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Neuropsychiatric Systemic Lupus Erythematosus (NPSLE) – Masquerade Diagnosis: A Case Report

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Abstract

Diagnostic challenge is seen in Systemic Lupus Erythematosus (SLE) due to insidious onset, unpredictable course and broad spectrum of clinical presentation [1]. SLE can mimic other diseases manifestation and possibility not fulfilling the classification criteria at the earlier course of the disease [1]. We highlighted a case of a man, lived with diagnosis of Schizophrenia for almost 15 years subsequently revealed him suffered from neuropsychiatric lupus as he fulfilled the SLE classification criteria. His initial presentation was altered sensorium treated as acute psychosis- the tip of an iceberg to the underlying disease [2]. His multiple admissions to psychiatric ward were believed contributed by episode of lupus flare [2,3]. Necessary investigation is important to exclude another medical condition before make a diagnosis of Schizophrenia using DSM-V criteria [2]. The misdiagnosis has significantly impaired his social life and untreated disease had leads to morbidity and severe organ damage. This case emphasizes on the crucial aspect of assessing patient as a whole, follow-up the progression and re-evaluates patient's condition for a new hint. Even though there is no cure yet for SLE, correct and early diagnosis is able to guide for individualized treatment and thus helping in good disease control that warrant better outcome [4].

Keywords: SLE; Schizophrenia; Neuropsychiatric lupus; Altered sensorium

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Introduction

Systemic Lupus Erythematosus (SLE) is a chronic multisystem heterogeneous autoimmune disorder of unknown cause with a broad spectrum of clinical presentation involving virtual organs and tissues [1]. SLE traditionally affecting women and is a rare disease in men [1]. The natural history of the SLE has various severity, follows a relapsing and remitting course due to chronic inflammatory process [1,2]. The signs and symptoms of SLE do not appear concurrently and initial presentation differs according to the target-organ involved. The course is typically by periodic involvement of one organ after another and may present with any combination of clinical features and immunological evidence [1]. The recent updated SLE classification criterion was by American College of Rheumatology (ACR) and the European League against Rheumatism (EULAR) in June 2018, which are designed to accurately identify SLE patients with early disease as early intervention may improve outcomes [5] (Figure 1).

Case Presentation

A 41-year-old single, unemployed man was diagnosed with schizophrenia since 15 years ago. He was under psychiatry follow-up until about 2 years ago when he was transferred to primary healthcare service for continuation of care. He was on intramuscular flupenthixol decanoate injection 40 mg monthly and T. Risperidone 4 mg on night. During routine clinic follow-up, noticed he had discoid rashes over the face, scalp, abdomen and extensor part of bilateral elbow in which initially treated as plaque psoriasis. The clinical examination demonstrated pallor and presence of moderate splenomegaly (Figure 2).

The clinical findings triggered for autoimmune disease screening. Full blood count revealed pancytopenia with Hb 8.4 with TWBC of 1.9 and Platelet of 36. The peripheral blood film reported as hypochromic microcytic anaemia with leucopenia and thrombocytopenia, no suspicious cells and platelet clumping seen. Anti-Nuclear Antibody (ANA) was positive, pattern mix of speckled and centromere with dilution of 1:640. Anti-extractable nuclear antigen showed positive SSA/Ro.

New ACR and EULAR criteria for classification of SLE

All patients classified as having systemic lupus erythematosus must have a serum titer of antinuclear antibody of at least 1:80 on human epithelial-2–positive cells or an equivalent positive test. In addition, a patient must tally at least 10 points from these criteria. A criterion is not counted if it has a more likely explanation than SLE. Occurrence of the criterion only once is sufficient to tally the relevant points, and the time when a patient is positive for one criterion need not overlap with the time when the patient is positive for other criteria. SLE classification requires points from at least one clinical domain, and if a patient is positive for more than one criterion in a domain only the criterion with the highest point value counts:

Clinical domains	Points
Constitutional domain	
Fever	2
Cutaneous domain	
Nonscarring alopecia	2
Oral ulcers	2
Subacute cutaneous or discoid lupus	4
Acute cutaneous lupus	6
Arthritis domain	
Synovitis in at least two joints or tenderness in at least two joints, and at least 30 min of morning stiffness	6
Neurologic domain	
Delirium	2
Psychosis	3
Seizure	5
Serositis domain	
Pleural or pericardial effusion	5
Acute pericarditis	6
Hematologic domain	
Leukopenia	3
Thrombocytopenia	4
Autoimmune hemolysis	4
Renal domain	
Proteinuria >0.5g/24 hr	4
Class II or V lupus nephritis	8
Class III or IV lupus nephritis	10

Immunologic domains	Points
Antiphospholipid antibody domain	
Anticardiolipin IgG >40 GPL or anti-β2GP1 IgG >40 units or lupus anticoagulant	2
Complement proteins domain	
Low C3 or low C4	3
Low C3 and low C4	4
Highly specific antibodies domain	
Anti-dsDNA antibody	6
Anti-Smith antibody	6

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Source: Dr. Johnson

Figure 1:

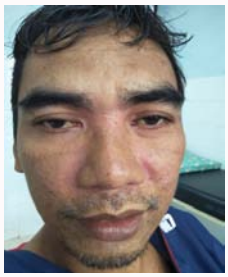

Figure 2:

His erythrocyte sedimentation rate was mildly raised with 35 mm/hour and both serum complement C3 and C4 titre were low.

Retrospectively, his initial presentations at the age of 26 were mood disturbance, cognitive impairment and acute psychosis. Throughout the course of illness, he had several hospitalizations to the psychiatric ward. He was unsure of the onset of skin manifestation and no history of unexplained fever or bleeding diathesis.

He had fulfilled criteria for SLE based on ACR and EULAR classification criteria in which he had positive ANA with titre of 1:640 and total score of 15 points for cutaneous domain (4 points), neurological domain (3 points), hematological domain (4 points) and complement protein domain (4 points). His final diagnosis was Neuropsychiatric SLE (NPSLE) with bone marrow infiltration. He was then referred to rheumatology clinic for further management. Family conference was arranged with counseling and psychotherapy was provided. His condition improved after initiation of oral prednisolone and Hydroxychloroquine (HCQ).

Discussions

The neuropsychiatric manifestation (altered mental state) presented was believed the earliest manifestation of SLE for this man as the auto-antibodies initially attack his brain [2,4,5]. However, at his initial presentation, the case was attributed as primary psychotic disorder that leads to the diagnosis of schizophrenia [2]. The approach for patient presented with altered sensorium should consider the probability of autoimmune disease cause [2,6]. SLE is one of the serious chronic disorder must not be missed as prompt diagnosis with appropriate treatment can shorten the period of flare, reduce the disease activity and prevent further insult to other vital organs.

The neurological domain that was elaborated in the SLE classification criteria is to avoid the pitfall that often missed in the diagnosis [6,7]. The neurological features for neuropsychiatric lupus was established in 1999, recommended by ACR that comprises of 19 cases definition divided into central and peripheral nervous system involvement [6,7]. This patient had psychosis, mood disorder and cognitive dysfunction suggest the central nervous system involvement [2-4,6]. As a result of untreated long-standing illness, the disease has progressed dramatically to involve bone marrow as end-target organ damage as evidence of anaemia of chronic disease and hypersplenism [1].

The previous classification criteria by SLICC in 2012 required fulfillment 4 criteria or more with at least 1 clinical criterion and 1 immunological criterion as minimum requirement [1]. Despite excellent sensitivity and specificity for patients with established disease, their sensitivity in the early disease phase is significantly lower [1].

The recent SLE classification criteria shift toward point system instead of yes or no decision as previous classification. The criteria now rely on ANA titre as an entry criterion and weighted by additional criteria [5]. The ANA have high sensitivity and specificity making an ideal screening test for the diagnosis of SLE. The classification scoring system require ANA test positive with titre of at least 1:80 with other domain of clinical features contribute to total score of at least 10 points [5].

Conclusion

The Murtagh's diagnostic approachis a good practice to achieve accurate and timely diagnosis. SLE should be one of the classical differential diagnosis to be kept in mind because the sign and symptoms are varies and the presentation is unique and dynamic according to the individual. The SLE classification criteria available as a diagnostic tool and checklist for systemic clinical presentation that usually presented at primary care setting. Schizophrenia hence is the diagnosis of exclusion after rule out other possible causes.

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