Review Article

SYSTEMIC LUPUS ERYTHEMATOUS

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ABSTRACT

Systemic lupus erythematosus (SLE) is a relapsing and unpredictable multisystem disease. The exact cause is unknown, although it appears to result from an immunoregulatory disturbance brought about by combination of genetic, hormonal, chemical, and environment factors. The major symptoms of SLE are malar rash, discoid rash, photosensitivity, oral ulcers, arthritis, serositis, renal manifestations, neurological manifestations, haematological manifestations, immunological manifestations, or a positive antinuclear antibody (ANA) result. Diagnostic criteria based on major criteria of SLE. Prognosis is good if the disease is controlled during the initial acute phase. Treatment depends on the severity of the disease as well as organ involved. In case of mild symptoms require the use of NSAIDS and in severe form require corticosteroids or immunosupressants. Therefore, SLE patients require a healthcare team of multiple disciplines and professions, with close monitoring and control to prevent the worsening of the disease or the development of complications.

Case presentation: case of 25-year-old female patient present to the hospital with progressive difficulty in breathing, chest pain, generalized edema, joint pain, alopecia, severe fatigue and sleep disturbance over the 6 month which is diagnosed as systemic lupus erythematosus

Conclusions: SLE is a chronic, autoimmune disease which induce renal, digestive, neurologic and hematologic disorders. It is characterized by erythema, myalgia and arthralgias. For prevention of complication, multidisciplinary health professional team is required.

INTRODUCTION

Systemic lupus erythematosus is a chronic autoimmune disease characterized by unpredictable flares, irreversible organ damage, adverse impacts on health-related quality of life, and increased mortality.[1] SLE is a chronic, unpredictable, relapsing and serious autoimmune disease, characterized by erythema discus, myalgia and arthralgias. It is a multisystem disease and can induce renal, digestive, neurologic and hematologic disorders. SLE is a prototypic multisystem disease of autoimmune origin and is characterized by a fundamental failure of mechanisms that maintain self-tolerance.[2,3]More than 90% of cases of SLE occur in women, frequently starting at childbearing age (15-44 years).[4,5]

The exact cause of SLE is not known, but several factors have been associated with the disease including genetic, environmental, and hormonal factors. The pathogenesis of SLE consists of antibodies (anti-nuclear antibody, anti-dsDNA, anti-histone antibody) and immune complexes, resulting in damage to many tissues and organs.[6] The disease results from the interaction of genes, environment, and random effects combining to lead to a loss of tolerance to self-antigens and active autoimmunity.[3]

SLE is characterized by the breaking of tolerance to nuclear self-antigens and the production of pathogenic autoantibodies. It involve several organs in the body such as skin, eyes, kidney, heart, muscles, and the joints.[7]SLEmay also have hematological as well as joint, kidney, and central nervous system involvements. Hematological abnormalities such as anemia, leukopenia, thrombocytopenia, and autoimmune hemolytic anemia are the best-known and most common hematological findings detected in SLE patients which are included in the classification criteria for SLE.[8]

CASE REPORT

The case is a 25-year-old female patient present to the

hospital with progressive difficulty in breathing, chest pain, generalized edema joint pain, alopecia, severe fatigue and sleep disturbance over the 6 month. The other problem noted with loss of appetite since illness. She also noted slightly heavier than usual menstrual periods and history of abortion. The patient was not on any medications, had no prior surgeries, no previous hospitalization and had no known allergies. The patient denied using alcohol, tobacco, or any illicit substances. She denied any history of anemia, easy bruising or bleeding, yellowing of skin, rashes, and prior history of blood transfusions.

On examination, her blood pressure was 100/70 mmHg, heart rate was 96 beats per minute, respiratory rate was 20 breaths per minute, temperature was 36.9°C (98.4°F), weight was 56 kg, height was 153 cm, and body mass index was 23.9 kg/m^2 and abdominal girth was 40 cm. She was alert, oriented, and looks sad, tired, limp walk.

Significant findings were included crackle sound over right side of lung, malar rash (butterfly rash), discoid rash over the whole body, diffuse anasarca, alopecia, oral ulcer, vasculitis.

There was evidence of cardiomegaly and right side pleural effusion on chest x-ray examination. Results of hematologic and biochemical evaluation were as following; white cells: 8200 in mm3, hematocrit 22,8%, hemoglobin 7,7g/dl, platelets 132.000 in mm3 , glucose: 91 mg/dl, urea: 58 mg/dl, creatinine: 1,7 mg/dl, lactated hydrogenase (LDH): 2600 u/lt, total protein: 6,3 mg/dl, proteinuria 300 mg/lt. Antibody (ANA), Sm (RNP-70, RNP-A and RNP-C) were positive. Anti-ds DNA and AMA-M2 were negative. Ribosomal protein was also positive.

It was suspected SLE because any four items from the diagnostic criteria of the American College of Rheumatology (ACR) were satisfied. There was presence of Positivity of ANA, Sm (RNP-70, RNP-A and RNP-C) and Ribosomal protein, photosensitivity, oral ulcers, cardiac enlargement, right side pleural effusion and low level of hemoglobin. These evidences have pointed out SLE, therefore the diagnosis was drawn on the basis of findings. The patient was admitted to the Medical ward where she received Inj methyl prednisolone 725 mg I/V start and TDS, Tab wysolone 50 mg P/O OD, Tab HCQS 200mg P/O BD, Inj Pireracillin 4.5 gm IV TDS, Inj Lasix 10 mg IV TDS, InjXone 1 gm IV BD, Inj Ondem 4mg IV BD, Cap Gemdol 1 cap P/O TDS, Tab Nicardia retard 20mg P/O OD and syrup Norvent 10ml P/O TDS, Nebulization w (A:I:NS). The patient was hospitalized for 10 days. The patient's clinical symptoms improved and she was discharged with on therapeutic regimen of wysolone 50mg P/O OD and instructed to follow-up as an outpatient for further workup.

DISCUSSION

SLE is a multisystem vasculitis disease and presence of autoantibodies. Auto antibodies are responsible for the multisystem involvement. Skin lesions and dermal sensitivity are the other clinical features of SLE as it was evident in this case. About 90% of patients with SLE are women, with the risk in disease increasing dramatically with the appearance of female sex hormones.[3] (Mbonu, et al., 2019).In our case, female patient and involvement of different organs such lung, kidney, cardiac and skin.

The reason of hemolytic anemia is the presence of auto antibodies against blood cells. Anemia causes fatigue, and exercise intolerance. Thrombocytopenia causes purpuras and mucosal hemorrhages.[6] (Kim, et al., 2015). In our case, there was anemia due to chronic hemolysis and leucocytosis due to infection.

Molar Rash is one of the criteria for SLE as per American college of rheumatology (ACR), present in patient with erythematous flat or raised rash across the bridge of the nose and cheeks, which usually spares nasolabial folds. In our case molar rash was demonstrated by butterfly rash over nasolabial folds.

Photosensitivity is another sign of SLE which is produce skin rash after exposure to sunlight. In our case photosensitivity was present.

Serositis, one of the criteria of systemic lupus erythematosus may be present in patients with pleuritis, pericarditis, pleural and pericardial effusion. Lung injury may cause long term chronic lung damage and respiratory complications in advanced systemic lupus and the other connective tissue. Main reason of serositis is immune mediated inflammation on serous membranes caused by auto antibodies. Pleuritis make a barrier for inspiration and causes dyspnea. In our case serositis was demonstrated by radiological investigation which revealed that right side pleural effusion, pericarditis, cardiac enlargement.

As per criteria of American College of Rheumatology (ACR), Arthritis is one the criteria of SLE. Majority of patient present with arthritis, joint pain, unable to move. In our case, joint pain specially in knee joint pain was present.

Lupus nephritis is an important prognostic indicator and it can lead to nephrotic syndrome and renal failure in developed patients by autoantibodies. Lupus nephritis develops in 50% of systemic lupus patients. One of the laboratory findings of lupus nephritis is proteinuria. In our patient, there was 300 mg/lt proteinuria and diffuse proliferative glomerulonephritis which was shown in biopsy material. Immune suppressive therapy inhibits progression of nephritis

Systemic lupus must be kept in mind in female patients who presents to hospital with arthralgia, fever and skin lesions. In our case, she was present with knee joint pain, fever.

The diagnosis can be confirmed by histopathology and serology. Serology showed high titre of ANA though Anti-ds and DNA. Anti-ds DNA antibodies are highly specific for SLE, which are also present in our case. The disease severity varies from mild to severe, and requires long-term and often aggressive treatment. Strict avoidance to sun is to be advised the patients with use of broad-spectrum sunscreens. Corticosteroids (1-3mg/kg/d) and hydroxychloroquine (4-6mg/kg/d) have shown excellent results in control of disease. Other options include azathioprine (0.5-2.5 mg/kg/d), cyclophosphamide (0.5-2.5 mg/kg/d), intravenous immunoglobulins 2 g per kg per dose and plasmapheresis. In our case methyle prednisolone 725 mg I/V start and TDS for two days and Tab wysolone 50 mg P/O OD.

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