

Infections

Viral

-HIV, EBV, CMV, HSV, VZV

Bacterial

-Mycobacteria

-Syphilis

-Rickettsia

-Brucella

Fungal

-Endemic mycosis

-Cryptococcus

Parasitic

-Toxoplasmosis

Neoplasm

Malignant

-Leukemia

-Lymphomas

-Metastasis

-Sarcoma

Benign

-Castleman

-Kikuchi

-Autoimmune lymphoproliferative

Autoimmune

-SLE

-Sarcoidosis

-Sjogren

-Still's Disease

-HLH

-Rheumatoid Arthritis

-Kawasaki Disease

Exposures

Toxins

-Silicosis

-Berylliosis

Meds

-Hydralazine

-Allopurinol

-Phenytoin

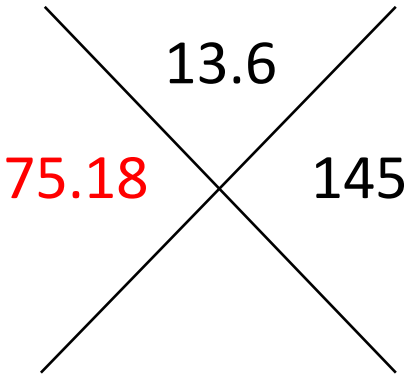
-Dress

Other

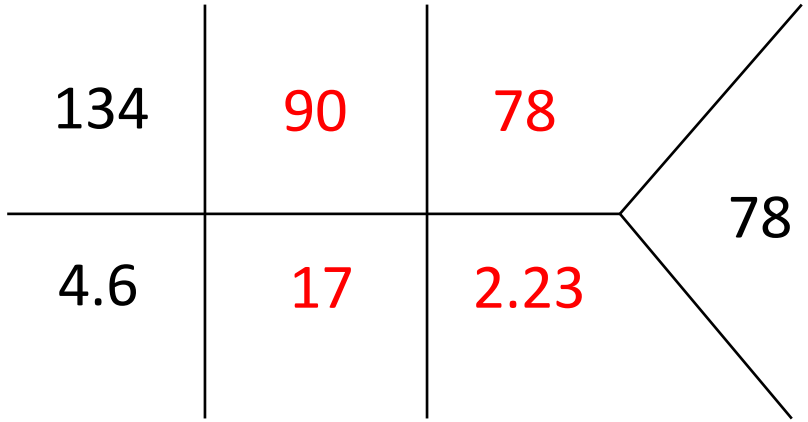
-Amyloidosis

-IgG4 related disease

Labs



89% neutrophils
8% Lymphs
2% Monos
1% metamyelocytes



Anion gap: 27

AST: 135
ALT: 90
Alk Phos: 278
Tbili: 8.1

Lactic Acid: 9.7
INR: 1.5
UA: Unremarkable
Bcx: Negative
Viral testing: Negative

Update the Differential?

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-HIV, EBV, CMV, HSV, VZV

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-Cryptococcus

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Neoplasm

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-Leukemia
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Autoimmune
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-Still's Disease
-HLH
-Rheumatoid Arthritis
-Kawasaki Disease

Exposures
Toxins
-Silicosis
-Beryllosis
Med
-Hydralazine
-Allopurinol
-Phenytoin
-Dress

Other
-Amyloidosis
-IgG4 related disease

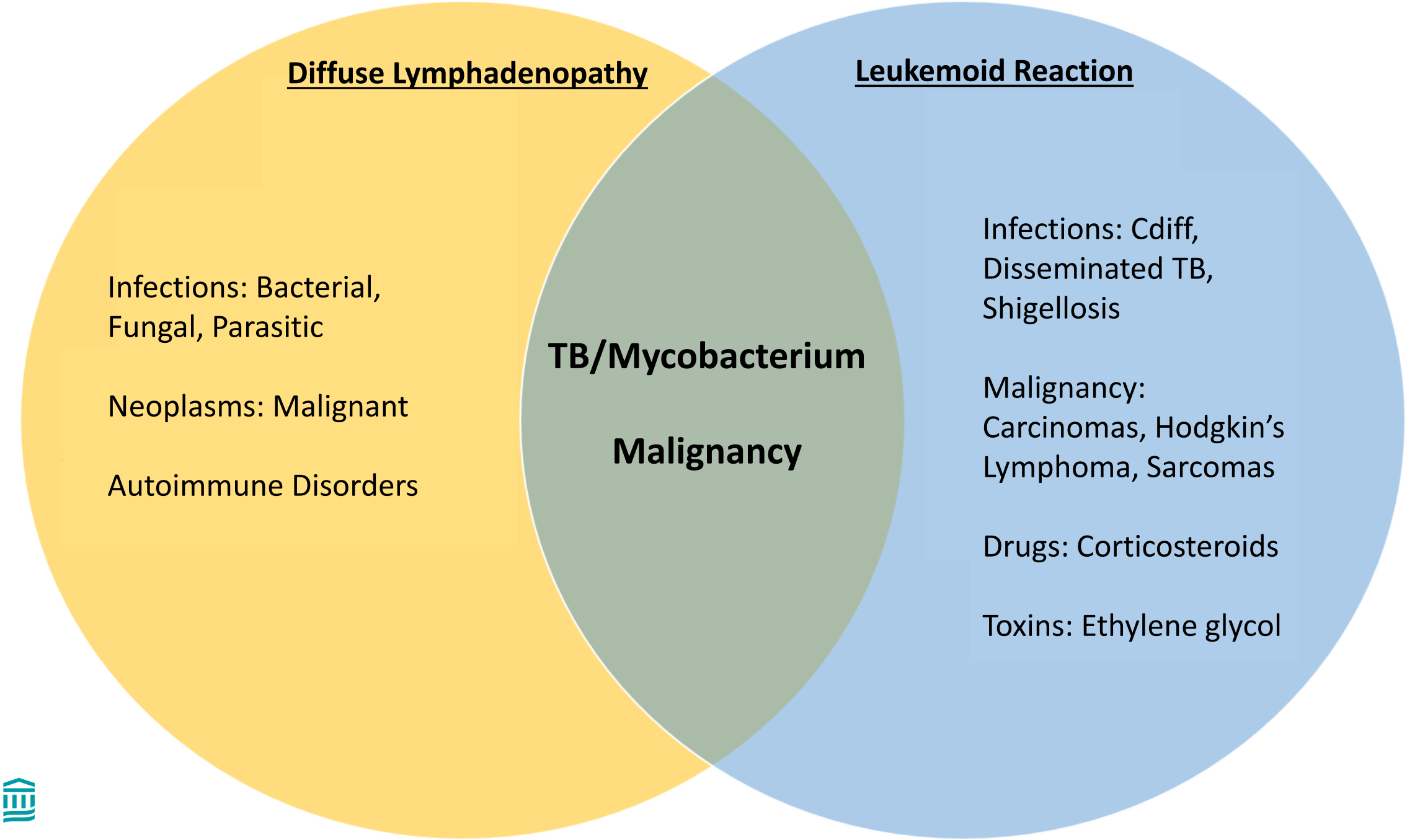
ED Course

- Loaded with amiodarone with mild improvement in rates, but in setting of ongoing tachycardia, respiratory distress, and severe acidosis, decision is made to admit to MICU
- A broad infectious work-up is sent, and the patient is empirically started on vanc/cefepime
- Oncology evaluates the patient and feels he is likely having a “a leukemoid reaction with dully bodies and toxic granulations.”

“Extreme Leukocytosis”

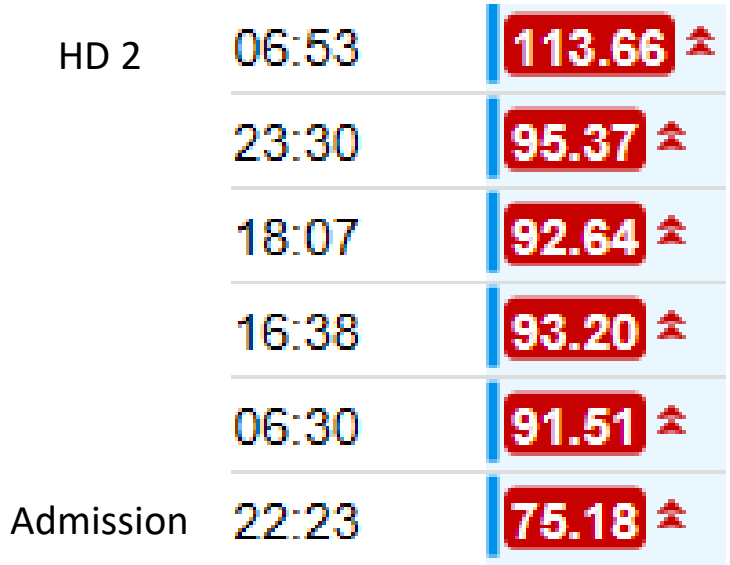
- Identified patients in the ED who presented with “extreme leukocytosis” (defined as leukocytes >25 × 10^9) and those with a moderate leukocytosis (defined as leukocytes 12–25 × 10^9)
- Those with “extreme leukocytosis” where more likely to have a “pathologic” CXR, had received prior antibiotics, and to have an identified infectious disease. There was no significant difference in vital signs
- “Extreme leukocytosis” was associated with a higher case fatality rate

Source: Y.R. Lawrence, D. Raveh, B. Rudensky, G. Munter, Extreme leukocytosis in the emergency department, *QJM: An International Journal of Medicine*, Volume 100, Issue 4, April 2007, Pages 217–223, https://doi.org/10.1093/qjmed/hcm006


Diffuse Lymphadenopathy
Infections: Bacterial, Fungal, Parasitic
Neoplasms: Malignant
Autoimmune Disorders
TB/Mycobacterium
Malignancy
Leukemoid Reaction
Infections: Cdiff, Disseminated TB, Shigellosis
Malignancy: Carcinomas, Hodgkin's Lymphoma, Sarcomas
Drugs: Corticosteroids
Toxins: Ethylene glycol

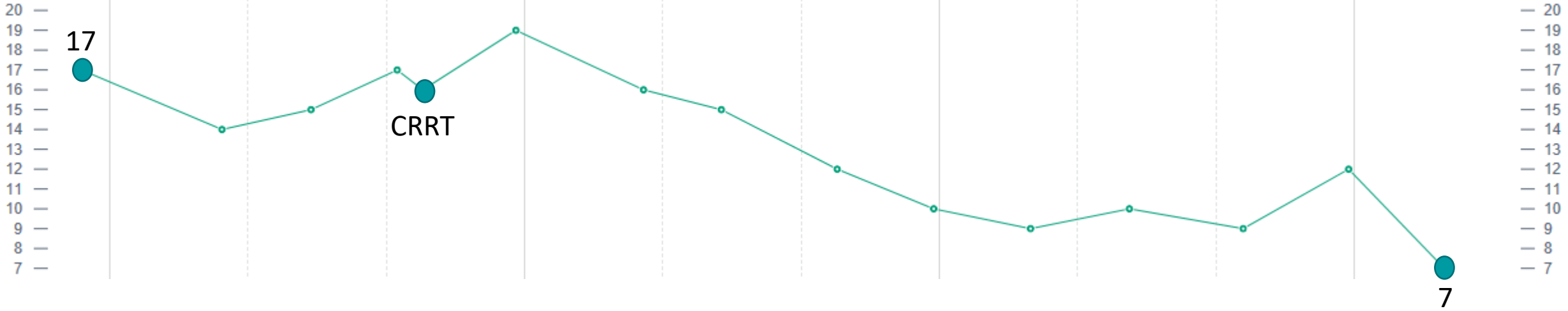
Hospital Day 1

- UOP drops significantly with worsening acidosis
- Broad infectious work-up is negative
- He develops worsening respiratory distress, requiring escalating non-invasive ventilation and ultimately not relieved by BiPAP



What would you do next?

- Patient is intubated, and post-intubation requires significant pressors and ventilatory support
- CVVH is initiated, but he continues to have worsening acidosis
- In addition to continuing broad antibiotics, the patient is started on dexamethasone



Corticosteroids in the Treatment of Neoplasms

	Solo Agent	Only in Combination with Chemotherapy	Little Efficacy
Acute Myeloid Leukemia			X
Acute Lymphoblastic Leukemia	X		
Chronic Lymphocytic Leukemia		X	
Chronic Myeloid Leukemia			X
Hodgkin Lymphoma	X		
Non-Hodgkin Lymphoma	X		
Multiple Myeloma	X		

Social Aspects of Care

- Prior to intubation, and given his worsening clinical trajectory, the team attempted to reach out to his HCP (niece). However, patient was unable to provide the contact information, and the prison only had phone numbers for two other family members, neither of whom could be reached
- On hospital day 2, following intubation, social work is able to track down his HCP and update her on the situation. The HCP requires permission from the superintendent from the prison before she is allowed to visit the patient

Who dictates visitor (and other) policies f ed patients?

SALEM HOSPITAL
Administrative Policy

Visitor access and Phone use will be at the discretion of the Law Enforcement /Correction Personnel based on each departments regulations.

Management of Patients in Custody of Law Enforcement Personnel

Brigham and Women's Faulkner Hospital
Policy

Forensic Patient Visitors - Forensic patients are not allowed visitors except in terminal and/or extraordinary cases. Those cases with special consideration are approved by the law enforcement authority in consultation with Nursing Leadership/Supervisor and the BWFH Security Department. Visitors must provide photo identification and may be subject to a criminal background check as well as a physical search of their person and belongings. Visits shall be limited to one adult visitor at a time and will not exceed 30 minutes in length.

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ity and Hospital Administration
ccommodate attorney and
clergy visits during and after regular visiting hours in conjunction with custodial agency policy, procedures and guidelines. Note: United States ors under any circumstances.

NEWTON-WELLESLEY HOSPITAL
NEWTON, MASSACHUSETTS

Visitors - Visitors to patient prisoners are not allowed except in terminal cases and those cases with special consideration as approved by the law enforcement/corrections authority and Hospital Administration in consultation with Public Safety.

Hospital Course Cont.

- Despite three pressors, broad spectrum antibiotics, steroids, CVVH, and maximum ventilatory support, his shock and hypotension continue to worsen
- His HCP notes difficulty obtaining permission from the superintendent for his remaining family members (patient's brother, son, and daughter) to visit, prompting the medical team to reach out on her behalf
- Visits are limited to 20 minutes except for the HCP
- Ultimately, in the setting of ongoing declining clinical status, the patient is transitioned to comfort measures and passes away an hour later

Autopsy

- Massachusetts law dictates that death of any patient who was in custody (including in the hospital), must be reported to the medical examiner
- Though family agrees to autopsy, they request it be completed by the BWH hospital instead of the medical examiner
- After extensive discussion with the office of medical examiner, despite the family request, they state that if the medical examiner deems an autopsy is necessary, their office will need to complete it

Challenges with Caring For Incarcerated Patients

- Constraints on advocacy and dealing with an opaque system (the correctional system)
- Whose responsibility is it to navigate the administrative process
- Limitations in allowing family visits and comforts, even near the end of life
- These and numerous other challenges can lead to distrust between the care team, patient, and family, and lead to suboptimal care near the end of life

- What other challenges have you struggled with?

Source: Panozzo S, Bryan T, Collins A, Marco D, Lethborg C, Philip JA. Complexities and constraints in end-of-life care for hospitalized prisoner patients. *Journal of Pain and Symptom Management*. 2020;60(5). doi:10.1016/j.jpainsymman.2020.05.024

Conclusion

- Ultimately, the medical examiner declines autopsy, and the patient is referred back to BWH for autopsy
- Anatomic autopsy did not clearly identify his underlying disease
- Lymph node and other tissue biopsy information is still pending

Learning Points

- Diffuse lymphadenopathy has a broad differential, including infectious, neoplastic, autoimmune, and toxic etiologies
- Leukemoid reactions though traditionally associated with infections, can also happen in response to neoplasms without being directly due to the neoplasm
- Corticosteroids alone have shown some efficacy in treating ALL, Hodgkin's lymphoma, non-Hodgkin's lymphoma, and multiple myeloma
- Care for incarcerated patients, especially around end of life, is complex with multiple challenges, partially due to policies outside of physician control