

Systemic Lupus Erythematosus with Initial Manifestation as Enlargement in Cervical, Axillary and Inguinal Lymphnodes: A Case Report

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Abstract

Systemic lupus erythematosus (SLE) is a chronic inflammatory disease that affects the nervous system, lungs, kidneys, joints, and other parts of the body. Changes in the distinctive characteristics and size of the lymph nodes help to identify lymphadenopathy. This is caused by a secondary reticuloendothelial proliferation in infection, malignancies, and inflammation. According to reports, 25–67% of SLE patients experience enlargement of the lymph node. Here, a thirty-three-year patient presented with lymph nodes enlargement in cervical, axillary, and inguinal regions, in addition to joint pain, renal involvement, and respiratory involvement is reported. The patient was diagnosed as Systemic lupus erythematosus according to ACR/ EULAR criteria, immunological and clinical domain SLICC (systemic lupus international collaborating clinics. In conclusion, lymphadenopathy was characterized as one of the manifestations of SLE. It might take years for the diagnosis of SLE to be made, as the instance given here demonstrates. It is possible that lymph node proliferative factors are also responsible for the development of autoantibodies.

Keywords: systemic lupus erythematosus, lymphadenopathy.

Introduction

A chronic inflammatory condition called systemic lupus erythematosus (SLE) that affects the nervous system, lungs, kidneys, joints, and other parts of the body. Reports state that 25–67% of SLE patients had enlarged lymph nodes ⁽¹⁻³⁾. However, SLE rarely causes an enlargement in the mediastinal and hilar lymph nodes ⁽⁴⁾.

Rarely SLE presents with lymph nodes in the neck. The American Rheumatology Criteria do not list lymphadenopathy as a criterion; however, some case documents in the literature describe generalized or localized lymphadenopathy as the initial sign of the illness ^(5,6). Modifications in the distinctive characteristics and size of the lymph nodes help to recognize lymphadenopathy. This is caused by

secondary reticuloendothelial proliferation in infection, malignancies, and inflammation. It is a benign sign that can appear at any stage of systemic lupus erythematosus illness⁽⁷⁻⁹⁾.

A patient presented with lymph nodes enlargement in cervical, axillary and inguinal regions, in addition to joint pain, renal involvement, and respiratory involvement is reported; she was diagnosed as an SLE according to ACR/EULAR criteria immunological and clinical domain SLICC (systemic lupus international collaborating clinics).

Case Report:

A 33 years old female patient from Kurdistan-Iraq presented in November 2020. The case started when she was pregnant on 24 weeks of gestation; she had joint pain in both hands, wrists, elbows, and knees with night pain, morning stiffness of more than 30 minutes with fever intermittent low grade. On 37 weeks of her pregnancy she developed recurrent episodes of lymphadenopathy, hepatomegaly, and fever; she delivered a live baby on her 38 weeks by normal vaginal delivery at the hospital.

In February 2021, 2 days post-partum, the patient developed anasarca, a generalized body oedema, productive cough with dyspnea mostly on exertion. Since that time, she developed arthritis in small joints of the hand, wrist, elbow, and knee with painful movement; she had hair loss, painless oral ulceration, with photosensitivity; at that time, she was admitted to the hematology ward with a fever, lymphadenopathy, ascites, and generalized edema.

After a referral from internist suspicion of lymphoma, biopsy of lymph node were done which reveal activation of lymph node due to rheumatic disease. The hematologist refers the patient to rheumatology and she was really admitted

in the rheumatology department, after assessing her vital signs checking: 130/85 mm Hg was her blood pressure, temperature was 38 °C, PR 100 bpm, SPO2 94% on room air, patient pale with bilateral leg oedema and ascites. There are enlarged lymphnodes in the inguinal, axillary, and cervical regions, their size ranging from one centimeter or more than one centimeters, investigations show hemoglobin was 9.1 g/dl, (11.5-16.5), WBC 9.7(4.5-10.0/L), PLT:48(130-400 per microliter). A rheumatoid factor was negative, CRP (C reactive protein):117.5 mg/l(5), ESR :118 mm/hr, albumin:2.14 low, LDH 538 high, chest-ray show mild left lower lung plural effusion, ECHO : ejection fraction 70% , trace pericardial effusion size (0.5 cm).

Infection from other sites of the body were excluded by sending to blood investigations, general urine examination, and blood culture.

The ultrasound of abdomen shows hepatomegaly 20 cm with moderate to severe amount of ascites, her ANA 14.6 + (N: less than 0.8), Anti- double strand DNA positive 44.5 (less than 25), lupus anticoagulant 47 positive, anticardiolipin negative, C3; 52 low (90-180 mg/l), C4: 8 (10-40 mg/l). Two days later, she develop oliguria with urea : 161 mg/dl high (15-45), creatinine: 2.5 protein in urine +++ ,granular cast 2-4 in urine ,and 24 hour urine collection was 375, albumin creatinine ratio elevated, according to 2019 CR/EULAR criteria, diagnosed as an SLE with nephrotic lupus nephritis. Renal biopsy was not done for the patient because when patient develop proteinuria and lupus nephritis, she was tired and her condition did not tolerate biopsy.

The patient received hydroxychloroquine tab 200 mg by 2, IV methylprednisolone 1g for 3 days, with IV cyclophosphamide 1g. Diagnostic and therapeutic

aspiration of ascites was done, the result of ascites fluid examination was exudative Sag ratio less than 1.1.

After a week of follow-up, her ESR became 24, CRP: 16 decreased, creatinine normalize, protein in urine become 1 plus. After 30 days of remaining in hospital, her ascites fluid starts to decrease and just small amount remains. Her edema subsides and weight at the first day of admission was 78 kg; now it became 60 kg after subsiding body edema, good urine output, stable vital signs.

Two days later, she developed ARDS (acute respiratory distress syndrome) admitted to RCU (Respiratory Care Unit). On examination, she was tired; hypoxic (SPO₂ 80 without Oxygen), chest was full of crepitation, her CRP reached to 117, PCR by nasopharyngeal swab for COVID-19 was negative, chest x ray showed feature of chest infection and pulmonary hemorrhage and intubated for 10 days, she received treatment (covered with steroid IV, three antibiotic's IV with daily follow-up of vital signs later her chest started to improve (and follow up x ray became better and extubated).

After 48 days in the hospital, the patient improved and discharged home on treatment prednisone tab started by 40 mg with tapering, Vit D tablet, hydroxychloroquine tab 200 mg by 2, with iv cyclophosphamide 1g for 6 cycle every month.

Discussion:

Fever with cervical lymphadenopathy are the first presentation of various diseases. For example, common causes include tuberculous lymphadenitis, lymphoma, and even rheumatic diseases, for instance SLE and sarcoidosis. A comprehensive physical examination and collection of a full medical history should allow for a more accurate assessment of each patient.

Evidence of infection or immunological disorder can be found by laboratory testing. The gold standard investigation is an excisional biopsy to distinguish malignant from benign association with inflammatory disease ⁽¹⁰⁾.

Multiple organs are affected by SLE, an autoimmune condition. Women are affected more than men. Four or more of the eleven American College of Rheumatology (ACR) criteria must exist to diagnose SLE ⁽¹¹⁾. The ACR criteria are not involved and do not include Lymphadenopathy. According to an investigation by Shapira et al. ⁽¹²⁾, out of 90 patients with SLE only 23 had lymphadenopathy; it was also observed that those patients had more systemic symptoms.

The estimated prevalence for lupus lymphadenopathy (LL) ranges from 5-7% at the illness beginning to 12-15% at the disease stage ⁽¹³⁾. LL mainly affects the axillary and cervical areas. The lymph nodes are mobile, soft, tender, and not attached to deep planes ⁽²⁾. Lymph node biopsy is advised to rule out lymphoproliferative or infectious diseases if there is severe lymphadenopathy ⁽¹⁻³⁾. There are two different categories of LL: focal: involving two chains of lymph nodes, and general, three or more ⁽¹⁴⁾.

Generalized lymphadenopathy was a frequent early symptom of SLE in children, according to research by Kitsanou et al ⁽¹⁵⁾. Regional or systemic lymphadenopathy is included as a symptom in much older English literature on SLE ^(16,17).

In conclusion, lymphadenopathy was characterized as one of the manifestations of SLE. It might take years for the diagnosis of SLE to be made, as the instance given here demonstrates; It is possible that lymph node proliferative factors are also responsible for the development of autoantibodies.

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Declaration of patient consent:

The patient has given her consent for information to be reported in the journal.

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Conflicts of interest

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