AMSER Case of the Month October 2022

47-year-old female presenting with fevers, muscle aches and a tender right cervical lymph node

Conor Rizzuto, MS4
Drexel University College of Medicine



Allegheny Health Network Dr. Matthew Hartman, M.D. Allegheny Health Network



Patient Presentation

HPI:

47 year old female with hx notable for EBV presented to the ED with a constellation of symptoms including subjective fevers, generalized muscle pains, flank pain and a right sided, painful lymph node.

She denied any rash or known tick bites but did state she was "jeeping" outdoors before symptoms began.

Physical Exam:

Vitals: BP 128/80 HR 81 RR 16 Temp 99 F SpO2 99%

Neck exam was notable for a tender, mobile cervical lymph node measuring 2 cm in diameter.



PMHx

- EBV
- Hypercholesterolemia
- Myocardial Infarction
- Scoliosis
- SVT
- Recurrent kidney stones complicated by pyelonephritis and hydronephrosis.



Initial Workup

- A CT of the abdomen and pelvis was ordered due to flank pain to assess for recurrent kidney stones which showed stones in the kidney but none in the ureter.
- Significant findings included splenomegaly at 16 cm with acute infarcts and retroperitoneal LAD.
- Given these findings and the tender cervical lymph node on exam:



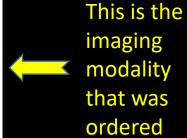
What additional imaging should we order?



Select the applicable ACR Appropriateness Criteria

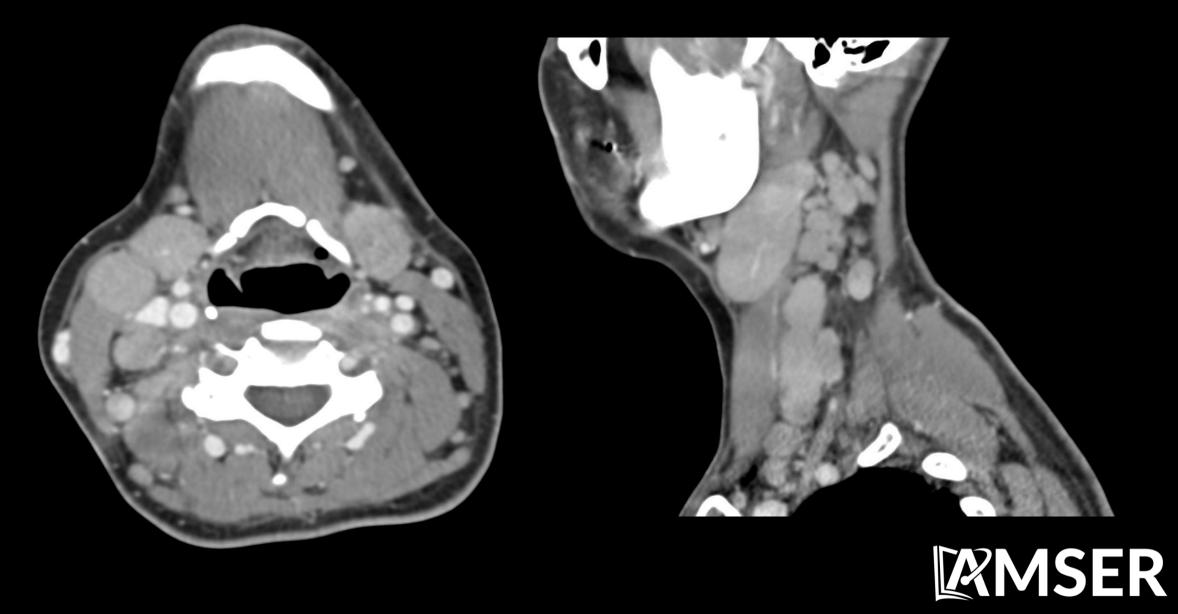
Variant 1:	Nonpulsatile neck mass(es). Not parotid region or thyroid. Initial imaging
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Procedure	Appropriateness Category	Relative Radiation Level
CT neck with IV contrast	Usually Appropriate	₩₩₩
MRI neck without and with IV contrast	Usually Appropriate	0
MRI neck without IV contrast	May Be Appropriate	0
US neck	May Be Appropriate	0
CT neck without IV contrast	May Be Appropriate	⊕⊕
CT neck without and with IV contrast	Usually Not Appropriate	⊕⊕
CTA neck with IV contrast	Usually Not Appropriate	⊕⊕
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	***
FDG-PET/MRI skull base to mid-thigh	Usually Not Appropriate	⊕⊕⊕
MRA neck without and with IV contrast	Usually Not Appropriate	0
Arteriography cervicocerebral	Usually Not Appropriate	***
MRA neck without IV contrast	Usually Not Appropriate	0

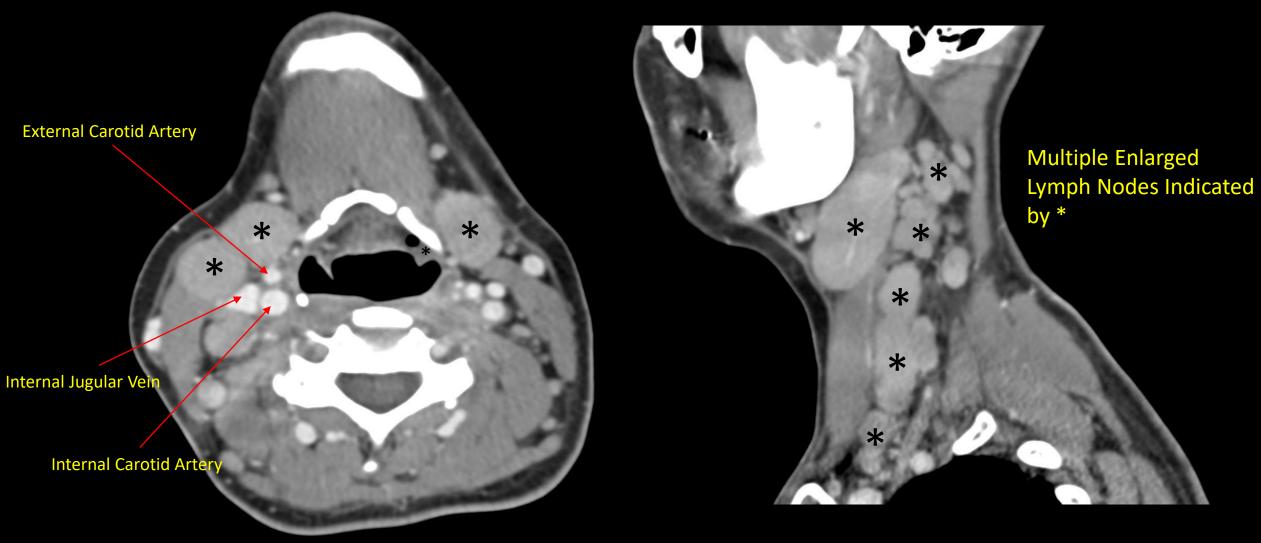




Findings (unlabeled)



Findings (Labeled)





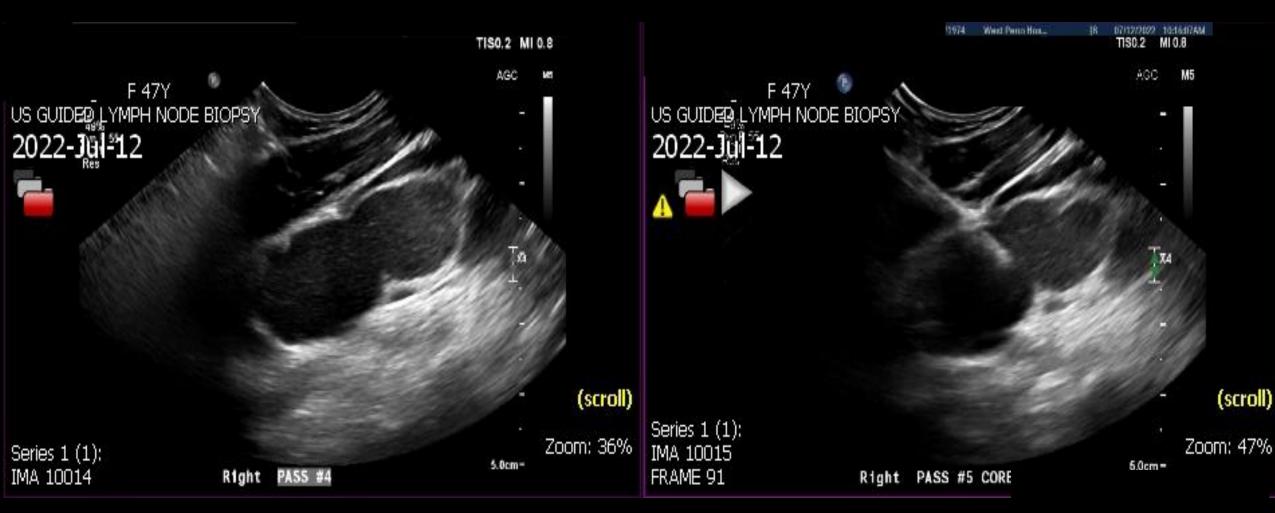
CT Head and Neck Findings

• Extensive bilateral cervical and supraclavicular adenopathy seen at the level of the internal jugular vein and the internal and external carotid arteries of the neck.

What procedure should be done next to help make a diagnosis?



Cervical Lymph Node US Guided Biopsy





Core Biopsy Results

- Pathology Report Summary
 - Atypical small sized lymphoid cells with ovoid nuclei, indistinct nucleoli and scant cytoplasm seen admixed with larger lymphoid cells which are clustered in prominent proliferation centers.
 - Via immunohistochemical staining, the atypical lymphocytes are CD20- positive B cells which coexpress CD5, CD23 and CD21.
 - The Ki67 proliferation index was estimated to be > 40%.



Diagnosis

- The core biopsy along with a bone marrow aspiration showed a CD5 positive lymphoma, most consistent with Chronic Lymphocytic Leukemia.
- The high Ki 67 proliferation index was noted to be concerning for aggressive disease.



Chronic Lymphocytic Leukemia

Overview:

- CLL is a hematologic malignancy involving the accumulation of phenotypically mature monoclonal B lymphocytes in the blood, bone marrow and lymph nodes.
- It is the most common adult leukemia in the Western world, and most cases are diagnosed in patients who are aged 60-74.
- There is a higher incidence of CLL in males, but studies have shown that women can have a more aggressive form of the disease.

• Clinical Presentation:

- History:
 - Most patients are asymptomatic when diagnosed but some may experience fatigue, weight loss, night sweats and fevers
 - Enlarged lymph nodes are the most common finding
- Physical Exam:
 - Enlarged, tender lymph nodes
 - Organomegaly
 - +/- fever



Chronic Lymphocytic Leukemia

• Evaluation:

- Labs:
 - CBC with immunophenotyping by flow cytometry
 - Peripheral blood smear
 - LN biopsy
 - Bone marrow aspiration
- Imaging:
 - CT of head/neck, chest, abdomen, pelvis for assessment of peripheral LAD

Management:

- Staging with FDG PET/CT is very helpful for treatment planning.
- The patient should be evaluated for treatment indications including threatened end-organ function, B symptoms, progressive anemia or thrombocytopenia, and/or complications from LAD or organomegaly.
- If indications for treatment are present, the following are possible treatment options:
 - Ibrutinib
 - Acalabrutinib with or with obinutuzumab
 - Venetoclax plus obinutuzumab



References

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- Johnson SA, Kumar A, Matasar MJ, Schöder H, Rademaker J. Imaging for Staging and Response Assessment in Lymphoma. *Radiology*. 2015;276(2):323-338. doi:10.1148/radiol.2015142088
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