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Diagnosis and Management of Systemic Lupus Erythematosus: A Case Report

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Abstract

Systemic lupus erythematosus (SLE) is a multisystem inflammatory disease with a broad clinical presentation, which is principally difficult to diagnose across the emergency departments (EDs). The immune system of the body in this disease mistakenly damages or attacks healthy tissues. Majority of the patients suffering from SLE tend to develop "secondary heart disease" once in a while throughout the course of their primary illness. This study aimed to report a case of a previously healthy 9-year-old Saudi female who presented with rheumatic fever and congestive heart failure accompanied by productive cough, chest, abdomen, and back pain. A 9-year-old Saudi female was presented to the emergency department with a history of progressive rheumatic fever, iron deficiency anemia, and pain in chest with productive cough. Examination revealed that patient felt extremely ill, pale, afebrile, with a loss of appetite, tachycardic, high grade fever (39 °C), tachypneic, and a peripheral oxygen saturation of 95% on 40% supplemented oxygen" with low blood pressure 105/70 was noted. The patient was assumed to be diagnosed with probable SLE. We started our patient on methylprednisolone, omeprazole, and prednisolon and noticed sustained improvements. Multisystemic and acute life-threatening conditions should arise the suspicion of autoimmune diseases, predominantly SLE in the ED. SLE treatment shall be planned separately with consideration to utilize the "best-suited therapy" for targeting the organ systems affected. Lack of an explicit biological marker, disease heterogeneity, as well as absence of a specific outcome measurement for improvement makes this procedure harder.

Keywords: Rheumatic fever; Juvenile rheumatoid arthritis; Lupus erythematosus; Purpura; Discoid; Thrombotic thrombocytopenic

Introduction

Systemic lupus erythematosus (SLE) is considered to be a "multisystem inflammatory disease" which is often hard to diagnose [1]. Specifically for the emergency department (ED) staff, it is critical to consider SLE as an option, when they observe an individual suffering from the signs and symptoms suggesting a disorder involving multiple systems. The complications observed in the EDs for SLE can be managed in a usual manner, the most widely found being pulmonary emboli, respiratory distress, hemoptysis, acute myocardial infarctions, and strokes [2, 3]. Various other complications including cerebritis, renal failure, pulmonary hemorrhage, and pericardial tamponade can be managed by proper consultations obtained from the subspecialist [1].

Congestive heart failure in patients with SLE is often multifactorial in origin [4]. Individuals having lupus possess a significantly high risk for stroke, atherosclerosis or premature "coronary heart disease" (CHD) and many other "cardiovascular-related conditions" as compared to those deprived of lupus [5]. During the onset SLE in childhood, there are numerous clinical symptoms more specifically found in comparison to adults, such as proteinuria, renal involvement, mucocutaneous involvement/ ulcers, malar rash, seizures, urinary cellular casts, fever, hemolytic anemia, thrombocytopenia, and lymphadenopathy. Among adults, sicca as well as Raynaud pleuritic are thought to be twice as common as in adolescents and children [6]. One such classic presentation of a rash, joint pain, and a triad of fever in females of childbearing age needs a rapid investigation into SLE diagnosis. People with SLE often experience a broad spectrum of symptoms as well as have multiple combinations of the organs involvement, while no definite test can establish the systemic lupus diagnosis [7].

The management along with the diagnosis of the congestive heart failure in SLE is not different significantly from the results obtained through other etiologies. For the purpose of reversing this procedure, prompt recognition by laboratory evaluation, signs and symptoms, with echocardiography, chest X-rays, and electrocardiogram is essential. To assist health care professionals to improve the SLE diagnosis accuracy, 11 such criteria were identified by the American Rheumatism Association [8]. These include discoid skin rash (redness in patches with hypopigmentation and hyperpigmentation tends to cause scarring), arthritis (more than two tender and swollen joints of the extremities), malar (observed on face mainly over the cheeks) "butterfly" rash, irritation of brain (manifested by psychosis or seizures referred to as "lupus cerebritis"), pericarditis or pleuritis, mucous membrane ulcers, antinuclear antibody, blood-count abnormalities, photosensitivity (rash observed in skin in reaction to exposure to ultraviolet light or sunlight),

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Table 1. Diagnostic Criteria of SLE [9]

Criterion	Definition/examples
Malar rash	Fixed erythema over the malar eminences, tending to spare the nasolabial folds
Discoid rash	Erythematosus raised patches, may scar
Photosensitivity	Skin rash as a result of unusual reaction to sunlight
Oral ulcers	Usually painless
Arthritis	Non-erosive: Jaccoud's arthropathy
Serositis	1) Pleuritis - pleuritic pain, pleural rub, pleural effusion 2) Pericarditis - ECG changes, rub, pericardial effusion
Renal disorder	1) Proteinuria (> 3+ or 0.5 g/day) 2) Cellular casts in urine
Neurological disorder	1) Seizures 2) Psychosis
Hematological disorder	1) Hemolytic anemia 2) Leukopenia 3) Lymphopenia 4) Thrombocytopenia
Immunological disorder	1) Anti-DNA antibodies 2) Anti-Sm antibodies 3) Anti-phospholipid antibodies
Anti-nuclear antibody	Exclude drug causes

A person is said to have SLE if he/she meets any four of these 11 criteria simultaneously or in succession.

and abnormalities in kidney (abnormal volume of urine protein or cellular elements clumps called casts measureable with a standard urinalysis) (Table 1) [8, 9]. This study aimed to report a case of a previously healthy 9-year-old Saudi female who presented with rheumatic fever and congestive heart failure accompanied by productive cough, chest, abdomen, and back pain.

Case Report

A 9-year-old Saudi female was admitted due to rheumatic fever (experienced at the age of 6 years) accompanied by iron deficiency anemia, juvenile rheumatoid arthritis (6 years and 7 months), and celiac disease (reported at 8 years of age). Two weeks prior to admission, she presented with fever along with productive cough, paleness, chest, abdomen, and back pain, loss of appetite, depressed mood, poor school performance, malar rash, and mouth ulcers.

On inspection, patient was feeling extremely ill, pale, afebrile, with a high grade fever documented to be 39 °C, tachypneic, tachycardic, and a "peripheral oxygen saturation of 95% on 40% supplemented oxygen" with low blood pressure (BP) 105/70 was noted. The patient had a history of juvenile rheumatoid arthritis, also showed typical congestive heart failure as of accepted case from the peripheral hospital at the age of 6 years, iron deficiency anemia, celiac disease and 2 months history of "inflammatory polyarthralgias" (joints pain) encompassing initially "interphalangeal joints", evolving, sometime later, generalized, progressive intermittent abdominal pain and fever (1 month), colicky in nature with no aggravating or relieving factors. At the age of 8 years, patient perceived failure to thrive and had frequent abdominal pain. Therefore, celiac profile and endoscopy was performed confirming celiac disease. One month prior to admission, she had symptoms of an upper respiratory tract infection followed by right and left ankle joint pain experienced for 2 weeks, both knees and wrist joints experienced for 1 month as migratory joints pain. For 1 month before admission, she was immobile (unable to walk) with severe pain. Back pain for 4 days increased with movement.

Her vital signs on presentation were noted for tachycardia 105 beats per minute, temperature 39 °C, BP 105/70 mm Hg, no distress, and scaring from the skin rashes reported previously. Neurological and respiratory examinations were otherwise average with RR 26/min, and O_2 saturation 92%.

Initial laboratory tests demonstrated anemia: hemoglobin of 7.5 g/dL (12.0 - 16.0 g/dL), platelets 798 × 10^9/mL, white blood cells (WBC) count of 12.9 × 10^3/µL, total bilirubin of 0.7 mg/dL, undetectable direct bilirubin, glucose of 120 mg/dL, and pH 7.40. Serology for human immunodeficiency virus (HIV), hepatitis, and direct coombs tests were also found to be negative. Based on these findings, renal biopsy was performed and diagnosis of SLE was made and therapy with lasix, captopril, antibiotics, nebulization hydroxychloroquine (plaquenil) 200 mg TW, vitamin D3 800, and international units PO once daily was initiated. We also continued methylprednisolone 30 mg/kg/dose, 1 g/day, given over 60 min, for 3 days, omeprazole 1 mg/kg/day, and also started prednisolon 10 mg PO OD for 1 month, with tapering doses prior to discharge.

Discussion

The patient's prognosis with the SLE or SLE is known to be considerably improved with 20 years of survival now roughly about 80% partly owing to the treatments which are effective [10]. SLE is thought to be a "multisystem autoimmune disease" that tends to impact different organs as well as tissue in an individual's body ultimately causing dysfunction and damage. Some of the patients with lupus often possess mild disease that can primarily be treated with simple medications, while others tend to have life-threatening and serious complications. Treatments should be extremely individualized as well as varies as per the inconsistency during the clinical presentation of a disease [11]. The utmost care for the patients with SLE comprises of support and education services in addition to the non-pharmacologic and pharmacologic treatments. For SLE, the drug therapies are formed to suppress the inflammation and immune responses.

Establishment of the patient management and diagnosis with SLE needs an incorporation of the results of physical examination, patient symptoms, and diagnostic test results. The SLE management is mainly based on the disease manifestation and severity [12], even though "hydroxychloroquine" plays a dominant role in all SLE patients considering its treatment in long term. The "lupus in minorities: nature versus nurture" or (LUMINA) study along with other trials offers evidences of a decline in prolonged life and flares in individuals given hydroxychloroquine, thus, creating it as a foundation for managing SLE. Specifically in the patient, the diagnostic criteria of SLE were accomplished when oral ulcers, malar rash, serositis renal disorder: lupus nephrites II hematologic disorder, microcytic hypochromic anemia, immunologic disorder, anti-DNA antibody positive, anti-Smith abs positive, antinuclear antibody positive were summed up.

The EULAR also is referred to as "European League against Rheumatism" released recommendations for SLE treatments in 2007 [13]. In individuals with SLE without the manifestation of the major organs, antimalarial and glucocorticoid agents are considerably useful. However, for short term, NSAIDs can be consumed in patients possessing a low risk for complications from such drugs. Considering different immunosuppressive agents such as methotrexate, mycophenolate mofetil, and azathioprine in refractory circumstances or at times when doses of steroid cannot be limited to levels for a longer-term practice [14]. The recommendations from EULAR for SLE management with neuropsychiatric manifestations support the treatment and evaluation of these symptoms in a similar manner, as they are treated and evaluated in patients without SLE; if such symptoms persist, management of these symptoms as an extension of SLE should be considered [13].

Conclusion

The current case report implicates that SLE should be considered during refractory heart failure to substantial conventional therapy, exclusively in young females. Furthermore, early corticosteroids treatment, either with or without immunosuppressive agents, might lead to improved and good outcomes. SLE treatments need to be planned on individual basis with primary consideration for utilizing best suited therapy so as to target the organ systems affected. Lack of certain biological markers, heterogeneity of the disease as well as absence of single outcome measurement for improvement often makes it a critical process. Lastly, this case specifically reminds a healthcare professional that even though lupus is considered to be a complicated disease on its own, some number of the patients do present with a more concomitant or second autoimmune disease that increases the complication. Due to this reason, the lupus diagnosis is just the starting point of a case. Furthermore, there is a need to raise awareness among SLE patients regarding this condition in order to guarantee concordance with treatment, as limited compliance subsidizes towards failure of treatment with flares in disease, accumulation of impairment such as amplified risk of death or renal failure.

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Competing Interest

The author declared that they have no competing interests.

Consent

Written informed consent was acquired from the patient for publication of this case report. The patient refused image publication.

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