

Let's not be rash...

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Internal Medicine, PGY-3

Dr. Seuss

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La Jolla, California 92037

Dear Children of Troy:

Read!

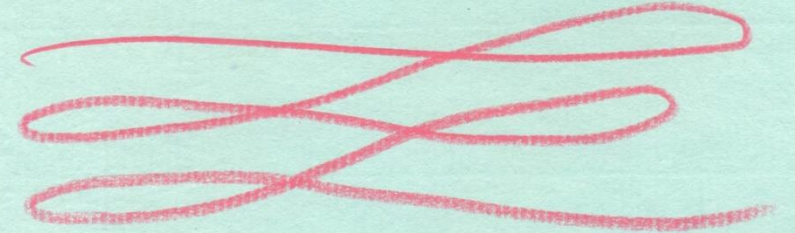
Read!

READ!

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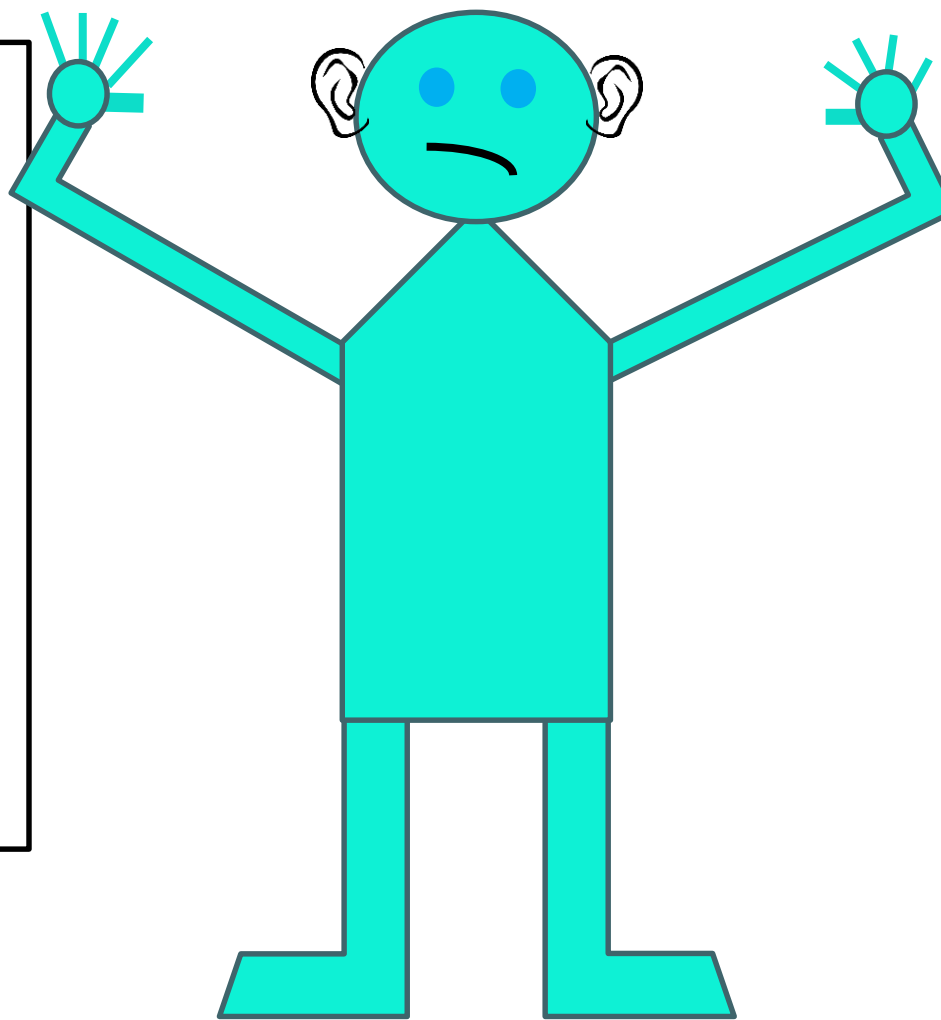
READ!



That's the advice
of your good friend, Dr. Seuss

Problem List

- Myelodysplastic syndrome

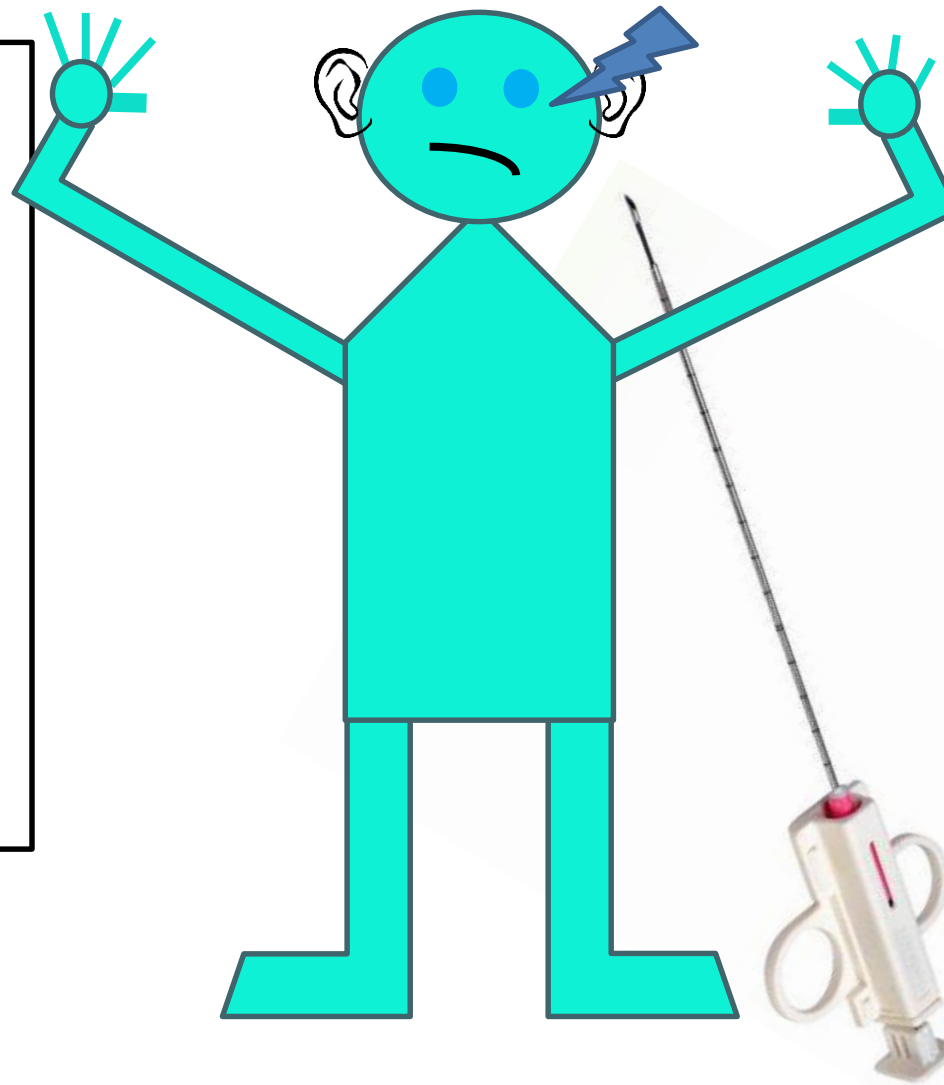


Problem List

- Myelodysplastic syndrome
- Giant cell arteritis

Treatments

- Prednisone 60 mg



-6 weeks

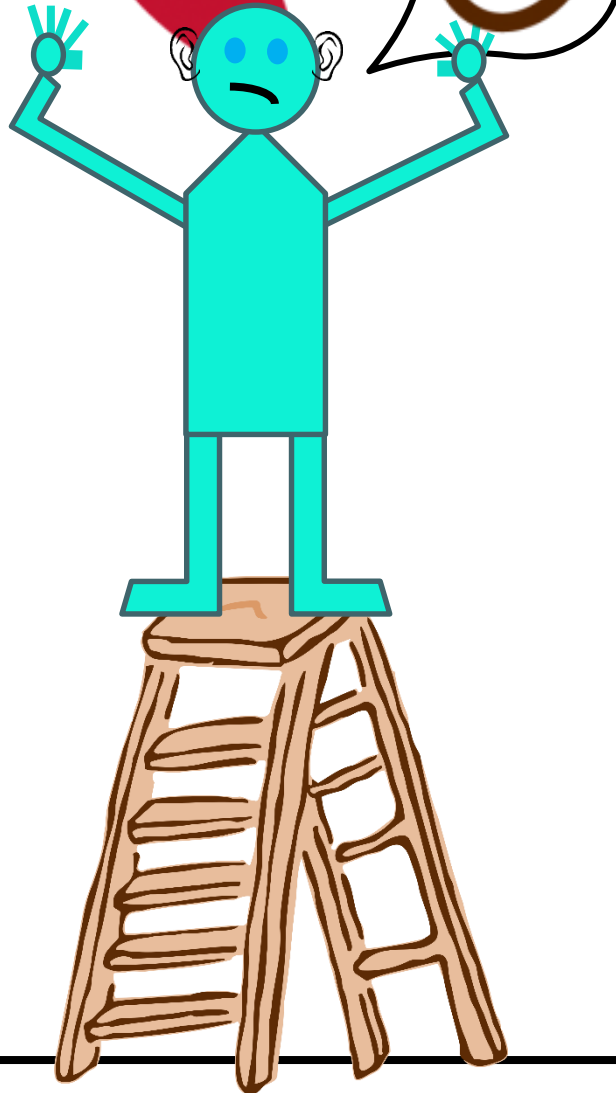


Problem List

- Melodysplastic syndrome
- Giant cell arteritis

Treatments

- Prednisone 60 mg



-6 weeks

-4 weeks

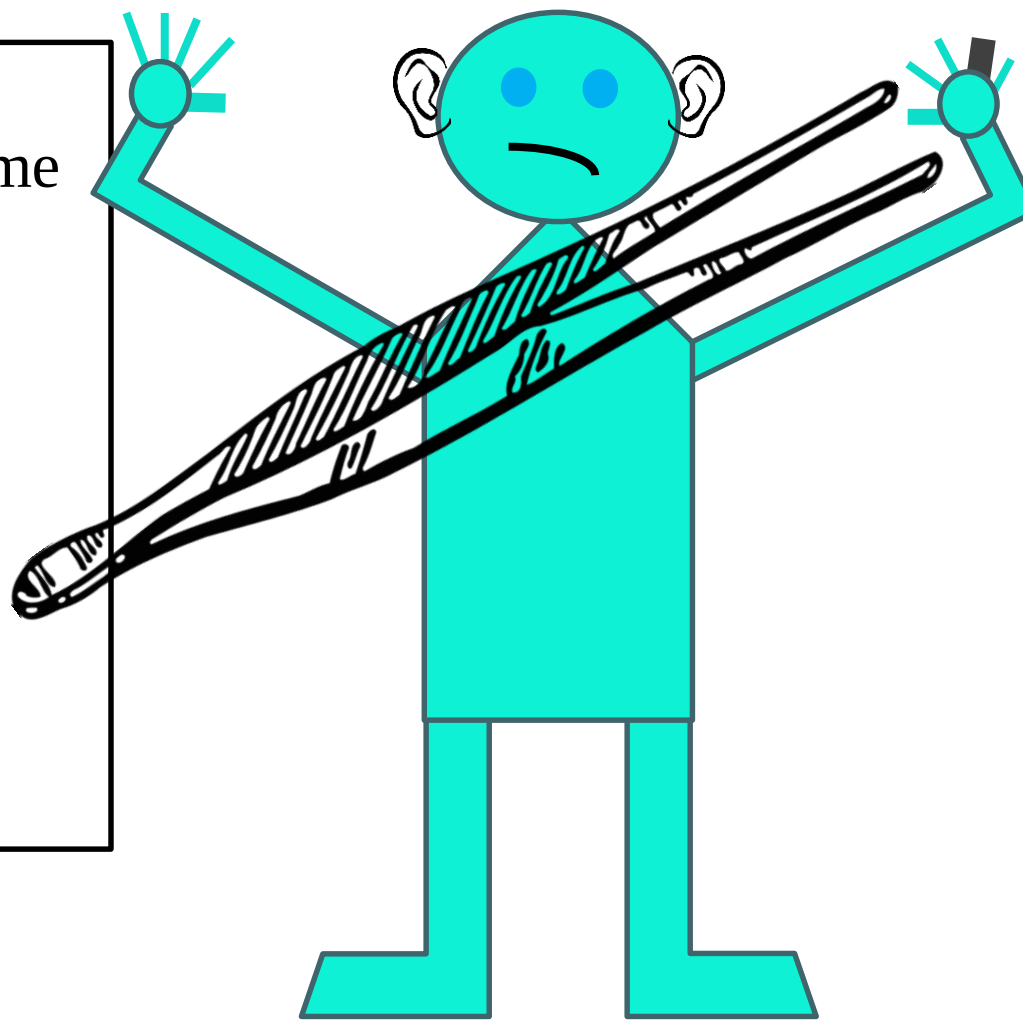
Dx of
GCA

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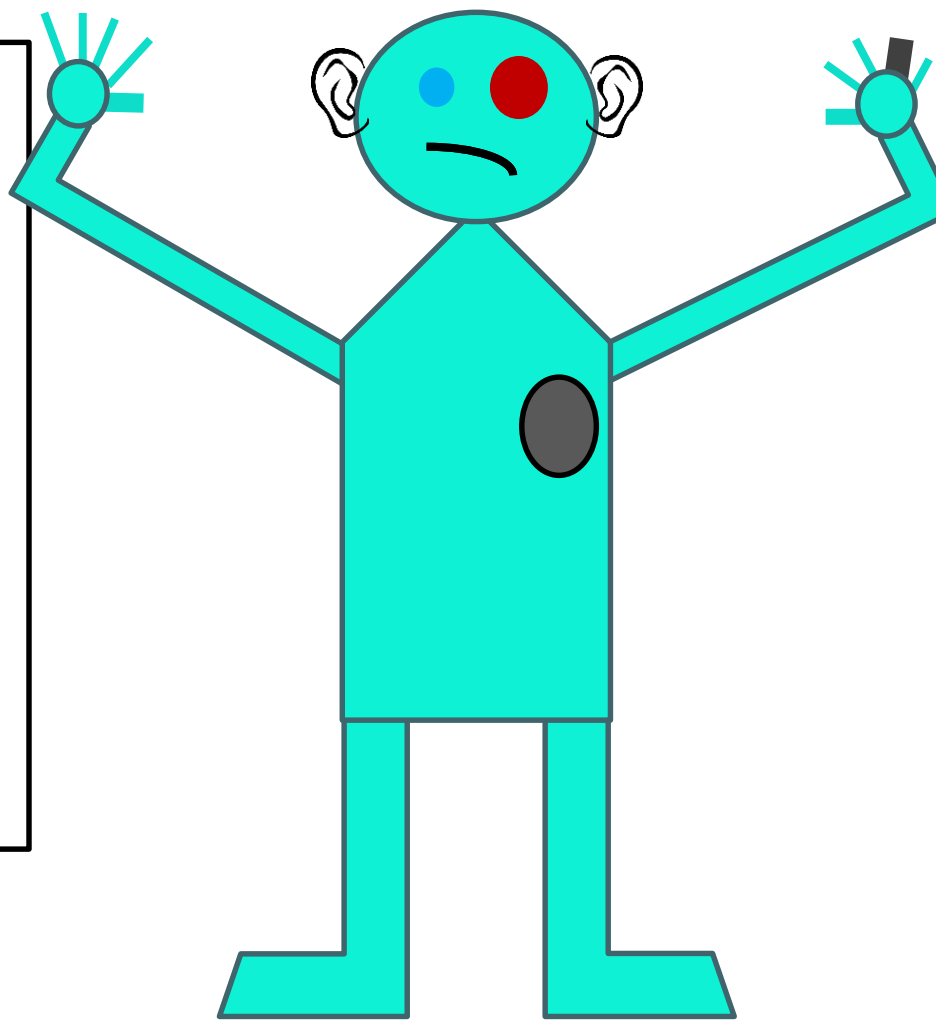
Dx of
GCA

Problem List

- Myelodysplastic syndrome
- Giant cell arteritis

Treatments

- Prednisone 60 mg



-6 weeks

-4 weeks

-2 weeks

Dx of
GCA

Cut
finger

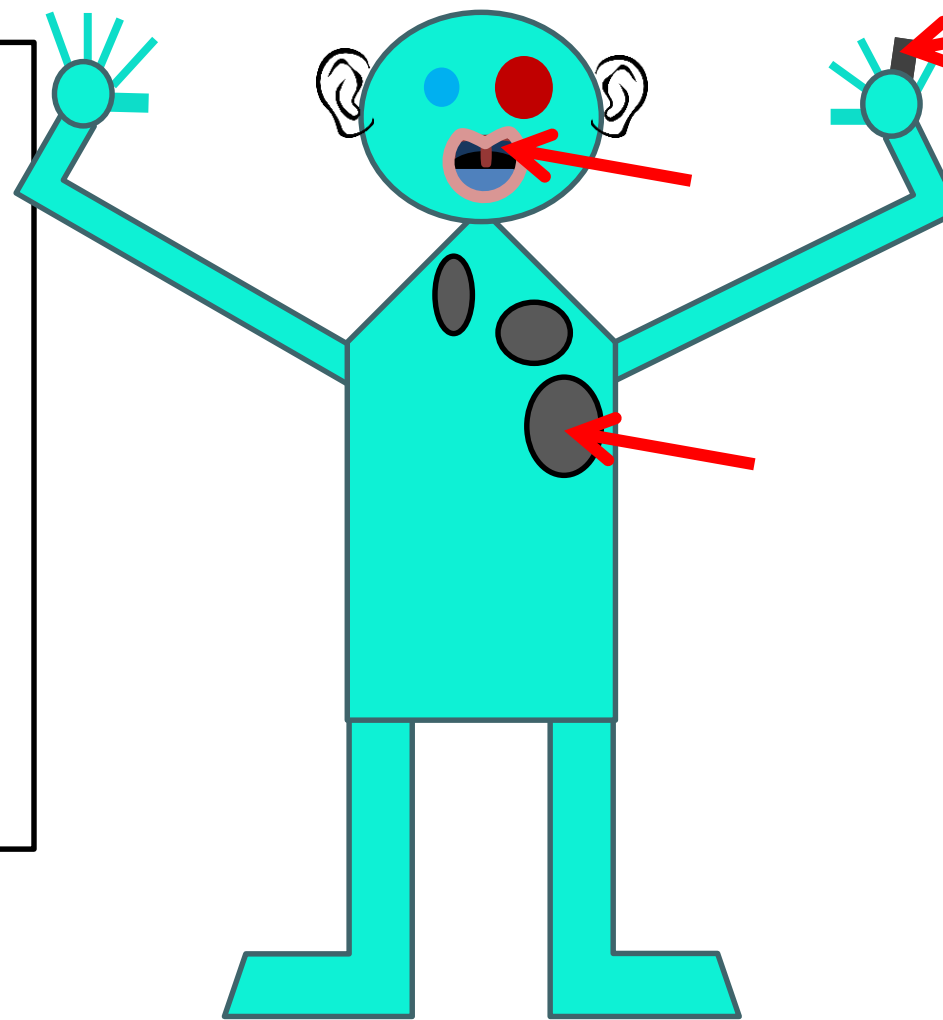


Problem List

- Myelodysplastic syndrome
- Giant cell arteritis
- Bilateral necrotic skin lesions
- Left fourth finger necrosis
- Maxillary mucosal necrosis
- Fever

Treatments

- Prednisone 60 mg
- Vancomycin and piperacillin/tazobactam



-6 weeks

-4 weeks

-2 weeks

Presents
to OSH

Dx of
GCA

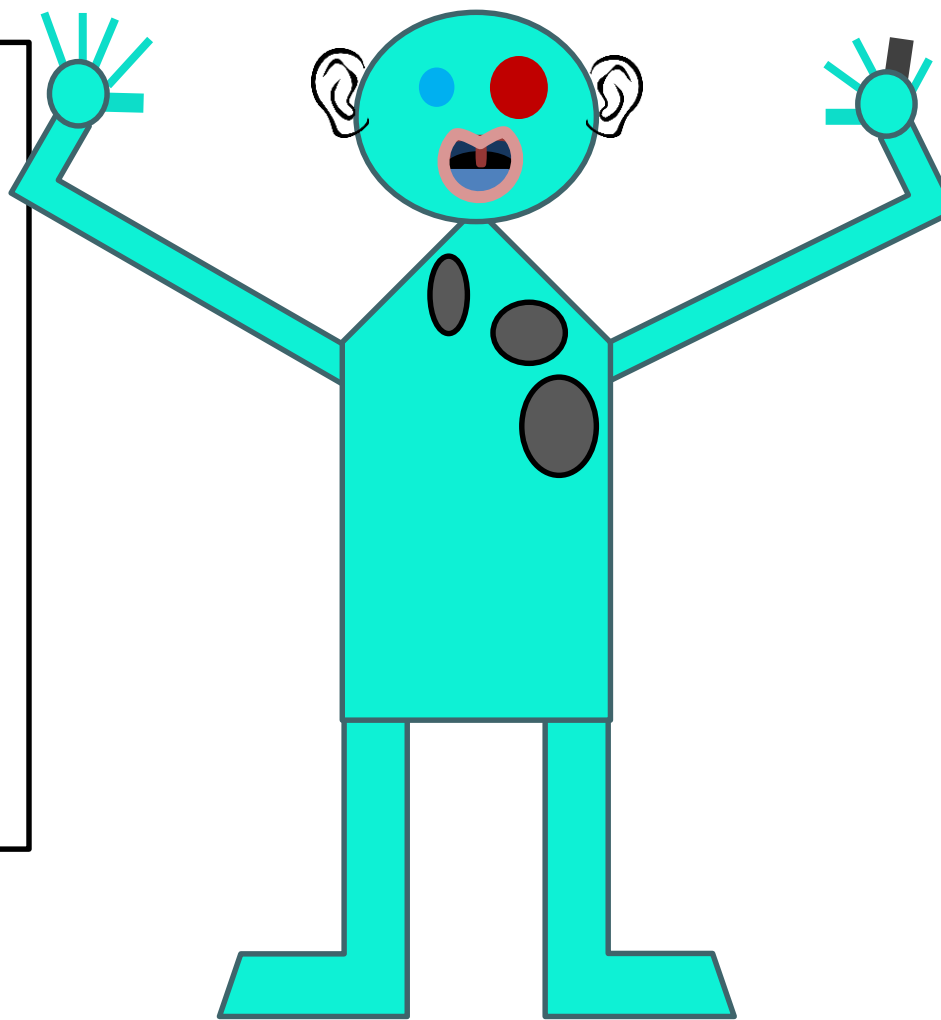
Cut
finger

Eye
pain



Problem List

- Myelodysplastic syndrome
- Giant cell arteritis
- Bilateral necrotic skin lesions
- Left fourth finger necrosis
- Maxillary mucosal necrosis
- Fever



Treatments

- Prednisone 60 mg → 40 mg
- ~~Vancomycin and piperacillin/tazobactam~~
- Voriconazole
- Daptomycin and meropenem

-6 weeks

-4 weeks

-2 weeks

Presents
to OSH

First
admit

Dx of
GCA

Cut
finger

Eye
pain

Broad
abx



Initial work-up

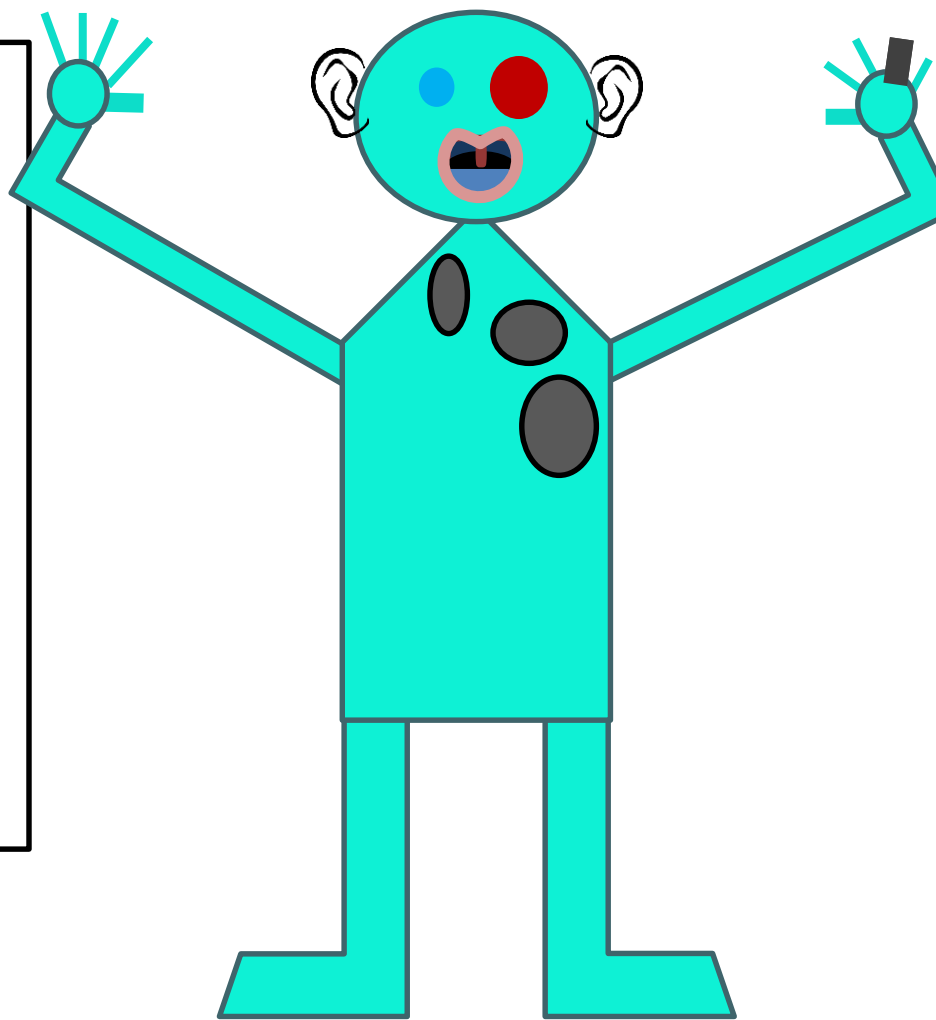
WBC	6.5x10 ⁹ /L	Hepatitis A, B, and C	Negative
Hemoglobin	8.5 g/dl	HIV	Negative
Platelets	132,000/ml	Rheumatoid factor	POSITIVE
Ferritin	1555 ng/ml	Hemoglobin A1c	6%
Vitamin B12	1274 pg/ml	Beta-2 glycoprotein IgM	POSITIVE
Absolute reticulocyte count	0.027	Cardiolipin IgM	Negative
Multiple sets of blood cultures	No growth	ESR	115 mm/hr
Total IgG	WNL	RPR	Negative
Cryoglobulins	Negative	Liver function tests	WNL
ANCAs	Negative	Histoplasma antibody	Negative
ANA	Negative	Aspergillus galactomannan	Negative
Bone marrow biopsy	Hypocellular	Protein C and S activity	WNL
Herpes simplex virus IgM	POSITIVE	HSV culture of mouth lesions	Negative

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Treatments

- Prednisone 60 mg
- ~~Vancomycin and piperacillin/tazobactam~~
- Voriconazole
- Daptomycin and meropenem
- Heparin drip
- Valacyclovir



-6 weeks

-4 weeks

-2 weeks

Presents
to OSH

First
admit

Dx of
GCA

Cut
finger

Eye
pain

Broad
abx

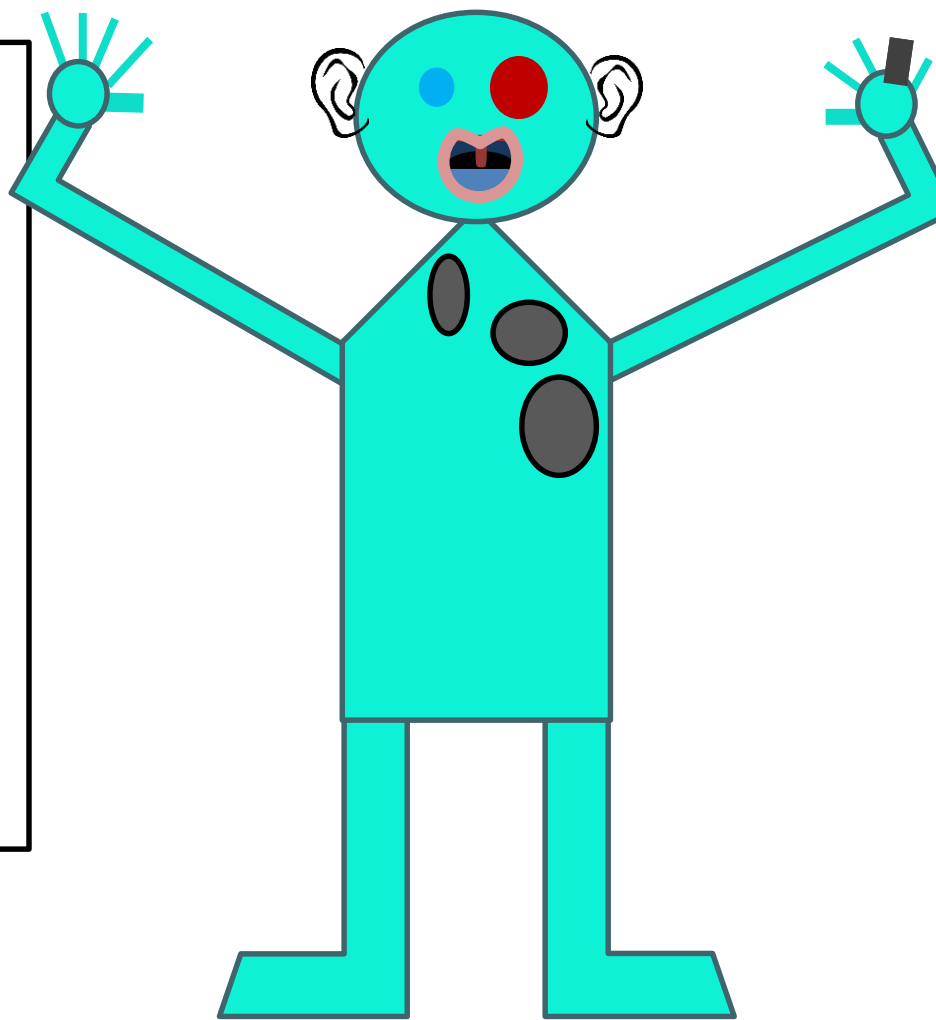


Problem List

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-6 weeks

-4 weeks

-2 weeks

Presents
to OSH

First
admit

Hospital
transfer

Dx of
GCA

Cut
finger

Eye
pain

Broad
abx

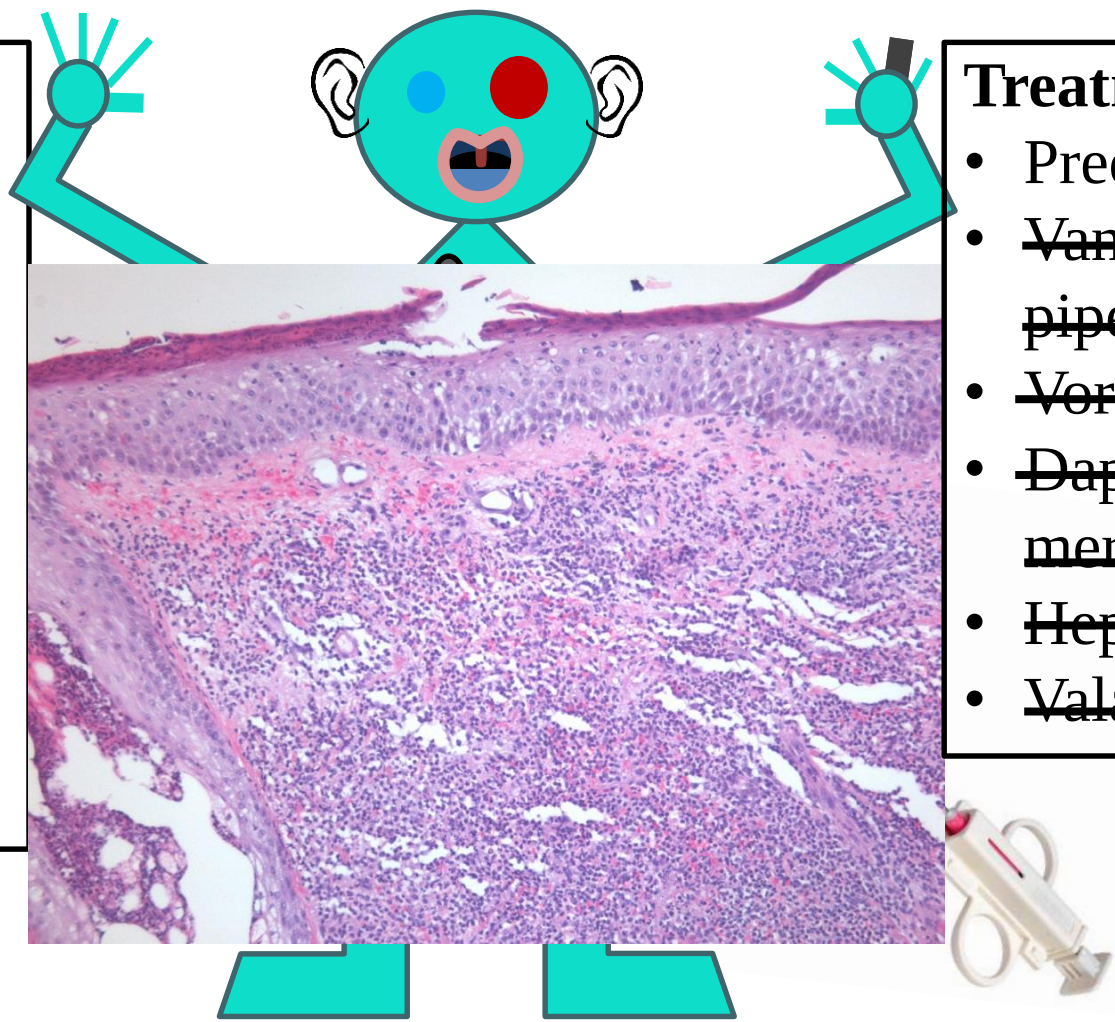
Heparin





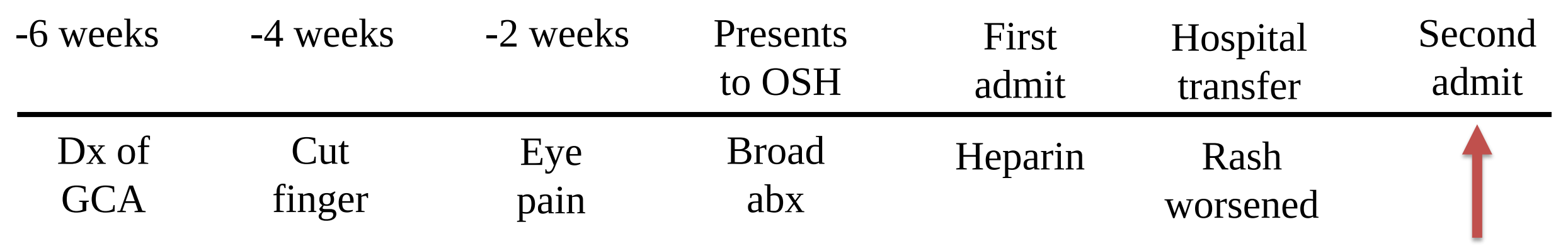
Problem List

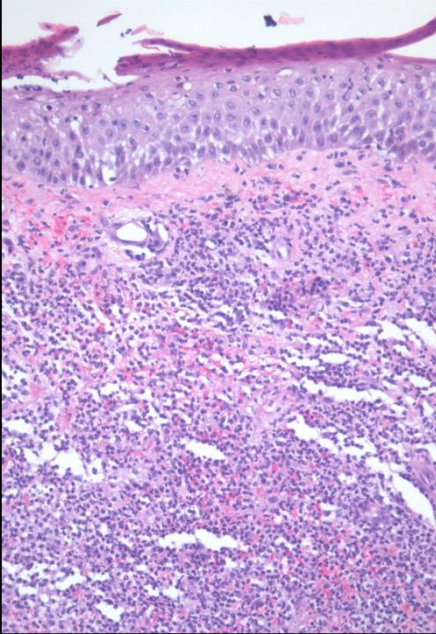
- Myelodysplastic syndrome
- Giant cell arteritis
- Left fourth finger necrosis
- Bilateral necrotic skin lesions
- Maxillary mucosal necrosis
- Fever
- Left conjunctival erosion
- Neutrophilic dermatitis



Treatments

- Prednisone 60 mg
- ~~Vancomycin and piperacillin/tazobactam~~
- ~~Voriconazole~~
- ~~Daptomycin and meropenem~~
- ~~Heparin drip~~
- ~~Valacyclovir~~





Problem List



PMHx

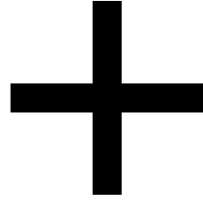
MDS

GCA



Despite steroids, antimicrobials, and anticoagulation





Malignancies

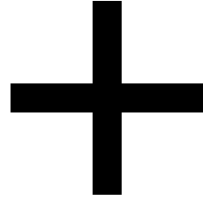
- Sweet syndrome
- Leukemia cutis
- Paraneoplastic syndromes

Atypical infections

- Sporotrichosis
- Leishmaniasis
- Atypical mycobacteria
- *M. tuberculosis*
- Cutaneous anthrax
- Nocardia
- Tularemia

Rheumatologic conditions

- Medium and large vessel vasculitides



Malignancies

- Sweet syndrome
- Leukemia cutis
- Paraneoplastic syndromes

Atypical infections

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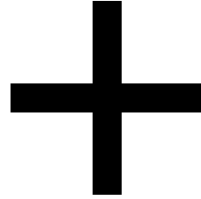
Rheumatologic conditions

- Medium and large vessel vasculitides

Additional work-up

- Computed tomography angiography (CTA) head and neck
- Positive emission tomography (PET scan)
- Computed tomography (CT) of chest, abdomen and pelvis
- Universal polymerase chain reaction (PCR) for bacterial and viral infections

SPEP	Normal
ESR	116 mm/hr
MMA	0.26 umol/L
LDH	402 U/L
Rapid plasma reagin	Negative
Repeat blood cultures	No growth
CRP	134 mg/L
Zinc	62 ug/mdl
Vitamin D, 25-hydroxy	31.8 ng/ml
Ferritin	1980 ng/mL
Cryptococcus antigen	Negative



Malignancies

- Sweet syndrome
- ~~Leukemia cutis~~
- ~~Paraneoplastic syndromes~~

?

~~Atypical infections~~

- ~~Sporotrichosis~~
- ~~Leishmaniasis~~
- ~~Atypical mycobacteria~~
- ~~*M. tuberculosis*~~
- ~~Cutaneous anthrax~~
- ~~Nocardia~~
- ~~Tularemia~~

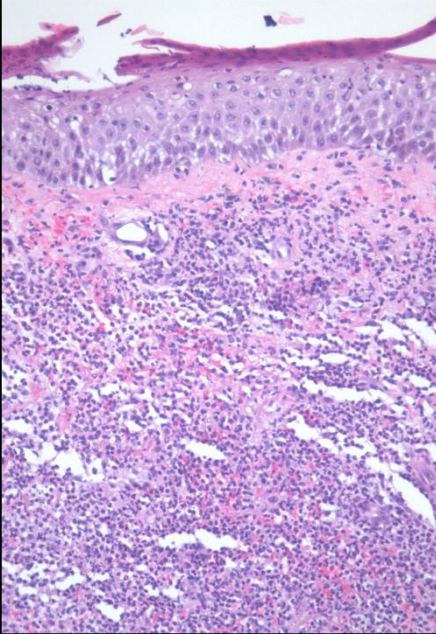
~~Rheumatologic conditions~~

- ~~Medium and large vessel vasculitides~~

A Lesson Learned from **Dr. Seuss**...

“[We] puzzled, and
puzzled, till [our]
puzzlers were sore.
And then, [we]
thought of something
[we] hadn’t before...”





Problem List



PMHx
MDS
GCA



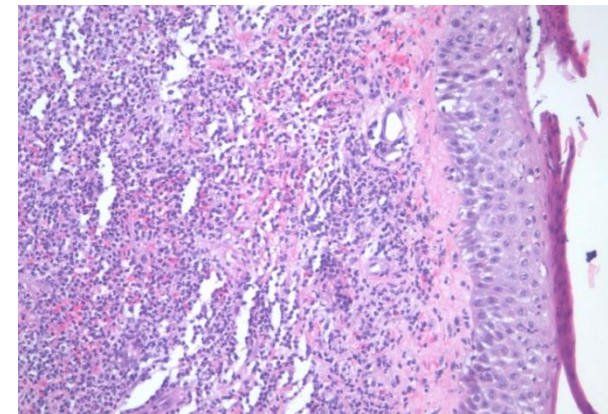
Despite steroids, antimicrobials, and anticoagulation



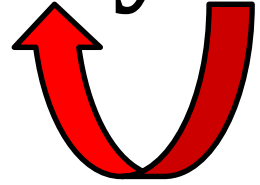
Pyodermatitis-Pyostomatitis Vegetans (PD-PSV)

- Neutrophilic dermatosis subtype
- Characterized by rash involving the skin and mucosa
- First described in 1898
- Strong association with inflammatory bowel disease (IBD)
- Clinical dx supported by histology
- High dose steroids are first-line tx

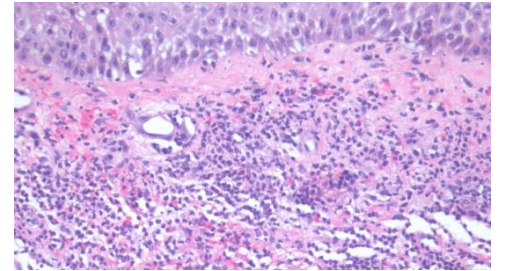
Pyodermite végétante



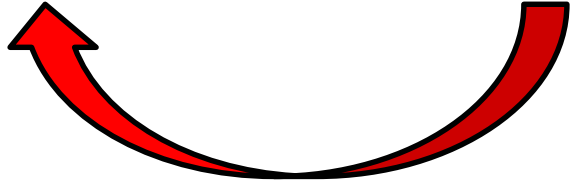
Pyodermatitis-Pyostomatitis Vegetans



- **Pyo=pus (neutrophils)**



Pyodermatitis-Pyostomatitis Vegetans



- **Pyo=pus (neutrophils)**
- **Dermatitis=inflammation of skin**



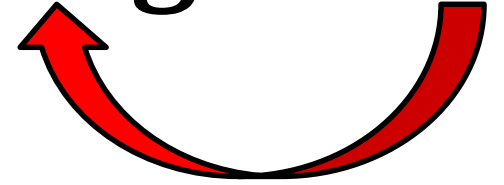
Pyodermatitis-Pyostomatitis Vegetans



- **Pyo=pus (neutrophils)**
- **Dermatitis=inflammation of skin**
- **Stomatitis=inflammation of mucosa**



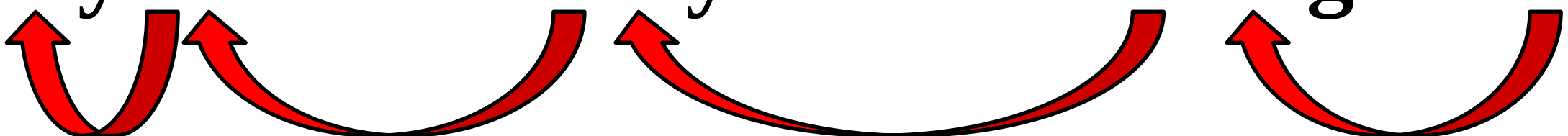
Pyodermatitis-Pyostomatitis Vegetans



- **Pyo=pus (neutrophils)**
- **Dermatitis=inflammation of skin**
- **Stomatitis=inflammation of mucosa**
- **Vegetans=proliferative growths**



Pyodermatitis-Pyostomatitis Vegetans



- Pyo=pus (neutrophils)
- Dermatitis=inflammation of skin
- Stomatitis=inflammation of mucosa
- Vegetans=proliferative growths



A Brief Primer on Neutrophilic Dermatoses

- Sweet syndrome
- Pyoderma gangrenosum

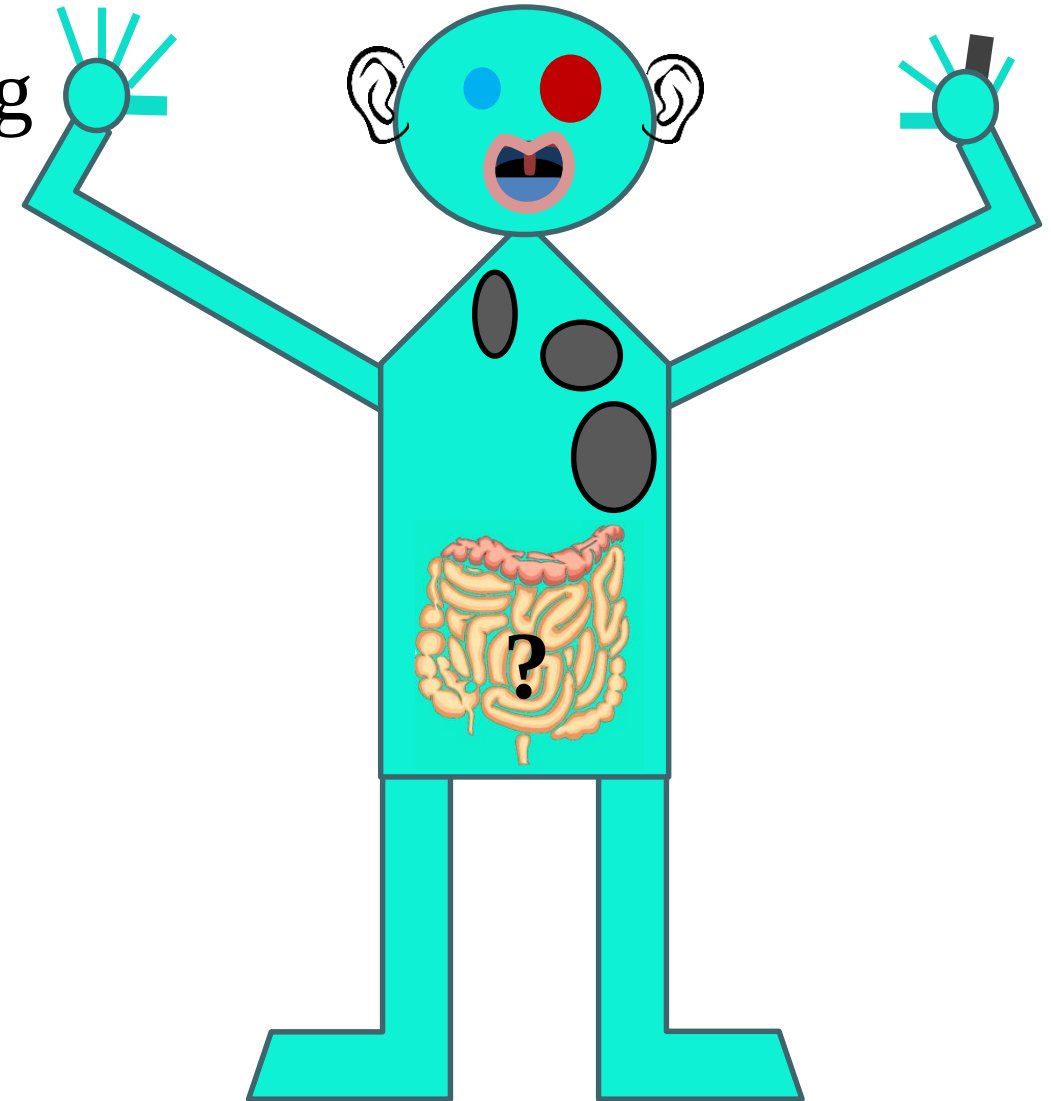
To name a few....

- Pustular psoriasis
- Drug-induced/acute generalized exanthematous pustulosis
- Keratoderma blennorrhagicum
- Sheddon-Wilkinson disease
- IgA pemphigus
- Amicrobial pustulosis of the hands
- Infantile acropustulosis
- Neutrophilic dermatosis of the dorsal hands
- DIRA syndrome
- Behcet's disease



What happened to our patient?

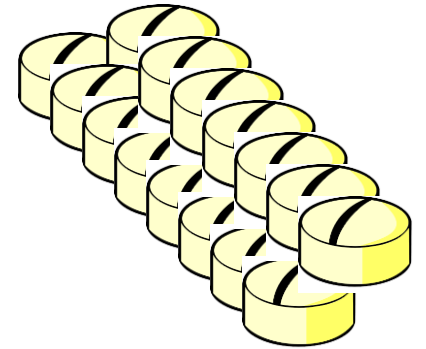
- Prednisone was increased to 1 mg/kg (prednisone 70 mg)
- Denied any symptoms of inflammatory bowel disease
- Fecal calprotectin was positive
- Referred to gastroenterology





Lessons to take away

- Puzzle and puzzle, till your puzzler is sore
- If you don't have enough of the right treatment, it doesn't work
- If there's neutrophilic inflammation without infection, think neutrophilic dermatoses
- PD-PSV involves skin and mucosa and is highly associated with IBD



Thank You to...

- Our patient, who graciously gave permission for his story and pictures to be shared.
- Our consulting teams: dermatology, ophthalmology, rheumatology, oral maxillofacial surgery, plastic surgery, and infectious diseases.
- Everyone who contributed to the care of the patient during his hospitalizations and follow-up.

References

- Clark, L.G. et al., Pyostomatitis vegetans (PSV)-pyodermatitis vegetans (PDV): A clinicopathologic study of 7 cases at a tertiary referral center. *J Am Acad Dermatol*. 2016;75(3):578-584. doi:10.1016/j.jaad.2016.03.047
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- Rosmaninho, A. Carvalho, S. Neutrophilic Dermatoses Revisited. *EMJ Dermatol*. 2014;2:77-85.

References for Outside Images

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Clip Art References

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- <https://konopcompanies.wordpress.com/cropped-christmas-lights-border-clip-art-png/>
- <https://openclipart.org/detail/280986/roughly-drawn-stepladder>
- <https://openclipart.org/detail/263267/tweezers>
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- <http://clipart-library.com/gut-cliparts.html>
- Illustrationsof.com, #1047791
- http://clipartbarn.com/pill-clipart_9718/
- <http://clipart-library.com/wink-smiley-face.html>

Another Case of PD-PSV



More Information about PD-PSV

- More strongly associated with ulcerative colitis (UC)
- Seems to be a correlation with poorly controlled IBD
- PD-PSV may predate IBD by up to 2 years (15% of patients)
- Other treatments tried include azathioprine, mesalamine, infliximab and dapsone
- Improvement with treatment of underlying IBD is also reported