

Grand Rounds

“Periorbital Swelling”



Kevin Lowder, MD – PGY4

08/9/2019

Patient Presentation

CC

Left periorbital swelling

HPI

- 63 yo WF presents with 6 months of progressive swelling around left eye and left side of face. Originally seen by outside provider who treated with steroids with no improvement. Seen by another provider who tried PO Bactrim x 10 days with no improvement. A third provider tried PO Amoxicillin and topical Zylet with no improvement. A fourth tried PO Doxycycline and prednisone 40mg with minimal improvement. This provider then tried to drain the lesion and ordered a CT of the orbits. The patient developed diffuse body rash (hives) and was then referred to Oculoplastics.



History (Hx)

Past Ocular Hx: as described in HPI

Past Medical Hx: Osteopenia, Hyperlipidemia

Fam Hx: Glaucoma (father), Cancer (mother)

Meds: Multivitamin, fish oil



History (continued)

Allergies: Doxycycline (rash), Prednisone (rash)

Social Hx: no drugs or EtOH

RoS: Diffuse rash (hives)



Physical Exam

	OD		OS
VAcc	20/20		20/25+3 PH
Pupils	Round. Brisk.	No rAPD	Round. Brisk.
IOP	14 mmHg		20 mmHg
EOM	full		full
CVF	full		Full
Adnexa	WNL		Large (2 cm x 2cm) subcutaneous mass along lateral orbital wall, firm, non-mobile, painful to touch. Prolene suture present. Significant upper and lower lid edema with palpebral fissure ~1-2mm due to mechanical ptosis. 2 cm firm painless submandibular node.



Physical Exam

Anterior Exam

SLE	OD		OS
Lids	WNL		Severe edema
Conj/Sclera	WNL		Mild Conj injection
Cornea	WNL		WNL
Ant Chamber	WNL		Normal depth
Iris	WNL		WNL
Lens	Clear		Clear
Vitreous	WNL		No view



Physical Exam

Posterior Exam

Fundus	OD		OS
Optic Nerve	WNL		WNL
Macula	WNL		WNL
Vessels	WNL		WNL
Periphery	WNL		WNL





CT Orbits

IMPRESSION:

There are 3 masses seen around the left orbit. The mass along the superior orbital rim demonstrates **cortical bone disruption and destruction**. The 2 masses along the lateral orbital rim appear more well-circumscribed with no bony involvement. The largest mass presumably representing the palpable abnormality demonstrates **central necrosis**. Biopsy recommended.



Assessment:

63 yo WF with periorbital swelling and 3 distinct masses as seen on CT Orbits with bony destruction and central necrosis:

DDx Includes:

1. Neoplasm/Malignancy
2. Inflammatory Disorder
3. Atypical Infection (fungal, etc.)
4. ~~Vascular malformation~~



Plan

Orbitotomy with biopsy/exploration OS:

- Frozen sections
- Fresh sample (Flow cytometry & immunohistochemical staining)



Biopsy Results

Frozen sections: *“Atypical lymphoid lesion”*



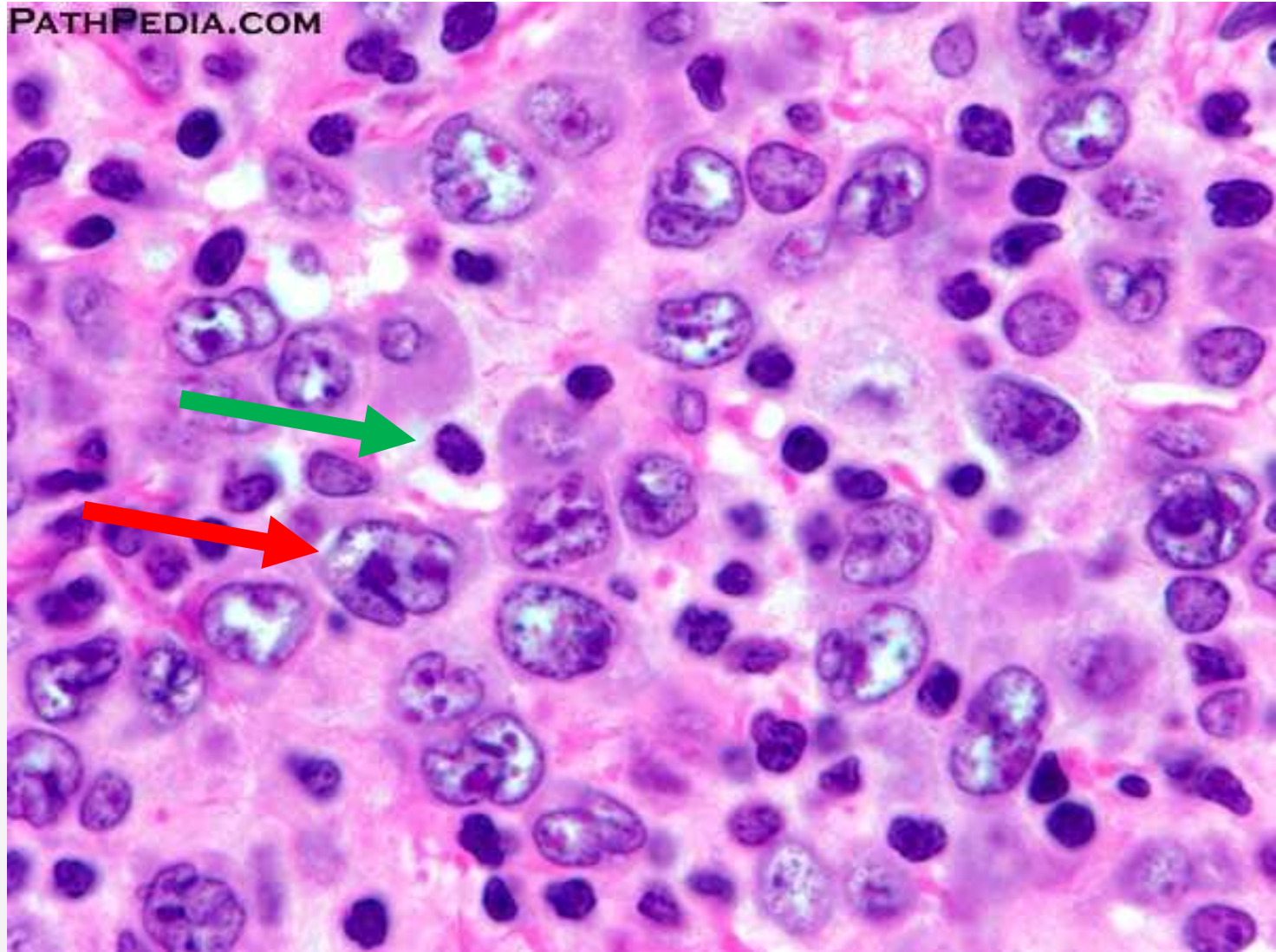
The atypical lymphoid infiltrate is composed of medium to large cells with irregular nuclear membranes and single to multiple prominent nucleoli. There is brisk mitotic activity consistent with a **high grade lymphoma**.



Diffuse large B-cell lymphoma, germinal center cell type



Representative Pathology Slide



Courtesy of:

https://www.pathpedia.com/education/eatlas/histopathology/lymph_node/diffuse_large_b-cell_lymphoma_nos.aspx



Discussion

Diffuse Large B Cell Lymphoma (DLBCL)

Definition/Criteria:

- At least 2x size of normal B cells
- Many different subsets based on immunohistochemical staining of biopsy

Epidemiology:

- Common in Western countries (25-30% of Non-Hodgkin Lymphoma)
- Affects all ages, but MC in elderly: median age 64
- Slightly MC in men vs women
- MC in white people



Discussion

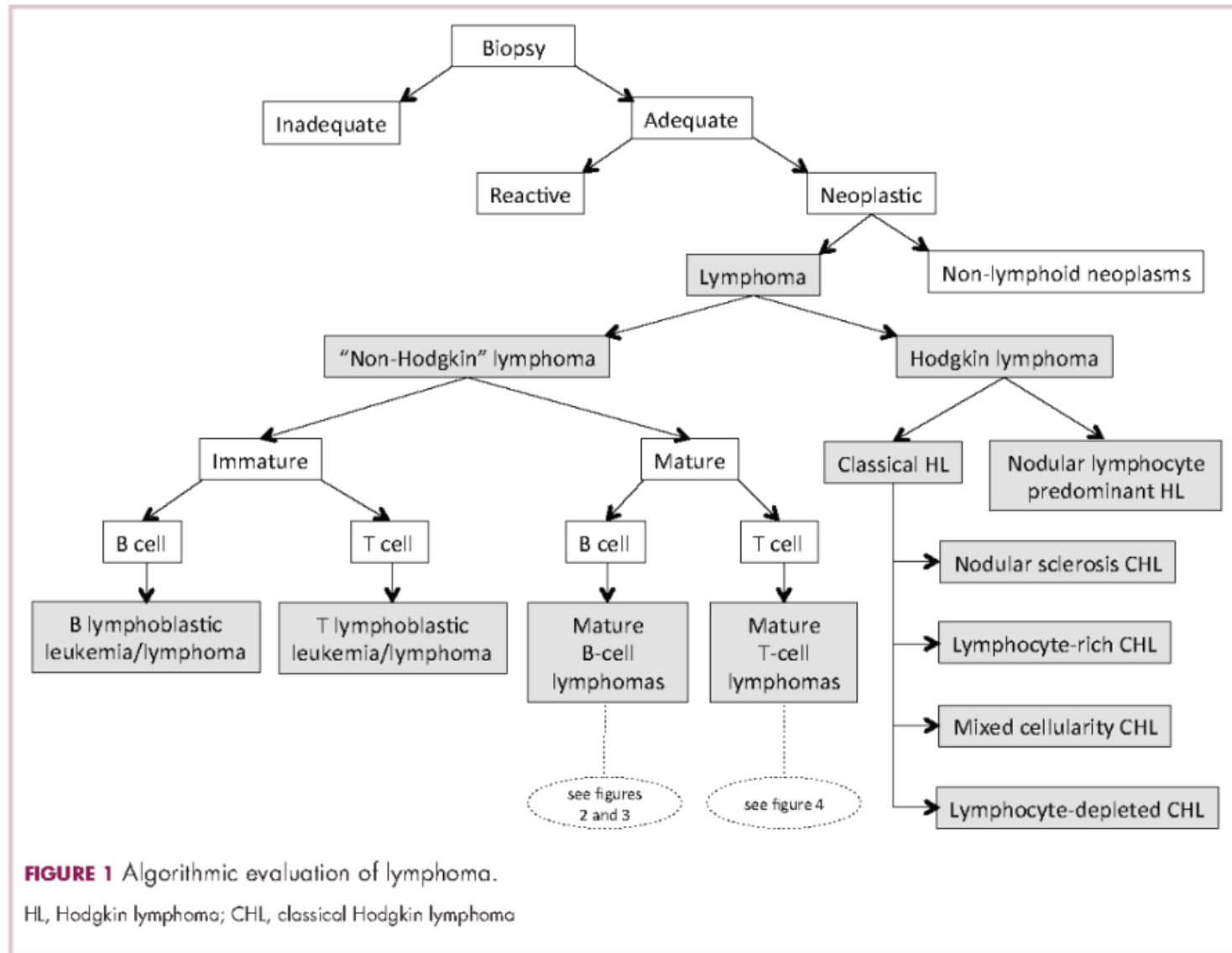
Diffuse Large B Cell Lymphoma (DLBCL)

Clinical Features:

- Often present as a single, rapidly growing nodal mass
- May be accompanied by “B symptoms”
 - Weight loss
 - Drenching night sweats
 - Fevers
- 30 – 40% are extranodal at diagnosis
- ~60% are stage III / IV at diagnosis
- Bone Marrow involved in ~27% of cases
- Overall cure rate ~67% if caught & treated in a timely manner



Discussion – Classifying Lymphoma



Follow Up

POW#1 Plan:

*Continue steroid taper.

*Scheduled PET/CT scan, LP, brain MRI.



Imaging Findings:

PET/CT:

- *Large left periorbital mass.
- *Left neck hypermetabolic LN.
- *Bulky scattered mesenteric adenopathy (5.0cm)

MRI Brain:

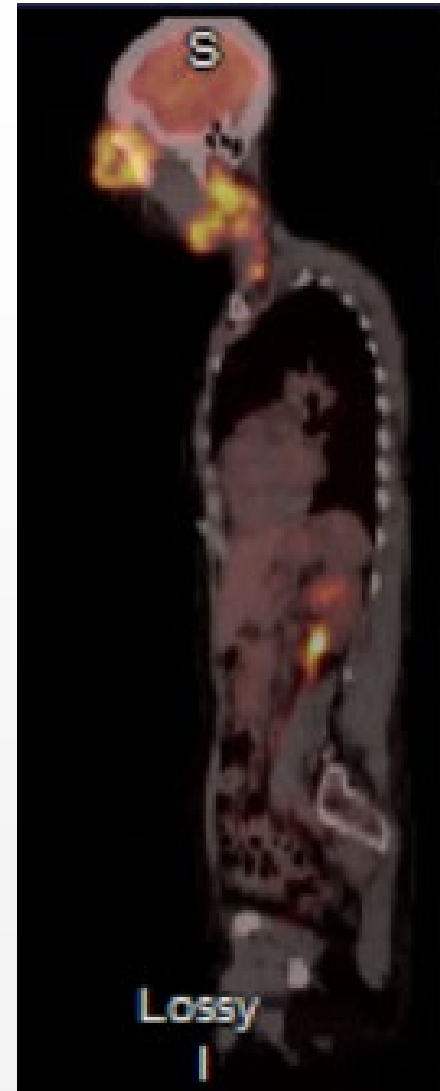
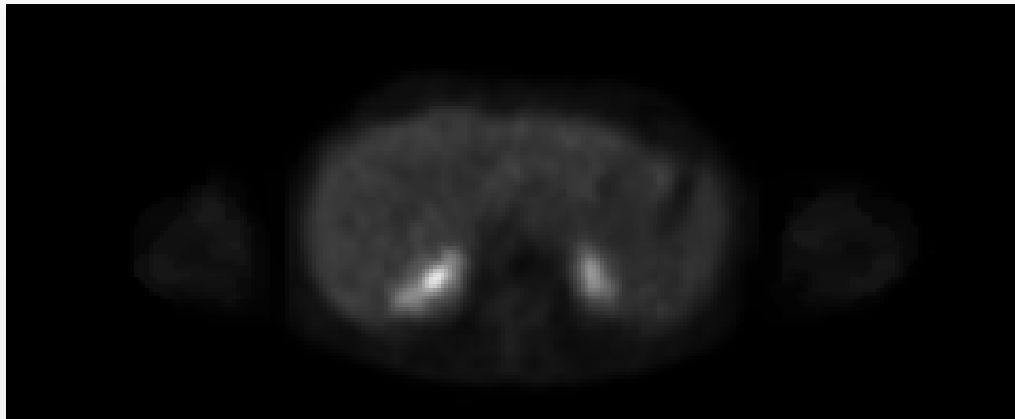
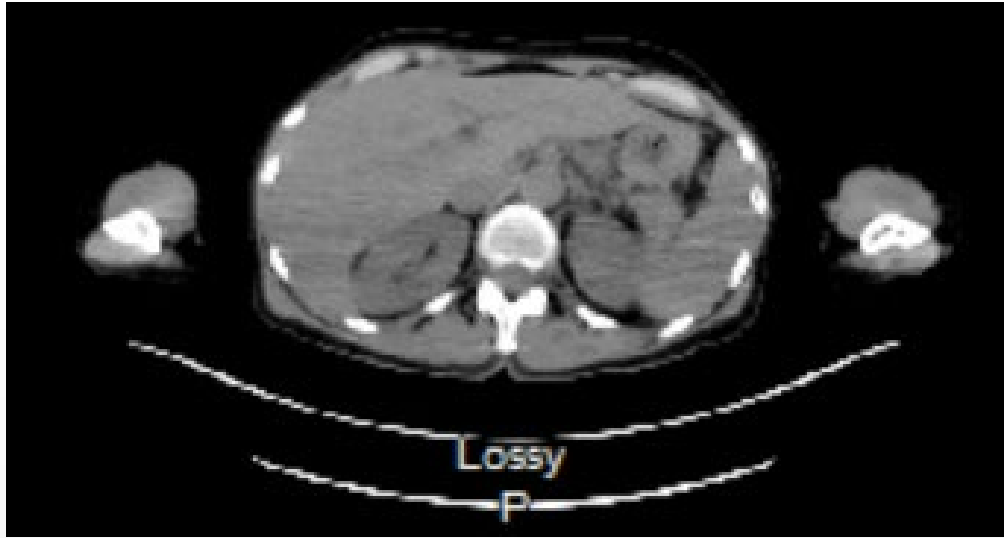
Negative for evidence of lymphoma.

LP:

No evidence of lymphoma



PET/CT



Discussion – Staging Lymphoma

Stage 1

*Only one group of lymph nodes involved (anywhere)

Stage 1E

*Extranodal, originating in a single body organ outside the lymphatic system.

Stage 2

*2+ groups of lymph nodes involved, all above diaphragm

Stage 2E

*Lymphoma started in one body organ, but is also in LN, all above the diaphragm

Stage 3

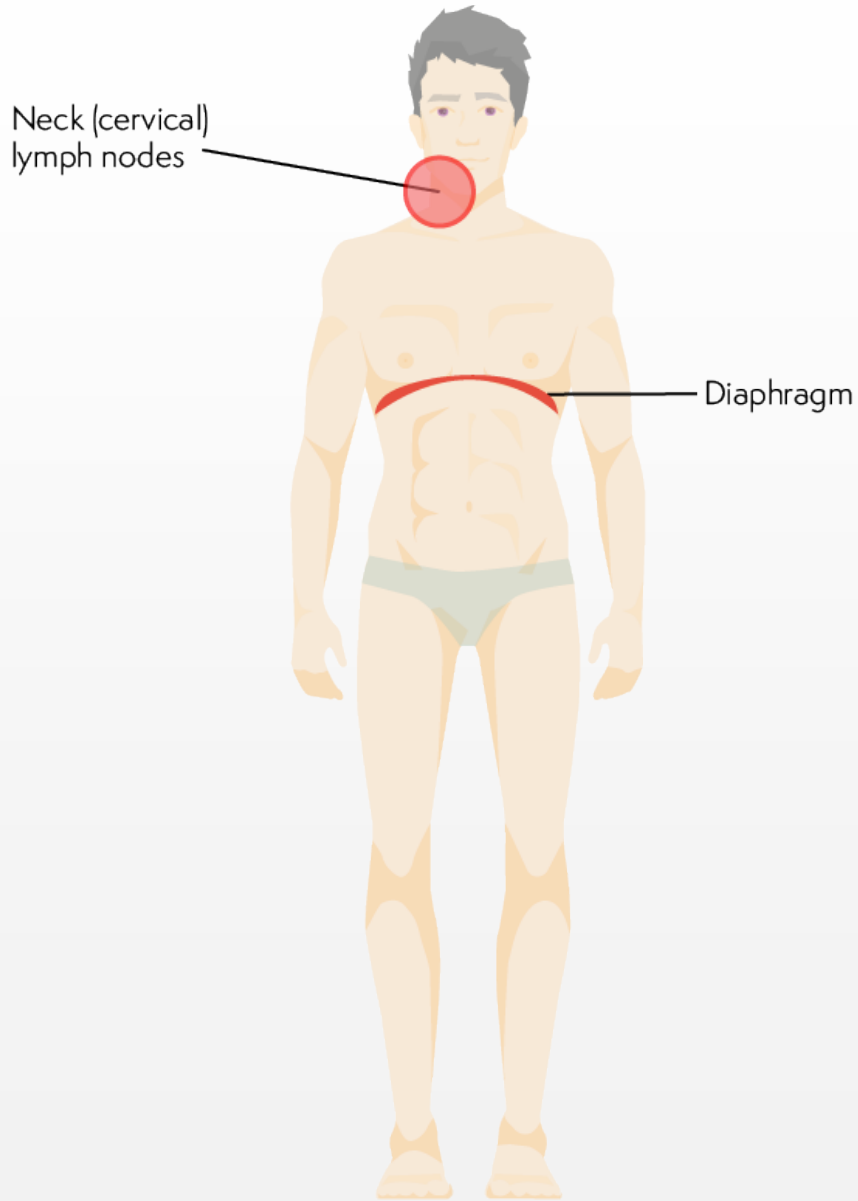
*Lymph nodes affected on both sides of diaphragm

Stage 4

*Spread from LN to organ systems



Stage 1



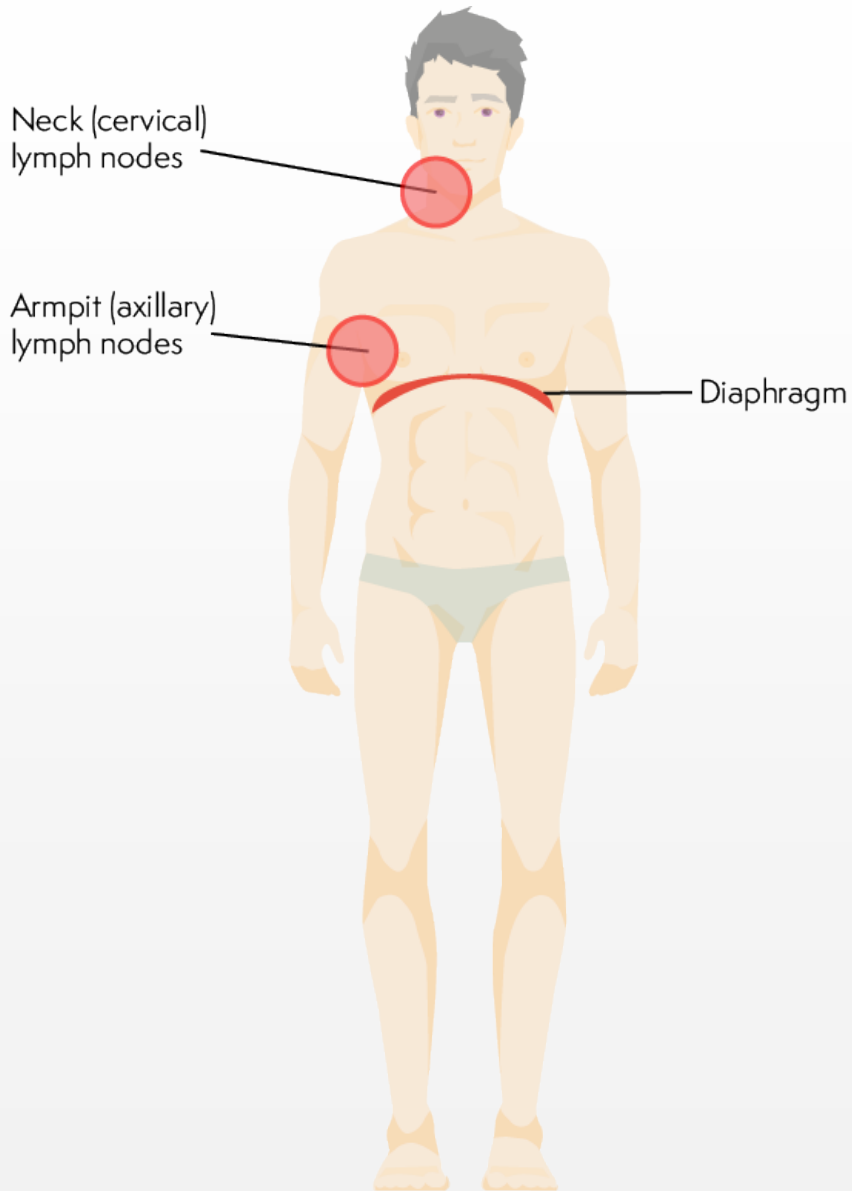
Stage 1

*Only one group of lymph nodes involved (anywhere)

Stage 1E

*Extranodal, originating in a single body organ outside the lymphatic system.

Stage 2



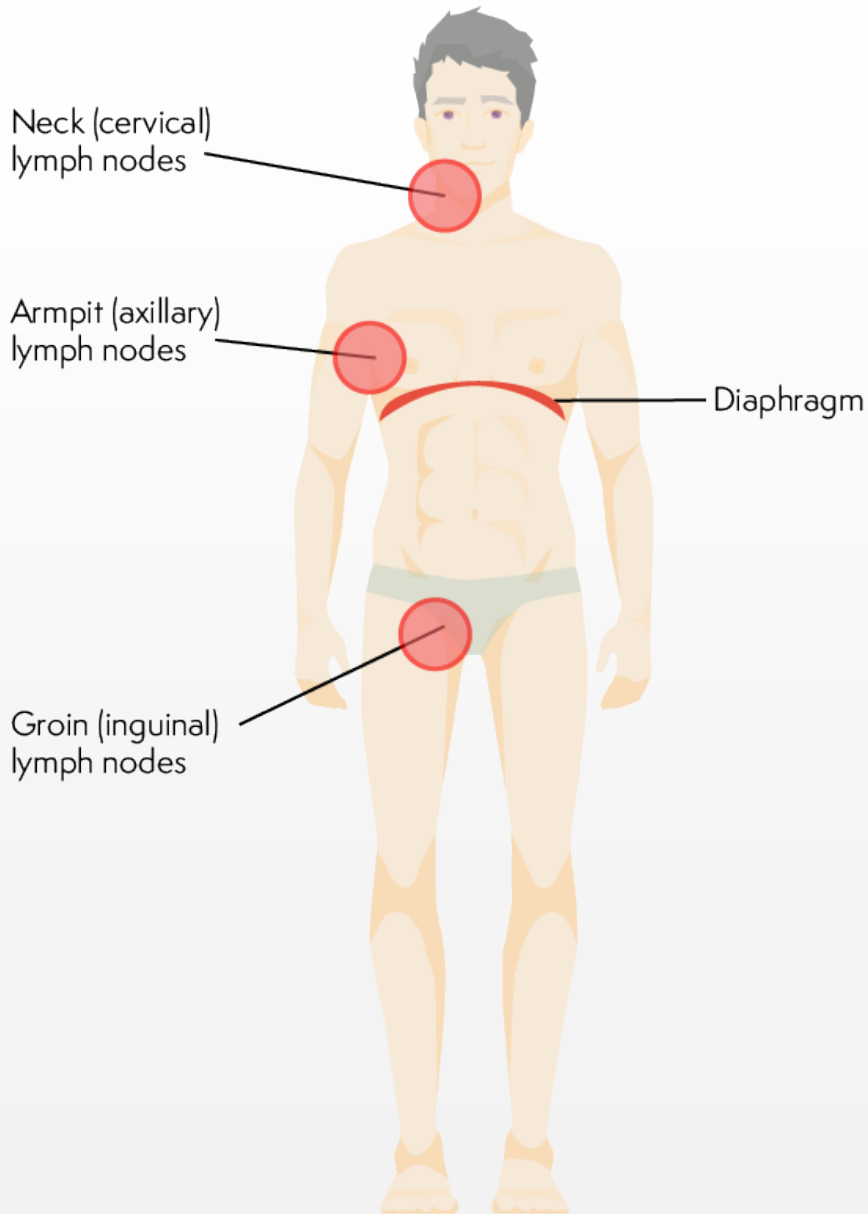
Stage 2

*2+ groups of lymph nodes involved, all above diaphragm

Stage 2E

*Lymphoma started in one body organ, but is also in LN, all above the diaphragm

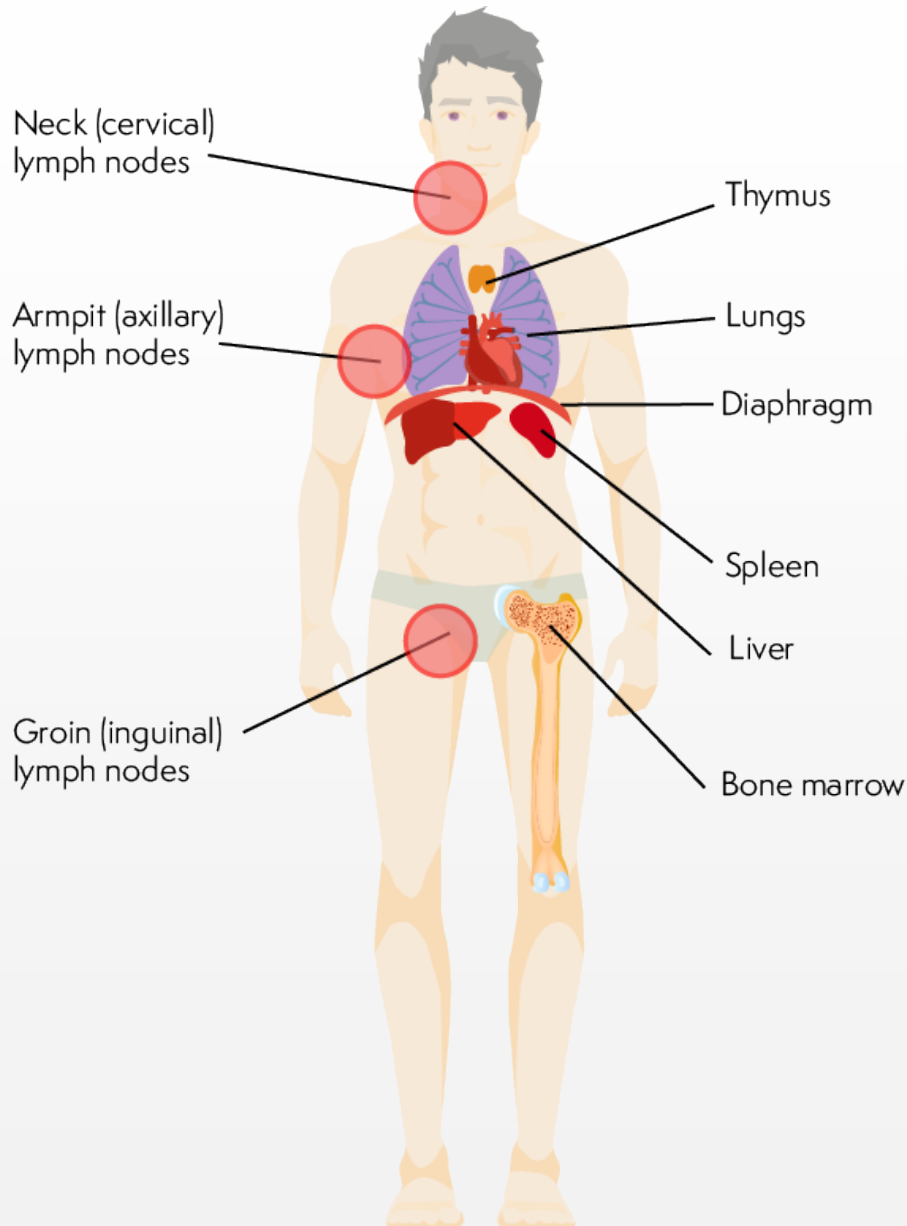
Stage 3



Stage 3

*Lymph nodes affected on both sides of diaphragm

Stage 4



Stage 4

*Spread from LN to organ systems

Discussion – Treating Lymphoma

Depends on Stage

- **Early stages** = 3 cycles of chemo + radiation
(if small enough for single field of radiation)
- **Later stages** = 6-8 cycles of chemo



Discussion – Treating Lymphoma

A decade of R-CHOP

Laurie H. Sehn

Blood 2010 116:2000–2001; doi: <https://doi.org/10.1182/blood-2010-07-293407>

Rituximab added to “CHOP” regimen:
Cyclophosphamide
Doxorubicin
Vincristine
Prednisone

**Improved 10 year progression-free rate
& overall survival by ~18% (absolute)**



Update on Our Patient



Conclusions

1. If there's persistent periorbital swelling despite multiple rounds of steroids and antibiotics, **ORDER IMAGING!**
2. Lymphoma needs a biopsy (with fresh sample), and then requires staging
3. Treatment depends on staging
 - *Chemo + Radiation vs. Chemo alone



References

American Cancer Society. "Treating B-Cell Non-Hodgkin Lymphoma." 20 June 2019, <https://www.cancer.org/cancer/non-hodgkin-lymphoma/treating/b-cell-lymphoma.html>

BCSC 2017-2018 Ophthalmic Pathology and Intraocular Tumors, pg 232

Caponetti, Gabriel, et al. "Demystifying the diagnosis and classification of lymphoma: a guide to the hematopathologist's galaxy." MD Edge Hematology & Oncology, Frontline Medical Communications Inc., 2017, <https://www.mdedge.com/hematology-oncology/article/135934/lymphoma-plasma-cell-disorders/demystifying-diagnosis-and>

Freedman A, Friedberg J. "Initial treatment of limited stage diffuse large B cell lymphoma." UpToDate, 22 Feb 2019

Lymphoma Action. "Staging of Lymphoma." <https://lymphoma-action.org.uk/about-lymphoma-tests-diagnosis-and-staging/stages-lymphoma>

Sangle, Nikhil. "Lymphoma and plasma cell neoplasms - B cell lymphoma subtypes: Diffuse large B cell lymphoma (DLBCL)." Pathology Outlines, 1 Feb 2011. <http://www.pathologyoutlines.com/topic/lymphomadiffuse.html>

Sehn, Laurie. "A decade of R-CHOP." Blood. 2010, 116:2000-2001



Special Thanks

Dr. Compton

Dr. Tomasko

