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Unveiling the Heterogeneity of Systemic Lupus Erythematosus: A Series of Interesting Cases

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ABSTRACT:

Systemic lupus erythematosus (SLE) presents a diagnostic and therapeutic challenge due to its diverse clinical manifestations and relapsing-remitting course. This review delves into the complexities of SLE, highlighting its predilection for young females and potential to affect any organ system. We explore the spectrum of clinical presentations, ranging from common symptoms like rash, fatigue, and arthritis to severe complications involving the kidneys, nervous system, and blood cell counts. The cornerstone of management is focusing on controlling acute flares, preventing future relapses, and alleviating daily symptoms. We discuss the crucial role of serological tests and tissue biopsies in establishing a definitive diagnosis. This case series presents [Number] cases of Systemic Lupus Erythematosus (SLE) diagnosed and managed at our institution. The series aims to illustrate the diverse clinical presentations of SLE.

Keywords: SLE, Autoimmune disease, Systemic sclerosis, Neuropsychiatric syphilis, Lupus Nephritis

1. Introduction:

The hallmarks of systemic lupus erythematosus (SLE) is that it's a multisystemic autoimmune disease, which includes immunological complex deposition, persistent inflammation in the traditional target organs—the skin, joints, and kidneys—and autoantibodies directed against

nuclear antigens. The burden of SLE remains high even with significant advancements in diagnosis and treatment. To enable early patient referral and diagnosis, it is critical to understand typical presentations and the diagnostic procedure. The majority of patients appear with constitutional, mucocutaneous, and musculoskeletal symptoms first; these symptoms may include myalgia, exhaustion, mouth ulcers, alopecia, joint pain, and rash unique to lupus. Individualized lifestyle interventions and patient education are used in non-pharmacotherapy management to enhance quality of life and medication adherence (e.g., hydroxychloroquine or immunosuppressant). A few significant advancements in lupus nephritis and SLE therapies have been made in the past ten years, including belimumab, anifrolumab, and voclosporin. Nonetheless, the disease's progression is still unpredictable, and the death rate is too high. We acknowledge that achieving remission or low disease activity is the primary objective of therapy. Vigilant preventive and management measures are required for comorbidities resulting from both disease activity and medication side effects, including infections, osteoporosis, and cardiovascular disease. Priority areas of managing SLE include balancing treatment-related comorbidities and customizing therapy strategies to achieve remission.

Case 1:

A 28year old female presented with fatigue, nonhealing oral ulcers, significant weight loss and excessive hair loss since 9 months. In past two years the patient had 2 abortions of unknown nature. On examination, BP 160/100 mm/Hg, PR 84/min, Temperature 38degree C. Systemic examination of Cardiovascular and Abdominal Systems were normal. Respiratory System-Coarse crepitations heard at the base bilaterally. HRCT showed infiltrations at the base bilaterally.

Table 1: Key Investigations pointing towards diagnosis of case 1

WBC	**8400 in mm3**	**Urine protein**	**300 mg/lt**
Hematocrit	23%,	Anti Nuclear Antibody (ANA)	Positive
Hemoglobin	7.6g/dl,	Anti ds DNA	Positive
Platelets	130000 in mm3	Anti cardiolipin antibody	Positive.
RBS	90mg/dl	All other biochemical tests	Normal
Urea	60 mg/dl	Anti HBs Ab, Anti HIV, Anti HCV titrations	Negative.
Creatinine	1.6mg/dl	Blood and Urine culture	Negative
LDH	2600 u/lt, Direct Coombs-Positive	USG Abdomen	Bilateral Renal paranchymal disease.
Total protein	63.4 mg/d	Renal biopsy	Diffuse proliferative glomerulonephritis

With the above findings the patient was diagnosed to have Systemic Lupus Erythematosus. The patient was treated with pulse steroids and the patient showed improvement symptomatically.

Case 2:

A female, 25 years of age, came with complaints of breathing difficulty, fever with chills and rigor, abdominal pain fatigue, weight loss(10kg), generalized body pain, alopecia and recurrent nonhealing oral ulcers since 2 months. In addition, she mentioned a history of blood transfusions due to anemia. There was no significant familial history of rheumatic diseases. Patient is on Levothyroxine75 mg/day for hypothyroidism since2 years. On examination, her

BP was 80/60 mmHg, temperature-38.9 °C, her pulse Rate 138 pm, and her SpO2 was 93%. Patient has pallor, bilateral pitting pedal edema; her JVP was raised. Cardiovascular examination revealed muffled heart sounds, Respiratory system-bilateral air entry decreased. Chest X-ray- increased cardiothoracic ratio and bilateral pleural effusion, ECG- sinus tachycardia. CT Chest- mild bilateral pleural effusion, large pericardial effusion (20–22 mm), hepatomegaly > 16 cm, moderate ascites. 2D ECHO -cardiac tamponade. Emergency pericardiocentesis was done. The pericardial fluid, peritoneal fluid analysis and other additional investigations excluded infectious and malignant etiology. Microscopic urinalysis showed proteinuria and microscopic hematuria. Based on a comprehensive clinical examination, medical history, and laboratory results, the presence of SLE was suspected, leading to the performance of further tests.

Table 2: Key Investigations pointing towards diagnosis of case 2

WBC	2500mm3	ANA	Positive
Hemoglobin	8 mg/dl	Anti-dsDNA	Positive
Plateleta	230k mm3	Protein 24 h urine	492 mg/24 h
ESR	70 mm/1 h	TSH	11.54
CRP	79 mg/L	C4	0.04 g/L
Creatinine	3.9 mg/dl	C3	0.28 g/L
LDH	728 U/L	ENA screen	Negative
Coombs direct	Positive	APL antibodies	Negative

The laboratory tests confirmed the diagnosis of SLE and the patient was treated with corticosteroids. The patient improved well symptomatically.

Case 3:

16-year-old female presented with an episode of seizure. She had fever, headache, behavioral changes, cognitive impairment and delusions since 3 days. On examination patient was in altered sensorium and vitals Bp:120/70mmHg; Pulse:90bpm; RR:14cpm; SpO2:99% @room air; Temp :98.4 F. Head to toe examination revealed she had bilateral pitting pedal oedema and rashes over cheeks, abdomen and legs. CNS examination: Bilateral pupils equally reacting to light. Plantar bilateral flexors. No signs of meningeal irritation. Other systemic examinations were normal.

Table 3: Key Investigations pointing towards diagnosis of case 3

WBC	**6800mm3**	**CT and MRI brain**	**No Abnormalities**
Hemoglobin	5.7 mg/dl	ANA	Strong Positive
Platelets	1.20k mm3	Anti-dsDNA	Positive
ESR	80 mm/1 h	Protein 24 h urine	312 mg/24 h
CRP	120 mg/L	C4	0.21 g/L
Creatinine	2.8 mg/dl	C3	1.09 g/L
LDH	620 U/L	APL antibodies	Negative
Coombs direct	Positive		

With the above evidence the patient was diagnosed to have Neuropsychiatric Presentation of Systemic Lupus Erythematosus. She was treated with pulse steroids, HCQ and Cyclophosphamide injections. The patient showed tremendous improvement.

2. Discussion and Conclusion:

All of our cases were females and is of reproduction age group. Each one of them has distinct presentation to hospital due to the heterogenic nature of the disease. Clinical suspicion was made based on the SLICCA and EULAR / ACR criteria for Systemic Lupus Erythematosus. The diagnosis of Systemic Lupus Erythematosus (SLE) relies on the examination of both clinical symptoms and laboratory test results. Nevertheless, in the present day, there frequently exists a substantial time gap between the appearance of symptoms and the determination of a diagnosis, despite the growing knowledge and understanding of SLE. Likewise, we encountered challenges in diagnosing the condition during the initial presentation. Prior to the diagnosis of SLE, the above cases have made several visits to the primary care center due to their symptoms. Individuals diagnosed with systemic lupus erythematosus (SLE) may exhibit a range of other clinical symptoms and conditions that resemble lupus. Lupus mimickers are a collection of illnesses that display similar clinical and laboratory characteristics to systemic lupus erythematosus (SLE). These conditions might include infections, neoplasms (abnormal growths) and drugs. There are other syndromes that have hallmarks of systemic lupus erythematosus (SLE) and also include features from other diseases such as rheumatoid arthritis (commonly known as "Rhupus"), polymyositis/dermatomyositis, systemic sclerosis, and Sjögren's syndrome. Glucocorticoids and antimalarial drug are the primary treatments for managing lupus, in addition to immunosuppressive or biologic medications. According to current recommendations, it is advised to consider using antimalarial medications, including hydroxychloroquine, for all patients with systemic lupus erythematosus (SLE).

Figures:

Figure 1: Alopecia-Localized Nonscarring alopecia in Systemic lupus erythematosus which can be considered as an indicatin of active disease

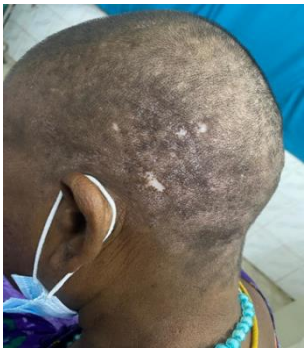

Figure 2/ Malar Rash, also known as Butterfly Rash, an erythematous flat or raised rash across the bridge of the nose and cheeks, which usually spares nasolabial folds

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