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A Rare Presentation of Systemic Lupus Erythematosus with Lymphadenitis

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Abstract

A 26 year old African American female presented with the chief complaint of persistent fever and a single painful axillary lymph node for two weeks. This is a unique case of systemic lupus erythematosus (SLE) presenting with painful lymphadenitis. Moreover, the result of her lymph node biopsy was also unusual, showing histiocytic necrotizing lymphadenitis (HNL) presenting in SLE.

She presented with a fever of 103 degrees Fahrenheit and a single painful yet mobile axillary lymph node. She had no rash, joint tenderness, or hepatosplenomegaly on physical exam. Initial lab work showed absolute neutropenia and lymphopenia as well as normocytic anemia. There was initial concern for a possible infectious etiology due to the painful lymphadenopathy, thus broad spectrum antibiotics were initiated. Despite three days of antibiotics, the patient continued to have low grade fevers. HIV, CMV, and EBV testing was negative. Concern for malignancy led to an excisional lymph node biopsy. Additional lab testing that was pending prior to lymph node biopsy subsequently came back positive for ANA (antinuclear antibody) with titer >1:1280 in homogeneous pattern, anti-ds DNA antibody, and anti-Smith antibody, thus pointing towards SLE. She was subsequently started on high dose steroids with improvement of symptoms and her fevers resolved.

Histiocytic necrotizing lymphadenitis (HNL) is a histologic feature most commonly associated with Kikuchi-Fujimoto Disease; there are few case reports that have shown an association between HNL and SLE [2, 3]. Given the strongly positive ANA, anti-ds DNA and anti-Smith antibodies, Kikuchi-Fujimoto Disease was deemed less likely and more consistent with SLE. Due to the variability of presentations and the vast differential that Lupus can encompass, Lupus can often be a difficult diagnosis to achieve. However none of these criteria include lymphadenopathy. In this case, the patient only met 3 out of 11 criteria. Lymphadenopathy is seen in 12-59% of patient with SLE, however is often generalized and painless [4,5,6], thus making her presentation so rare. Due to the solitary lymph node that was painful, this initially pointed towards a possible infectious etiology. Also, Lupus can often be mistaken for malignancy; malignancy must be ruled out as patients with SLE are at an increased risk of developing non-Hodgkin lymphoma, Hodgkin lymphoma, and leukemia [1,7]. Therefore, even in patients who do not meet ACR criteria, SLE should still be investigated, if suspected, as patients may be presenting early in the disease course or in an atypical presentation.

ACR criteria for diagnosing Lupus

At least four of the eleven criteria of lupus from the American College of Rheumatology are usually present for lupus to be diagnosed.

- 1) Malar rash – a butterfly shaped rash across cheeks and nose
- 2) Skin rash – raised red patches
- 3) Photosensitivity – unusually strong reaction to sun light, causing a rash or flare
- 4) Mouth or nose ulcers – usually painless
- 5) Nonerosive arthritis – inflammation in two or more joints.
- 6) Cardio-pulmonary involvement – inflammation of the heart lining and/or lungs
- 7) Neurologic disorder – seizures and/or psychosis
- 8) Kidney disorder – increased protein or clumps of red cells in urine
- 9) Blood disorder – anemia caused by damaged red cells, low white cells or low platelet count
- 10) Immunological disorder – when your immune system attacks healthy cells
- 11) Antinuclear antibodies (ANA) – positive blood test not induced by drugs

Introduction

- A 26 year old African American female with no significant past medical history presented with the chief complaint of a single painful lymph node for the past week. She was also spiking high fevers at home. She was treated with a course of antibiotics from her primary care provider for the initial concern of folliculitis and was without improvement after the course of antibiotics.
- On presentation, vital signs were stable, expect she was spiking high fevers between 101-103 degrees Fahrenheit. She had no tachycardia. She denied any rashes, joint pain, night sweats, weight loss, or fatigue. She denies any trauma , but did admit to shaving the area. She denied any travel or sick contacts.
- Her mother was concerned for malignancy as the patient’s mother and aunt both had young onset breast cancer before the age of 30.
- She denies any history of any personal or family history of autoimmune disorders.

Differential and Workup

- The initial differential was extensive. Top differentials included infectious, malignant, or autoimmune etiologies.
- Due to the lymph node being painful, infectious etiologies needed to be worked up. The patient had high fevers as well as a low white count. Initial lab work showed absolute neutropenia and lymphopenia as well as normocytic anemia. There was concern for infection, however blood cultures, urine analysis and chest x-ray all came back negative. HIV, CMV, and EBV testing was negative. However, Infectious Disease was consulted and the patient was started on broad spectrum antibiotics . Despite three days of antibiotics, the patient continued to have fevers.
- Due to the patient’s family history of young onset breast cancer in her mother and aunt before the age of thirty, malignancy needed to be ruled out. On physical exam, the patient had no other areas of lymphadenopathy. Therefore a biopsy of the lymph node was taken, but took it took some time before the results were back.
- While the lymph nodes biopsy results were pending, autoimmune workup was underway. ANA (antinuclear antibody) came back positive with titer >1:1280 in homogeneous pattern. Also, anti-ds DNA antibody, and anti-Smith antibody came back positive as well. Thus, patient was diagnosed with Systemic Lupus Erythematosus, high dose steroids were started, and patient’s fevers resolved.

Discussion

- Due to the variability of presentations and the vast differential that Lupus can encompass, Lupus can often be a difficult diagnosis to achieve
- Lymph node biopsy came back as Histiocytic necrotizing lymphadenitis (HNL) which is a histologic feature most commonly associated with Kikuchi-Fujimoto Disease; there are few case reports that have shown an association between HNL and SLE [2, 3]
- Given the strongly positive ANA, anti-ds DNA and anti-Smith antibodies, Kikuchi-Fujimoto Disease was deemed less likely and more consistent with SLE
- Malignancy must be ruled out! Patients with SLE are at an increased risk of developing non-Hodgkin lymphoma, Hodgkin lymphoma, and leukemia [1,7]
- Four or more out of 11 criteria must be met of the American College of Rheumatology (ACR) criteria for classifying as possible systemic lupus diagnosis [1]. Lymphadenitis is not one of the criteria
- In this case, the patient only met 3 criteria. Lymphadenopathy is seen in 12-59% of patient with SLE, however is often generalized and painless [4,5,6], thus making her presentation so rare
- Even in patients who do not meet ACR criteria, SLE should still be investigated, if suspected, as patients may be presenting early in the disease course or in an atypical presentation

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